Section 1

General Orthopaedics

Orthopaedic diagnosis 1

Orthopaedics is concerned with bones, joints, muscles,tendons and nerves – the skeletal system and all that makes it move. Conditions that affect these structures fall into seven easily remembered pairs:

1. Congenital and developmental abnormalities.

2. Infection and inflammation.

3. Arthritis and rheumatic disorders.

4. Metabolic and endocrine disorders.

5. Tumours and lesions that mimic them.

6. Neurological disorders and muscle weakness.

7. Injury and mechanical derangement.

Diagnosis in orthopaedics, as in all of medicine, is the identification of disease. It begins from the very first encounter with the patient and is gradually modified and fine-tuned until we have a picture, not only of a pathological process but also of the functional loss and the disability that goes with it. Understanding evolves from the systematic gathering of information from the history, the physical examination, tissue and organ imaging and special investigations. Systematic, but never mechanical; behind the enquiring mind there should also be what D. H. Lawrence has called ‘the intelligent heart’. It must never be forgotten that the patient has a unique personality, a job and hobbies, a family and a home; all have a bearing upon, and are in turn affected by, the disorder and its treatment.

HISTORY

‘Taking a history’ is a misnomer. The patient tells a story; it is we the listeners who construct a history.The story may be maddeningly disorganized; the history has to be systematic. Carefully and patiently compiled, it can be every bit as informative as examination or laboratory tests. As we record it, certain key words and phrases will inevitably stand out: injury, pain, stiffness, swelling, deformity, instability, weakness, altered sensibility and loss of function or inability to do certain things that were easily accomplished before.Each symptom is pursued for more detail: we need to know when it began, whether suddenly or gradually, spontaneously or after some specific event; how it has changed or progressed; what makes it worse; what makes it better. While listening, we consider whether the story fits some pattern

that we recognize, for we are already thinking of a diagnosis. Every piece of information should be thought of as part of a larger picture which gradually unfolds in our understanding. The surgeonphilosopher Wilfred Trotter (1870–1939) put it well: ‘Disease reveals itself in casual parentheses’.

SYMPTOMS

Pain

Pain is the most common symptom in orthopaedics. It is usually described in metaphors that range from inexpressively bland to unbelievably bizarre – descriptions that tell us more about the patient’s state of mind than about the physical disorder. Yet there are clearly differences between the throbbing pain of an abscess and the aching pain of chronic arthritis, between the ‘burning pain’ of neuralgia and the ‘stabbing pain’ of a ruptured tendon. Severity is even more subjective. High and low pain thresholds undoubtedly exist, but to the patient pain is as bad as it feels, and any system of ‘pain grading’ must take this into account. The main value of estimating severity is in assessing the progress of the disorder or the response to treatment. The commonest method is to invite the patient to mark the severity on an analogue scale of 1–10, with 1 being mild and easily ignored and 10 being totally unbearable. The problem about this type of grading is that patients who have never experienced very severe pain simply do not know what 8 or 9 or 10 would feel like. The following is suggested as a simpler system:

• Grade I (mild) Pain that can easily be ignored.

• Grade II (moderate) Pain that cannot be ignored, interferes with function and needs attention or treatment from time to time.

• Grade III (severe) Pain that is present most of the time, demanding constant attention or treatment.

• Grade IV (excruciating) Totally incapacitating pain.

Referred pain

Pain arising in or near the skin is usually localized accurately. Pain arising in deep structures is more diffuse and is sometimes of unexpected distribution; thus, hip disease may manifest with pain in the knee (so might an obturator hernia). This is not because sensory nerves connect the two sites; it is due to inability of the cerebral cortex to differentiate clearly between sensory messages from separate but embryologically related sites. A common example is ‘sciatica’ – pain at various points in the buttock, thigh and leg, supposedly following the course of the sciatic nerve. Such pain is not necessarily due to pressure on the sciatic nerve or the lumbar nerve roots; it may be ‘referred’ from any one of a number of structures in the lumbar spine, the pelvis and the posterior capsule of the hip joint.

Autonomic pain We are so accustomed to matching pain with some discrete anatomical structure and its known sensory nerve supply that we are apt to dismiss any pain that does not fit the usual pattern as ‘atypical’ or ‘inappropriate’ (i.e. psychologically determined). But pain can also affect the autonomic nerves that accompany the peripheral blood vessels and this is

much more vague, more widespread and often associated with vasomotor and trophic changes. It is poorly understood, often doubted, but nonetheless real.

Stiffness

Stiffness may be generalized (typically in systemic disorders such as rheumatoid arthritis and ankylosing spondylitis) or localized to a particular joint. Patients often have difficulty in distinguishing localized stiffness from painful movement; limitation of movement should never be assumed until verified by examination. Ask when it occurs: regular early morning stiffness of many joints is one of the cardinal symptoms of rheumatoid arthritis, whereas transient stiffness of one or two joints after periods of inactivity is typical of osteoarthritis.

Locking 'Locking' is the term applied to the sudden inability to complete a particular movement. It suggests a mechanical block - for example, due to a loose body or a torn meniscus becoming trapped between the articular surfaces of the knee. Unfortunately, patients tend to use the term for any painful limitation of movement; much more reliable is a history of 'unlocking', when the offending body slips out of the way.

Swelling

Swelling may be in the soft tissues, the joint or the bone; to the patient they are all the same. It is important to establish whether it followed an injury, whether it appeared rapidly (think of a haematoma or a haemarthrosis) or slowly (due to inflammation, a joint effusion, infection or a tumour), whether it is painful (suggestive of acute inflammation, infection or a tumour), whether it is constant or comes and goes, and whether it is increasing in size.

Deformity

The common deformities are described by patients in terms such as round shoulders, spinal curvature, knock knees, bow legs, pigeon toes and flat feet. Deformity of a single bone or joint is less easily described and the patient may simply declare that the limb is 'crooked'. Some 'deformities' are merely variations of the normal (e.g. short stature or wide hips); others disappear spontaneously with growth (e.g. flat feet or bandy legs in an infant). However, if the deformity is progressive, or if it affects only one side of the body while the opposite joint or limb is normal, it may be serious.

Weakness

Generalized weakness is a feature of all chronic illness, and any prolonged joint dysfunction will inevitably lead to weakness of the associated muscles. However, pure muscular weakness – especially if it is confined to one limb or to a single muscle group – is more specific and suggests some neurological or muscle disorder. Patients sometimes say that the limb is 'dead' when it is actually weak, and this can be a source of confusion. Questions should be framed to discover precisely which movements are affected, for this may give important clues, if not to the exact diagnosis at least to the site of the lesion.

Instability

The patient may complain that the joint 'gives way' or 'jumps out of place'. If this happens repeatedly, it suggests abnormal joint laxity, capsular or ligamentous deficiency, or some type of internal derangement such as a torn meniscus or a loose body in the joint. If there is a history of injury, its precise nature is important.

Change in sensibility
Tingling or numbness signifies interference with nerve function – pressure from a neighbouring structure (e.g. a prolapsed intervertebral disc), local ischaemia (e.g. nerve entrapment in a fibro-osseous tunnel) or a peripheral neuropathy. It is important to establish its exact distribution; from this we can tell whether the fault lies in a peripheral nerve or in a nerve root. We should also ask what makes it worse or better; a change in posture might be the trigger, thus focussing attention on a particular site.

Loss of function
Functional disability is more than the sum of individual symptoms and its expression depends upon theneeds of that particular patient. The patient may say 'I can't stand for long' rather than 'I have backache'; or'I can't put my socks on' rather than 'My hip is stiff'. Moreover, what to one patient is merely inconvenient may, to another, be incapacitating. Thus a lawyer or a teacher may readily tolerate a stiff knee provided it is painless, but to a plumber or a parson the same disorder might spell economic or spiritual disaster. One question should elicit the important information: 'What can't you do now that you used to be able to do?'

PAST HISTORY
Patients often forget to mention previous illnesses or accidents, or they may simply not appreciate their relevance to the present complaint. They should be asked specifically about childhood disorders, periods of incapacity and old injuries. A 'twisted ankle' many years ago may be the clue to the onset of osteoarthritis in what is otherwise an unusual site for this condition. Gastrointestinal disease, which in the patient's mind has nothing to do with bones, may be important in the later development of ankylosing spondylitis or osteoporosis. Similarly, certain rheumatic disorders may be suggested by a history of conjunctivitis, iritis, psoriasis or urogenital disease. Metastatic bone disease may erupt many years after a mastectomy for breast cancer. Patients should also be asked about previous medication: many drugs, and especially cortico - steroids, have long-term effects on bone. Alcohol and drug abuse are important, and we must not be afraid to ask about them.

FAMILY HISTORY
Patients often wonder (and worry) about inheriting a disease or passing it on to their children. To the doctor, information about musculoskeletal disorders in the patient's family may help with both diagnosis and counselling.
When dealing with a suspected case of bone or joint infection, ask about communicable diseases, such as tuberculosis or sexually transmitted disease, in other members of the family.
SOCIAL BACKGROUND
No history is complete without enquiry about the patient's background. There are the obvious things such as the level of care and nutrition in children; dietary constraints which may cause specific deficiencies; and, in certain cases, questions about smokinghabits, alcohol consumption and drug abuse, all of which call for a special degree of tact and non judgemental enquiry. Find out details about the patient's work practices, travel and recreation: could the disorder be due to a particular repetitive activity in the home, at work or on the sportsfield? Is the patient subject to any unusual occupational strain? Has he or she travelled to another country where tuberculosis is common?
Finally, it is important to assess the patient's home circumstances and the level of support by family and friends. This will help to answer the question: 'What has the patient lost and what is he or she hoping to regain?'

EXAMINATION

In A Case of Identity Sherlock Holmes has the following conversation with Dr Watson.
Watson: You appeared to read a good deal upon [your client] which was quite invisible to me.
Holmes: Not invisible but unnoticed, Watson.

Some disorders can be diagnosed at a glance: who would mistake the facial appearance of acromegaly or the hand deformities of rheumatoid arthritis for anything else? Nevertheless, even in these cases systematic examination is rewarding: it provides information about the patient's particular disability, as distinct from the clinicopathological diagnosis; it keeps reinforcing good habits; and, never to be forgotten, it lets the patient know that he or she has been thoroughly attended to. The examination actually begins from the moment we set eyes on the patient. We observe his or her general appearance, posture and gait. Can you spot any distinctive feature: Knock-knees? Spinal curvature? A short limb? A paralysed arm? Does he or she appear to be in pain? Do their movements look natural? Do they walk with a limp, or use a stick? A tell-tale gait may suggest a painful hip, an unstable knee or a foot-drop. The clues are endless and the game is played by everyone (qualified or lay) at each new encounter throughout life. In the clinical setting the assessment needs to be more focussed.
When we proceed to the structured examination, the patient must be suitably undressed; no mere rolling up of a trouser leg is sufficient. If one limb is affected, both must be exposed so that they can be compared.
We examine the good limb (for comparison), then the bad. There is a great temptation to rush in with both hands – a temptation that must be resisted. Only by proceeding in a purposeful, orderly way can we avoid missing important signs.
Alan Apley, who developed and taught the system used here, shied away from using long words where short ones would do as well. (He also used to say 'I'm neither an inspector nor a manipulator, and I am definitely not a palpator'.) Thus the traditional clinical routine, inspection, palpation, manipulation, was replaced by look, feel, move. With time his teaching has been extended and we now add test, to include the special manoeuvres we employ in assessing neurological integrity and complex functional attributes.

Look

Abnormalities are not always obvious at first sight. A systematic, step by step process helps to avoid mistakes. Shape and posture The first things to catch one's attention are the shape and posture of the limb or the body or the entire person who is being examined. Is the patient unusually thin or obese? Does the overall posture look normal? Is the spine straight or unusually curved? Are the shoulders level? Are the limbs normally positioned? It is important to look for deformity in three planes, and always compare the affected part with the normal side. In many joint disorders and in most nerve lesions the limb assumes a characteristic posture. In spinal disorders the entire torso may be deformed. Now look more closely for swelling or wasting – one often enhances the appearance of the other! Or is there a definite lump?
Skin- Careful attention is paid to the colour, quality and markings of the skin. Look for bruising, wounds and ulceration. Scars are an informative record of thepast – surgical archaeology, so to speak. Colour reflects vascular status or pigmentation – for example the pallor of ischaemia, the blueness of cyanosis, the redness of inflammation, or the dusky purple of an old bruise. Abnormal creases, unless due to fibrosis, suggest underlying deformity which is not always obvious; tight, shiny skin with no creases is typical of oedema or trophic change.

General survey Attention is initially focussed on the symptomatic or most obviously abnormal area, but we must also look further afield. The patient complains of the joint that is hurting now, but we may see at a glance that several other joints are affected as well.

Feel

Feeling is exploring, not groping aimlessly. Know your anatomy and you will know where to feel for the landmarks; find the landmarks and you can construct a virtual anatomical picture in your mind's eye.

The skin Is it warm or cold; moist or dry; and is sensation normal? The soft tissues Can you feel a lump; if so, what are its characteristics? Are the pulses normal? The bones and joints Are the outlines normal? Is the synovium thickened? Is there excessive joint fluid?

Tenderness -Once you have a clear idea of the structural features in the affected area, feel gently for tenderness. Keep your eyes on the patient's face; a grimace will tell you as much as a grunt. Try to localize any tenderness to a particular structure; if you know precisely where the trouble is, you are halfway to knowing what it is.

Move

'Movement' covers several different activities: active movement, passive mobility, abnormal or unstable movement, and provocative movement.

Active movement Ask the patient to move without your assistance. This will give you an idea of the degree of mobility and whether it is painful or not. Active movement is also used to assess muscle power.

Passive movement Here it is the examiner who moves the joint in each anatomical plane. Note whether there is any difference between the range of active and passive movement. Range of movement is recorded in degrees, starting from zero which, by convention, is the neutral or anatomical position of the joint and finishing where movement stops, due either to pain or anatomical limitation.

Describing the range of movement is often made to seem difficult. Words such as 'full', 'good', 'limited' and 'poor' are misleading. Always cite the range or span, from start to finish, in degrees. For example, 'knee flexion 0–140°' means that the range of flexion is from zero (the knee absolutely straight) through an arc of 140 degrees (the leg making an acute angle with the thigh). Similarly, 'knee flexion 20–90°' means that flexion begins at 20 degrees (i.e. the joint cannot extend fully) and continues only to 90 degrees.

For accuracy you can measure the range of movement with a goniometer, but with practice you will learn to estimate the angles by eye. Normal ranges of movement are shown in chapters dealing with individual joints. What is important is always to compare the symptomatic with the asymptomatic or normal side.

While testing movement, feel for crepitus. Joint crepitus is usually coarse and fairly diffuse; tenosynovial crepitus is fine and precisely localized to the affected tendon sheath.

Unstable movement This is movement which is inherently unphysiological. You may be able to shift orcangulate a joint out of its normal plane of movement, thus demonstrating that the joint is unstable. Such abnormal movement may be obvious (e.g. a wobbly knee); often, though, you have to use special manoeuvres to pick up minor degrees of instability.

Provocative movement One of the most telling clues to diagnosis is reproducing the patient's symptoms by applying a specific, provocative movement. Shoulder pain due to impingement

of the subacromial structures may be ‘provoked’ by moving the joint in a way that is calculated to produce such impingement; the patient recognizes the similarity between this pain and his or her daily symptoms. Likewise, a patient who has had a previous dislocation or subluxation can be vividly reminded of that event by stressing the joint in such a way that it again threatens to dislocate; indeed, merely starting the movement may be so distressing that the patient goes rigid with anxiety at the anticipated result – this is aptly called the apprehension test.

Test

The apprehension test referred to in the previous paragraph is one of several clinical tests that are used to elicit suspected abnormalities: some examples are Thomas’ test for flexion deformity of the hip, Trendelenburg’s test for instability of the hip, McMurray’s test for a torn meniscus of the knee, Lachman’s test for cruciate ligament instability and various tests for intra-articular fluid. These and others are described in the relevant chapters in Section 2. Tests for muscle tone, motor power, reflexes and various modes of sensibility are part and parcel of neurological examination.

Caveat

We recognize that the sequence set out here may sometimes have to be modified. We may need to ‘move’ before we ‘look’: an early scoliotic deformity of the spine often becomes apparent only when the patient bends forwards. The sequence may also have to be altered because a patient is in severe pain or disabled: you would not try to move a limb at all in someone with a suspected fracture when an x-ray can provide the answer. When examining a child you may have to take your chances with look or feel or move whenever you can!

TERMINOLOGY

Colloquial terms such as front, back, upper, lower, inner aspect, outer aspect, bow legs, knock knees have the advantage of familiarity but are not applicable to every situation. Universally acceptable anatomical definitions are therefore necessary in describing physical attributes. Bodily surfaces, planes and positions are always described in relation to the **anatomical position** - as if the person were standing erect, facing the viewer, legs together with the knees pointing directly forwards, and arms held by the sides with the palms facing forwards.

The principal planes of the body are named **sagittal, coronal and transverse**; they define the direction across which the body (or body part) is viewed in any description.

Sagittal planes, parallel to each other, pass vertically through the body from front to back; the **midsagittal** or **median plane** divides the body into right and left halves.

Coronal planes are also orientated vertically, corresponding to a frontal view, at right angles to the saggital planes; **transverse planes** pass horizontally across the body.

Anterior signifies the frontal aspect and **posterior** the rear aspect of the body or a body part. The terms **ventral** and **dorsal** are also used for the front and the back respectively. Note, though, that the use of these terms is somewhat confusing when it comes to the foot: here the upper surface is called the **dorsum** and the sole is called the **plantar surface**.

Medial means facing towards the median plane or midline of the body, and **lateral** away from the median plane. These terms are usually applied to a limb, the clavicle or one half of the pelvis. Thus the inner aspect of the thigh lies on the medial side of the limb and the outer part of the thigh lies on the lateral side. We could also say that the little finger lies on the medial or **ulnar side** of the hand and the thumb on the lateral or **radial side** of the hand.

Proximal and **distal** are used mainly for parts ofthe limbs, meaning respectively the upper end and the lower end as they appear in the anatomical position. Thus the knee joint is formed by the distal end of the femur and the proximal end of the tibia.
Axial alignment describes the longitudinal arrangement of adjacent limb segments or parts of a single bone.
The knees and elbows, for example, are normally angulated slightly outwards (**valgus)** while the opposite - 'bow legs' - is more correctly described as **varus** (see on page 13, under Deformity). Angulation in the middle of a long bone would always be regarded as abnormal.
Rotational alignment refers to the tortile arrangement of segments of a long bone (or an entire limb) around a single longitudinal axis. For example, in the anatomical position the patellae face forwards while the feet are turned slightly outwards; a marked difference in rotational alignment of the two legs is abnormal.
Flexion and extension are joint movements in the sagittal plane, most easily imagined in hinge joints like the knee, elbow and the joints of the fingers and toes.
In elbows, knees, wrists and fingers flexion means bending the joint and extension means straightening it. In shoulders and hips flexion is movement in an anterior direction and extension is movement posteriorwards.
In the ankle flexion is also called **plantarflexion** (pointing the foot downwards) and extension is called **dorsiflexion** (drawing the foot upwards). Thumb movements are the most complicated and are described in Chapter 16.
Abduction and adduction are movements in the coronal plane, away from or towards the median plane. Not quite for the fingers and toes, though: here abduction and adduction mean away from and towards the longitudinal midline of the hand or foot!
Lateral rotation and medial rotation are twisting movements, outwards and inwards, around a longitudinal axis.
Pronation and supination are also rotatory movements, but the terms are applied only to movements of the forearm and the foot.
Circumduction is a composite movement made up of a rhythmic sequence of all the other movements. It is possible only for ball-and-socket joints such as the hip and shoulder.
Specialized movements such as opposition of the thumb, lateral flexion and rotation of the spine, and inversion or eversion of the foot, will be described in the relevant chapters.

EXAMINING INFANTS AND CHILDREN

Paediatric practice requires special skills. You may have no first-hand account of the symptoms; a baby screaming with pain will tell you very little, and overanxious parents will probably tell you too much. When examining the child, be flexible. If he or she is moving a particular joint, take your opportunity to examine movement then and there. You will learn much more by adopting methods of play than by applying a rigid system of examination. And leave any test for tenderness until last!

INFANTS AND SMALL CHILDREN

The baby should be undressed, in a warm room, and placed on the examining couch. Look carefully for birthmarks, deformities and abnormal movements –or absence of movement. If there is no urgency or distress, take time to examine the head and neck, including facial features which may be characteristic of specific dysplastic syndromes. The back and limbs are then examined for abnormalities of position or shape.
Examining for joint movement can be difficult. Active movements can often be stimulated by gently stroking the limb. When testing for passive mobility, be careful to avoid frightening or hurting the child. In the neonate, and throughout the first two years of life, examination of the

hips is mandatory, even if the child appears to be normal. This is to avoid missing the subtle signs of developmental dysplasia of the hips (DDH) at the early stage when treatment is most effective.
It is also important to assess the child's general development by testing for the normal milestones which are expected to appear during the first two years of life.

NORMAL DEVELOPMENTAL MILESTONES
Newborn Grasp reflex present
Morrow reflex present
3–6 months Holds head up unsupported
6–9 months Able to sit up
9–12 months Crawling and standing up
9–18 months Walking
18–24 months Running

OLDER CHILDREN
Most children can be examined in the same way as adults, though with different emphasis on particular physical features. Posture and gait are very important; subtle deviations from the norm may herald the appearance of serious abnormalities such as scoliosis or neuromuscular disorders, while more obvious 'deformities' such as knock knees and bow legs may be no more than transient stages in normal development; similarly with mild degrees of 'flat feet' and 'pigeon toes'. More complex variations in posture and gait patterns, when the child sits and walks with the knees turned inwards (medially rotated) or outwards (laterally rotated) are usually due to anteversion or retroversion of the femoral necks, sometimes associated with compensatory rotational 'deformities' of the femora and tibiae. Seldom need anything be done about this; the condition usually improves as the child approaches puberty and only if the gait is very awkward would one consider performing corrective osteotomies of the femora.

PHYSICAL VARIATIONS AND DEFORMITIES
JOINT LAXITY
Children's joints are much more mobile than those of most adults, allowing them to adopt postures that would be impossible for their parents. An unusual degree of joint mobility can also be attained by adults willing to submit to rigorous exercise and practice, as witness the performances of professional dancers and athletes, but in most cases, when the exercises stop, mobility gradually reverts to the normal range.
Persistent generalized joint hypermobility occurs in about 5% of the population and is inherited as a simple mendelian dominant. Those affected describe themselves as being 'double-jointed': they can hyperextend their metacarpophalangeal joints beyond a right angle, hyperextend their elbows and knees and bend over with knees straight to place their hands flat on the ground; some can even 'do the splits' or place their feet behind their neck!
It is doubtful whether these individuals should be considered 'abnormal'. However, epidemiological studies have shown that they do have a greater than usual tendency to recurrent dislocation (e.g. of the shoulder or patella). Some experience recurrent episodes of aching around the larger joints; however, there is no convincing evidence that hypermobility by itself predisposes to osteoarthritis. Generalized hypermobility is not usually associated with any obvious disease, but severe laxity is a feature of certain rare connective tissue disorders such as Marfan's syndrome, Ehlers–Danlos syndrome, Larsen's disease and osteogenesis imperfecta.

Deformity

The boundary between variations of the normal and physical deformity is blurred. Indeed, in the development of species, what at one point of time might have been seen as a deformity could over the ages have turned out to be so advantageous as to become essential for survival.

So too in humans. The word 'deformity' is derived from the Latin for 'misshapen', but the range of 'normal shape' is so wide that variations should not automatically be designated as deformities, and some undoubted 'deformities' are not necessarily pathological; for example, the generally accepted cut-off points for 'abnormal' shortness or tallness are arbitrary and people who in one population might be considered abnormally short or abnormally tall could, in other populations, be seen as quite ordinary. However, if one leg is short and the other long, no-one would quibble with the use of the word 'deformity'!

Specific terms are used to describe the 'position' and 'shape' of the bones and joints. Whether, in any particular case, these amount to 'deformity' will be determined by additional factors such as the extent to which they deviate from the norm, symptoms to which they give rise, the presence or absence of instability and the degree to which they interfere with function.

Varus and valgus It seems pedantic to replace 'bow legs' and 'knock knees' with 'genu varum' and 'genu valgum', but comparable colloquialisms are not available for deformities of the elbow, hip or big toe; and, besides, the formality is justified by the need for clarity and consistency. Varus means that the part distal to the joint in question is displaced towards the median plane, valgus away from it.

Kyphosis and lordosis Seen from the side, the normal spine has a series of curves: convex posteriorly in the thoracic region (kyphosis), and convex anteriorly in the cervical and lumbar regions (lordosis). Excessive curvature constitutes kyphotic or lordotic deformity (also sometimes referred to as hyperkyphosis and hyperlordosis). Colloquially speaking, excessive thoracic kyphosis is referred to as 'round-shouldered'.

Scoliosis Seen from behind, the spine is straight. Any curvature in the coronal plane is called scoliosis. The position and direction of the curve are specified by terms such as thoracic scoliosis, lumbar scoliosis, convex to the right, concave to the left, etc.

Postural deformity A postural deformity is one which the patient can, if properly instructed, correct voluntarily: e.g. thoracic 'kyphosis' due to slumped shoulders.

Postural deformity may also be caused by temporary muscle spasm.

Structural deformity A deformity which results from a permanent change in anatomical structure cannot be voluntarily corrected. It is important to distinguish postural scoliosis from structural (fixed) scoliosis. The former is non-progressive and benign; the latter is usually progressive and may require treatment.

'Fixed deformity' This term is ambiguous. It seems to mean that a joint is deformed and unable to move.

Not so – it means that one particular movement cannot be completed. Thus the knee may be able to flex fully but not extend fully – at the limit of its extension it is still 'fixed' in a certain amount of flexion. This would be called a 'fixed flexion deformity'.

CAUSES OF JOINT DEFORMITY

There are six basic causes of joint deformity:

1. Contracture of the overlying skin This is seen typically when there is severe scarring across the flexor aspect of a joint, e.g. due to a burn or following surgery.
2. Contracture of the subcutaneous fascia The classical example is Dupuytren's contracture in the palm of the hand.
3. Muscle contracture Fibrosis and contracture of muscles that cross a joint will cause a fixed deformity of the joint. This may be due to deep infection or fibrosis following ischaemic necrosis (Volkmann's ischaemic contracture).
4. Muscle imbalance Unbalanced muscle weakness or spasticity will result in joint deformity which, if not corrected, will eventually become fixed. This is seen most typically in poliomyelitis and cerebral palsy. Tendon rupture, likewise, may cause deformity.
5. Joint instability Any unstable joint will assume a 'deformed' position when subjected to force.
6. Joint destruction Trauma, infection or arthritis may destroy the joint and lead to severe deformity.

CAUSES OF BONE DEFORMITY
Bone deformities in small children are usually due to genetic or developmental disorders of cartilage and bone growth; some can be diagnosed in utero by special imaging techniques (e.g. achondroplasia); some become apparent when the child starts to walk, or later still during one of the growth spurts (e.g. hereditary multiple exostosis); and some only in early adulthood (e.g. multiple epiphyseal dysplasia). There are a myriad genetic disorders affecting the skeleton, yet any one of these conditions is rare. The least unusual of them are described in Chapter 8. Acquired deformities in children may be due to fractures involving the physis (growth plate); ask about previous injuries. Other causes include rickets,endocrine disorders, malunited diaphyseal fractures and tumours. Acquired deformities of bone in adults are usually the result of previous malunited fractures. However, causes such as osteomalacia, bone tumours and Paget's disease should always be considered.

BONY LUMPS
A bony lump may be due to faulty development, injury, inflammation or a tumour. Although x-ray examination is essential, the clinical features can be highly informative.
Size A large lump attached to bone, or a lump that is getting bigger, is nearly always a tumour.
Site A lump near a joint is most likely to be a tumour (benign or malignant); a lump in the shaft may be fracture callus, inflammatory new bone or a tumour.
Margin A benign tumour has a well-defined margin; malignant tumours, inflammatory lumps and callus have a vague edge.
Consistency A benign tumour feels bony hard; malignant tumours often give the impression that they can be indented.
Tenderness Lumps due to active inflammation, recent callus or a rapidly growing sarcoma are tender.
Multiplicity Multiple bony lumps are uncommon: they occur in hereditary multiple exostosis and in Ollier's disease.

JOINT STIFFNESS
The term 'stiffness' covers a variety of limitations.
We consider three types of stiffness in particular: (1) all movements absent; (2) all movements limited; (3) one or two movements limited.
All movements absent Surprisingly, although movement is completely blocked, the patient may retain such good function that the restriction goes unnoticed until the joint is examined.

Surgical fusion is called ‘arthrodesis’; pathological fusion is called ‘ankylosis’. Acute suppurative arthritis typically ends in bony ankylosis; tuberculous arthritis heals by fibrosis and causes fibrous ankylosis – not strictly a ‘fusion’ because there may still be a small jog of movement.

All movements limited- After severe injury, movementmay be limited as a result of oedema and bruising.

Later, adhesions and loss of muscle extensibility may perpetuate the stiffness.

With active inflammation all movements are restricted and painful and the joint is said to be ‘irritable’.

In acute arthritis spasm may prevent all but a few degrees of movement.

In osteoarthritis the capsule fibroses and movements become increasingly restricted, but pain occurs only at the extremes of motion.

Some movements limited When one particular movement suddenly becomes blocked, the cause is usually mechanical. Thus a torn and displaced meniscus may prevent extension of the knee but not flexion.

Bone deformity may alter the arc of movement, such that it is limited in one direction (loss of abduction in coxa vara is an example) but movement in the opposite direction is full or even increased.

These are all examples of ‘fixed deformity’.

BONE MINERAL DENSITOMETRY

Bone mineral density (BMD) measurement is now widely used in identifying patients with osteoporosis and an increased risk of osteoporotic fractures.

Various techniques have been developed, including radiographic absorptiometry (RA), quantitative computed tomography (QCT) and quantitative ultrasonometry (QUS). However, the most widely used technique is dual energy x-ray absorptiometry (DXA). RA uses conventional radiographic equipment and measures bone density in the phalanges. QCT measures trabecular bone density in vertebral bodies, but is not widely available and involves a higher dose of ionizing radiation than DXA. QUS assesses bone mineral density in the peripheral skeleton (e.g. the wrist and calcaneus) by measuring both the attenuation of ultrasound and the variation of speed of sound through the bone.

DXA employs columnated low-dose x-ray beams of two different energy levels in order to distinguish the density of bone from that of soft tissue. Although this involves the use of ionizing radiation, it is an extremely low dose. A further advantage of DXA is the development of a huge international database that allows expression of bone mineral density values in comparison to both an age and sex matched population (Z score) and also to the peak adult bone mass (T score). The T score in particular allows calculation of relative fracture risk. Individual values for both the lumbar spine and hips are obtained as there is often a discrepancy between these two sites and the fracture risk is more directly related to the value at the target area. By World Health Organization (WHO) criteria, T scores of <–1.0 indicate ‘osteopenia’ and T scores of <–2.5 indicate ‘osteoporosis’.

Infection 2

Micro-organisms may reach the musculoskeletal tissues by (a) direct introduction through the skin (a pinprick, an injection, a stab wound, a laceration, an open fracture or an operation), (b) direct spread from a contiguous focus of infection, or (c) indirect spread via the blood stream from a distant site such as the nose or mouth, the respiratory tract, the bowel or the genitourinary tract.
Depending on the type of invader, the site of infection and the host response, the result may be a pyogenic osteomyelitis, a septic arthritis, a chronic granulomatous reaction (classically seen in tuberculosis of either bone or joint), or an indolent response to an unusual organism (e.g. a fungal infection). Softtissue infections range from superficial wound sepsis to widespread cellulitis and life-threatening necrotizing cellulitis. Parasitic lesions such as hydatid disease also are considered in this chapter, although these are infestations rather than infections.

GENERAL ASPECTS OF INFECTION

Infection – as distinct from mere residence of microorganisms – is a condition in which pathogenic organisms multiply and spread within the body tissues. This usually gives rise to an acute or chronic inflammatory reaction, which is the body's way of combating the invaders and destroying them, or at least immobilizing them and confining them to a single area. The signs of inflammation are recounted in the classical mantra: redness, swelling, heat, pain and loss of function.
In one important respect, bone infection differs from soft-tissue infection: since bone consists of a collection of rigid compartments, it is more susceptible than soft tissues to vascular damage and cell death from the build-up of pressure in acute inflammation. Unless it is rapidly suppressed, bone infection will inevitably lead to necrosis.
Host susceptibility to infection is increased by (a) local factors such as trauma, scar tissue, poor circulation, diminished sensibility, chronic bone or joint disease and the presence of foreign bodies, as well as (b) systemic factors such as malnutrition, general illness, debility, diabetes, rheumatoid disease, corticosteroid administration and all forms of immunosuppression, either acquired or induced. Resistance is also diminished in the very young and the very old. Bacterial colonization and resistance to antibiotics is enhanced by the ability of certain microbes (including Staphylococcus) to adhere to avascular bone surfaces and foreign implants, protected from both host defences and antibiotics by a protein-polysaccharide slime (glycocalyx).
Acute pyogenic bone infections are characterized by the formation of pus – a concentrate of defunct leucocytes, dead and dying bacteria and tissue debris – which is often localized in an abscess. Pressure builds up within the abscess and infection may then extend into a contiguous joint or through the cortex and along adjacent tissue planes. It may also spread further afield via lymphatics (causing lymphangitis and lymphadenopathy) or via the blood stream (bacteraemia and septicaemia). An accompanying systemic reaction varies from a vague feeling of lassitude with mild pyrexia to severe illness, fever, toxaemia and shock. The generalized effects are due to the release of bacterial enzymes and endotoxins as well as cellular breakdown products from the host tissues.

Chronic pyogenic infection may follow on unresolved acute infection and is characterized by persistence of the infecting organism in pockets of necrotic tissue. Purulent material accumulates and may be discharged through sinuses at the skin or a poorly healed wound. Factors which favour this outcome are the presence of damaged muscle, dead bone or a foreign implant, diminished local blood supply and a weak host response. Resistance is likely to be depressed in the very young and the very old, in states of malnutrition or immunosuppression, and in certain diseases such as diabetes and leukaemia.

Factors predisposing to bone infection
Malnutrition and general debility
Diabetes mellitus
Corticosteroid administration
Immune deficiency
Immunosuppressive drugs
Venous stasis in the limb
Peripheral vascular disease
Loss of sensibility
Iatrogenic invasive measures
Trauma

Chronic non-pyogenic infection may result from invasion by organisms that produce a cellular reaction leading to the formation of granulomas consisting largely of lymphocytes, modified macrophages and multinucleated giant cells; this type of granulomatous infection is seen most typically in tuberculosis.
Systemic effects are less acute but may ultimately be very debilitating, with lymphadenopathy, splenomegaly and tissue wasting.
The principles of treatment are: (1) to provide analgesia and general supportive measures; (2) to rest the affected part; (3) to identify the infecting organism and administer effective antibiotic treatment or chemotherapy; (4) to release pus as soon as it is detected; (5) to stabilize the bone if it has fractured; (6) to eradicate avascular and necrotic tissue; (7) to restore continuity if there is a gap in the bone; and (8) to maintain soft-tissue and skin cover. Acute infections, if treated early with effective antibiotics, can usually be cured. Once there is pus and bone necrosis, operative drainage will be needed.

ACUTE HAEMATOGENOUS OSTEOMYELITIS

Aetiology and pathogenesis

Acute haematogenous osteomyelitis is mainly a disease of children. When adults are affected it is usually because their resistance is lowered. Trauma may determine the site of infection, possibly by causing a small haematoma or fluid collection in a bone, in patients with concurrent bacteraemia. The incidence of acute haematogenous osteo - myelitis in western European children is thought to have declined in recent years, probably a reflection of improving social conditions. A study from Glasgow, Scotland, covering the period 1990–99, suggests that it is less than 3 cases per 100 000 per year (Blyth et al., 2001). However, it is almost certainly much higher among less affluent populations.
The causal organism in both adults and children is usually Staphylococcus aureus (found in over 70% of cases), and less often one of the other Gram-positive cocci, such as the Group A beta-haemolytic streptococcus (Streptococcus pyogenes) which is found in chronic skin infections, as well as Group B streptococcus (especially in new-born babies) or the alphahaemolytic diplococcus S. pneumoniae. In children between 1 and 4 years of age the

Gramnegative Haemophilus influenzae used to be a fairly common pathogen for osteomyelitis and septic arthritis, but the introduction of H. influenzae type B vaccination about 20 years ago has been followed by a much reduced incidence of this infection in many countries. In recent years its place has been taken by Kingella kingae, mainly following upper respiratory infection in young children. Other Gram-negative organisms (e.g. Escherichia coli, Pseudomonas aeruginosa, Proteus mirabilis and the anaerobic Bacteroides fragilis) occasionally cause acute bone infection. Curiously, patients with sickle-cell disease are prone to infection by Salmonella typhi.
Anaerobic organisms (particularly Peptococcus magnus) have been found in patients with osteomyelitis, usually as part of a mixed infection. Unusual organisms are more likely to be found in heroin addicts and as opportunistic pathogens in patients with compromised immune defence mechanisms.
The blood stream is invaded, perhaps from a minor skin abrasion, treading on a sharp object, an injection point, a boil, a septic tooth or – in the newborn – from an infected umbilical cord. In adults the source of infection may be a urethral catheter, an indwelling arterial line or a dirty needle and syringe.
In children the infection usually starts in the vascular metaphysis of a long bone, most often in the proximal tibia or in the distal or proximal ends of the femur. Predilection for this site has traditionally been attributed to the peculiar arrangement of the blood vessels in that area (Trueta, 1959): the non-anastomosing terminal branches of the nutrient artery twist back in hairpin loops before entering the large network of sinusoidal veins; the relative vascular stasis and consequent lowered oxygen tension are believed to favour bacterial colonization. It has also been suggested that the structure of the fine vessels in the hypertrophic zone of the physis allows bacteria more easily to pass through and adhere to type 1 collagen in that area (Song and Sloboda, 2001).
In infants, in whom there are still anastomoses between metaphyseal and epiphyseal blood vessels, infection can also reach the epiphysis.
In adults, haematogenous infection accounts for only about 20% of cases of osteomyelitis, mostly affecting the vertebrae.
Staphylococcus aureus is the commonest organism but Pseudomonas aeruginosa often appears in patients using intravenous drugs. Adults with diabetes, who are prone to soft-tissue infections of the foot, may develop contiguous bone infection involving a variety of organisms.

Pathology
Acute haematogenous osteomyelitis shows a characteristic progression marked by inflammation, suppuration, bone necrosis, reactive new bone formation and, ultimately, resolution and healing or else intractable chronicity. However, the pathological picture varies considerably, depending on the patient's age, the site of infection, the virulence of the organism and the host response.
Acute osteomyelitis in children The 'classical' picture is seen in children between 2 and 6 years. The earliest change in the metaphysis is an acute inflammatory reaction with vascular congestion, exudation of fluid and infiltration by polymorphonuclear leucocytes. The intraosseous pressure rises rapidly, causing intense pain, obstruction to blood flow and intravascular thrombosis. Even at an early stage the bone tissue is threatened by impending ischaemia and resorption due to a combination of phagocytic activity and the local accumulation of cytokines, growth factors, prostaglandin and bacterial enzymes. By the second or third day, pus forms within the bone and forces its way along the Volkmann canals to the surface where it produces a subperiosteal abscess. This is much more evident in children, because of the relatively loose attachment of the periosteum, than in adults. From

the subperiosteal abscess pus can spread along the shaft, to re-enter the bone at another level or burst into the surrounding soft tissues. The developing physis acts as a barrier to direct spread towards theepiphysis, but where the metaphysis is partly intracapsular (e.g. at the hip, shoulder or elbow) pus may discharge through the periosteum into the joint. The rising intraosseous pressure, vascular stasis, small-vessel thrombosis and periosteal stripping increasingly compromise the blood supply; by the end of a week there is usually microscopic evidence of bone death. Bacterial toxins and leucocytic enzymes also may play their part in the advancing tissue destruction. With the gradual ingrowth of granulation tissue the boundary between living and devitalized bone becomes defined. Pieces of dead bone may separate as sequestra varying in size from mere spicules to large necrotic segments of the cortex in neglected cases.

Macrophages and lymphocytes arrive in increasing numbers and the debris is slowly removed by a combination of phagocytosis and osteoclastic resorption. A small focus in cancellous bone may be completely resorbed, leaving a tiny cavity, but a large cortical or cortico-cancellous sequestrum will remain entombed, inaccessible to either final destruction or repair.

Another feature of advancing acute osteomyelitis is new bone formation. Initially the area around the infected zone is porotic (probably due to hyperaemia and osteoclastic activity) but if the pus is not released, either spontaneously or by surgical decompression, new bone starts forming on viable surfaces in the bone and from the deep layers of the stripped periosteum. This is typical of pyogenic infection and fine streaks of subperiosteal new bone usually become apparent on x-ray by the end of the second week. With time this new bone thickens to form a casement, or involucrum, enclosing the sequestrum and infected tissue. If the infection persists, pus and tiny sequestrated spicules of bone may discharge through perforations (cloacae) in the involucrum and track by sinuses to the skin surface. If the infection is controlled and intraosseous pressure released at an early stage, this dire progress can be halted. The bone around the zone of infection becomes increasingly dense; this, together with the periosteal reaction, results in thickening of the bone. In some cases the normal anatomy may eventually be reconstituted; in others, though healing is sound, the bone is left permanently deformed. If healing does not occur, a nidus of infection may remain locked inside the bone, causing pus and sometimes bone debris to be discharged intermittently through a persistent sinus (or several sinuses). The infection has now lapsed into chronic osteomyelitis, which may last for many years.

Acute osteomyelitis in infants The early features of acute osteomyelitis in infants are much the same as those in older children. However, a significant difference, during the first year of life, is the frequency with which the metaphyseal infection spreads to the epiphysis and from there into the adjacent joint. In the process, the physeal anlage may be irreparably damaged, further growth at that site is severely retarded and the joint will be permanently deformed. How this comes about is still argued over. Following Trueta (1957) it has long been held that, during the first 6–9 months of life, small metaphyseal vessels penetrate the physeal cartilage and this permits the infection to spread into the cartilaginous epiphysis. Others have disagreed with this hypothesis (Chung, 1976), but what is indisputable is that during infancy osteomyelitis and septic arthritis often go together. Another feature in infants is an unusually exuberant periosteal reaction resulting in sometimes bizarre new bone formation along the diaphysis; fortunately, with longitudinal growth and remodelling the diaphyseal anatomy is gradually restored.

Acute osteomyelitis in adults Bone infection in the adult usually follows an open injury, an operation or spread from a contiguous focus of infection (e.g. a neuropathic ulcer or an infected diabetic foot). True haematogenous osteomyelitis is uncommon and when it does

occur it usually affects one of the vertebrae (e.g. following a pelvic infection) or a small cuboidal bone. A vertebral infection may spread through the end-plate and the intervertebral disc into an adjacent vertebral body. If a long bone is infected, the abscess is likely to spread within the medullary cavity, eroding the cortex and extending into the surrounding soft tissues. Periosteal new bone formation is less obvious than in childhood and the weakened cortex may fracture. If the bone end becomes involved there is a risk of the infection spreading into an adjacent joint. The outcome is often a gradual slide towards subacute and chronic osteomyelitis.

Clinical features

Children The patient, usually a child over 4 years, presents with severe pain, malaise and a fever; in neglected cases, toxaemia may be marked. The parents will have noticed that he or she refuses to use one limb or to allow it to be handled or even touched. There may be a recent history of infection: a septic toe, a boil, a sore throat or a discharge from the ear. Typically the child looks ill and feverish; the pulse rate is likely to be over 100 and the temperature is raised. The limb is held still and there is acute tenderness near one of the larger joints (e.g. above or below the knee, in the popliteal fossa or in the groin). Even the gentlest manipulation is painful and joint movement is restricted ('pseudoparalysis'). Local redness, swelling, warmth and oedema are later signs and signify that pus has escaped from the interior of the bone. Lymphadenopathy is common but non-specific. It is important to remember that all these features may be attenuated if antibiotics have been administered.
Infants In children under a year old, and especially in the newborn, the constitutional disturbance can bemisleadingly mild; the baby simply fails to thrive and is drowsy but irritable. Suspicion should be aroused by a history of birth difficulties, umbilical artery catheterization or a site of infection (however mild) such as an inflamed intravenous infusion point or even a heel puncture. Metaphyseal tenderness and resistance to joint movement can signify either osteomyelitis or septic arthritis; indeed, both may be present, so the distinction hardly matters. Look for other sites – multiple infection is not uncommon, especially in babies who acquire the infection in hospital. Radionuclide bone scans may help to discover additional sites.
Adults The commonest site for haematogenous infection is the thoracolumbar spine. There may be a history of some urological procedure followed by a mild fever and backache. Local tenderness is not very marked and it may take weeks before x-ray signs appear; when they do appear the diagnosis may still need to be confirmed by fine-needle aspiration and bacteriological culture. Other bones are occasionally involved, especially if there is a background of diabetes, malnutrition, drug addiction, leukaemia, immunosuppressive therapy or debility.
In the very elderly, and in those with immune deficiency, systemic features are mild and the diagnosis is easily missed.

Laboratory investigations
The most certain way to confirm the clinical diagnosis is to aspirate pus or fluid from the metaphyseal subperiosteal abscess, the extraosseous soft tissues or an adjacent joint. This is done using a 16- or 18-gauge trocar needle. Even if no pus is found, a smear of the aspirate is examined immediately for cells and organisms; a simple Gram stain may help to identify the type of infection and assist with the initial choice of antibiotic. A sample is also sent for detailed microbiological examination and tests for sensitivity to antibiotics. Tissue aspiration will give a positive result in over 60% of cases; blood cultures are positive in less

than half the cases of proven infection. The C-reactive protein (CRP) values are usually elevated within 12–24 hours and the erythrocyte sedimentation rate (ESR) within 24–48 hours after the onset of symptoms. The white blood cell (WBC) count rises and the haemoglobin concentration may be diminished. In the very young and the very old these tests are less reliable and may show values within the range of normal.
Antistaphylococcal antibody titres may be raised. This test is useful in atypical cases where the diagnosis is in doubt. Osteomyelitis in an unusual site or with an unusual organism should alert one to the possibility of heroin addiction, sickle-cell disease (Salmonella may be cultured from the faeces) or deficient host defence mechanisms including HIV infection.

CARDINAL FEATURES OF ACUTE
OSTEOMYELITIS IN CHILDREN
Pain
Fever
Refusal to bear weight
Elevated white cell count
Elevated ESR
Elevated CRP

Differential diagnosis
Cellulitis This is often mistaken for osteomyelitis. There is widespread superficial redness and lymphangitis. The source of skin infection may not be obvious and should be searched for (e.g. on the sole or between the toes). If doubt remains about the diagnosis,
MRI will help to distinguish between bone infection and soft-tissue infection. The organism is usually staphylococcus or streptococcus. Mild cases will respond to high dosage oral antibiotics; severe cases need intravenous antibiotic treatment.
Acute suppurative arthritis Tenderness is diffuse, and movement at the joint is completely abolished by muscle spasm. In infants the distinction between metaphyseal osteomyelitis and septic arthritis of the adjacent joint is somewhat theoretical, as both often
coexist. A progressive rise in C-reactive protein values over 24–48 hours is said to be suggestive of concurrent septic arthritis (Unkila-Kallis et al., 1994). Streptococcal necrotizing myositis Group A betahaemolytic streptococci (the same organisms which
are responsible for the common ‘sore throat’) occasionally invade muscles and cause an acute myositis which, in its early stages, may be mistaken for cellulitis or osteomyelitis. Although the condition is rare, it should be kept well to the foreground in the differential diagnosis because it may rapidly spiral out of control towards muscle necrosis, septicaemia and death. Intense pain and board-like swelling of the limb in a patient with fever and a general feeling of illness are warning signs of a medical emergency. MRI will reveal muscle swelling and possibly signs of tissue breakdown.
Immediate treatment with intravenous antibiotics is essential. Surgical debridement of necrotic tissue – and sometimes even amputation – may be
needed to save a life.
Acute rheumatism The pain is less severe and it tends to flit from one joint to another. There may also be signs of carditis, rheumatic nodules or erythema marginatum.
Sickle-cell crisis The patient may present with features indistinguishable from those of acute osteomyelitis. In areas where Salmonella is endemic it would be wise to treat such patients with suitable antibiotics until infection is definitely excluded.
Gaucher’s disease ‘Pseudo-osteitis’ may occur with features closely resembling those of osteomyelitis. The diagnosis is made by finding other stigmata of the disease, especially enlargement of the spleen and liver.

Treatment

If osteomyelitis is suspected on clinical grounds, blood and fluid samples should be taken for laboratory investigation and then treatment started immediately without waiting for final confirmation of the diagnosis. There are four important aspects to the management of the patient:

- Supportive treatment for pain and dehydration.
- Splintage of the affected part.
- Appropriate antimicrobial therapy.
- Surgical drainage.

GENERAL SUPPORTIVE TREATMENT

The distressed child needs to be comforted and treated for pain. Analgesics should be given at repeated intervals without waiting for the patient to ask for them. Septicaemia and fever can cause severe dehydration and it may be necessary to give fluid intravenously.

SPLINTAGE

Some type of splintage is desirable, partly for comfort but also to prevent joint contractures. Simple skin traction may suffice and, if the hip is involved, this also helps to prevent dislocation. At other sites a plaster slab or half-cylinder may be used but it should not obscure the affected area.

ANTIBIOTICS

Blood and aspiration material are sent immediately for examination and culture, but the prompt intravenous administration of antibiotics is so vital that treatment should not await the result. Initially the choice of antibiotics is based on the findings from direct examination of the pus smear and the clinician's experience of local conditions – in other words, a 'best guess' at the most likely pathogen.

Staphylococcus aureus is the most common at all ages, but treatment should provide cover also for other bacteria that are likely to be encountered in each age group; a more appropriate drug which is also capable of good bone penetration can be substituted, if necessary, once the infecting organism is identified and its antibiotic sensitivity is known. Factors such as the patient's age, general state of resistance, renal function, degree of toxaemia and previous history of allergy must be taken into account. The following recommendations are offered as a guide.

- Neonates and infants up to 6 months of age Initial antibiotic treatment should be effective against penicillin-resistant Staphylococcus aureus, Group B streptococcus and Gram-negative organisms. Drugs of choice are flucloxacillin plus a third-generation cephalosporin like cefotaxime. Alternatively, effective empirical treatment can be provided by a combination of flucloxacillin (for penicillin-resistant staphylococci), benzylpenicillin (for Group B streptococci) and gentamicin (for Gram-negative organisms).
- Children 6 months to 6 years of age Empirical treatment in this age group should include cover against Haemophilus influenzae, unless it is known for certain that the child has had an anti-haemophilus vaccination. This is best provided by a combination of intravenous flucloxacillin and cefotaxime or cefuroxime.
- Older children and previously fit adults The vast majority in this group will have a staphylococcal infection and can be started on intravenous flucloxacillin and fusidic acid. Fusidic acid is preferred to benzylpenicillin partly because of the high prevalence of penicillin-resistant staphylococci and because it is particularly well concentrated in bone. However, for a known streptococcal infection benzylpenicillin

is better. Patients who are allergic to penicillin should be treated with a second- or third generation cephalosporin.
• Elderly and previously unfit patients In this group there is a greater than usual risk of Gram-negative infections, due to respiratory, gastro-intestinal, or urinary disorders and the likelihood of the patient needing invasive procedures. The antibiotic of choice would be a combination of flucloxacillin and a second- or third-generation cephalosporin.
• Patients with sickle-cell disease These patients are prone to osteomyelitis, which may be caused by a staphylococcal infection but in many cases is due to salmonella and/or other Gram-negative organisms. Chloramphenicol, which is effective against Grampositive, Gram-negative and anaerobic organisms, used to be the preferred antibiotic, though there were always worries about the rare complication of aplastic anaemia. Nowadays the antibiotic of choice is a third-generation cephalosporin or a fluoroquinolone like ciprofloxacin.
• Heroin addicts and immunocompromised patients Unusual infections (e.g. with Pseudomonas aeruginosa, Proteus mirabilis or anaerobic Bacteroides species) are likely in these patients. Infants with human immunodeficiency virus (HIV) infection may also have picked up other sexually transmitted organisms during birth. All patients with this type of background are therefore best treated empirically with a broad-spectrum antibiotic such as one of the third-generation cephalosporins or a fluoroquinolone preparation, depending on the results of sensitivity tests.
• Patients considered to be at risk of meticillin-resistant Staphylococcus aureus (MRSA) infection Patients admitted with acute haematogenous osteomyelitis and who have a previous history of MRSA infection, or any patient with a bone infection admitted to a hospital or a ward where MRSA is endemic, should be treated with intravenous vancomycin (or similar antibiotic) together with a third-generation cephalosporin.
The usual programme is to administer the drugs intravenously (if necessary adjusting the choice of antibiotic once the results of antimicrobial sensitivity become available) until the patient's condition begins to improve and the CRP values return to normal levels – which usually takes 2–4 weeks depending on the virulence of the infection and the patient's general degree of fitness. By that time the most appropriate antibiotic would have been prescribed, on the basis of sensitivity tests; this can then be administered orally for another 3–6 weeks, though if bone destruction is marked the period of treatment may have to be longer. While patients are on oral antibiotics it is important to track the serum antibiotic levels in order to ensure that the minimal inhibitory concentration (MIC) is maintained or exceeded. CRP, ESR and WBC values are also checked at regular intervals and treatment can be discontinued when these are seen to remain normal.

Complications
A lethal outcome from septicaemia is nowadays extremely rare; with antibiotics the child nearly always recovers and the bone may return to normal. But morbidity is common, especially if treatment is delayed or the organism is insensitive to the chosen antibiotic.
Epiphyseal damage and altered bone growth In neonates and infants whose epiphyses are still entirely cartilaginous, metaphyseal vessels penetrate the physis and may carry the infection into the epiphysis. If this happens, the physeal growth plate can be irrevocably damaged and the cartilaginous epiphysis may be destroyed, leading to arrest of growth and shortening of the bone. At the hip joint, the proximal end of the femur may be so badly damaged as to result in a pseudarthrosis.
Suppurative arthritis This may occur: (1) in very young infants, in whom the growth disc is not an impenetrable barrier; (2) where the metaphysis is intracapsular, as in the upper femur; or (3) from metastatic infection. In infants it is so common as almost to be taken for granted,

especially with osteomyelitis of the femoral neck. Ultrasound will help to demonstrate an effusion, but the definitive diagnosis is given by joint aspiration.
Metastatic infection This is sometimes seen – generally in infants – and may involve other bones, joints, serous cavities, the brain or lung. In some cases the infection may be multifocal from the outset. It is easy to miss secondary sites of infection when attention is focussed on one particular area; it is important to be alert to this complication and to examine the child all over and repeatedly. Pathological fracture Fracture is uncommon, but it may occur if treatment is delayed and the bone is weakened either by erosion at the site of infection or by overzealous debridement.
Chronic osteomyelitis Despite improved methods of diagnosis and treatment, acute osteomyelitis sometimes fails to resolve. Weeks or months after the onset of acute infection a sequestrum appears in the follow-up x-ray and the patient is left with a chronic infection and a draining sinus. This may be due to late or inadequate treatment but is also seen in debilitated patients and in those with compromised defence mechanisms.

POST-TRAUMATIC OSTEOMYELITIS
Open fractures are always contaminated and are therefore prone to infection. The combination of tissue injury, vascular damage, oedema, haematoma, dead bone fragments and an open pathway to the atmosphere must invite bacterial invasion even if the wound is not contaminated with particulate dirt. This is the most common cause of osteomyelitis in adults. Staphylococcus aureus is the usual pathogen, but other organisms such as E. coli, Proteus mirabilis and Pseudomonas aeruginosa are sometimes involved. Occasionally, anaerobic organisms (clostridia, anaerobicstreptococci or Bacteroides) appear in contaminated wounds.

CHRONIC OSTEOMYELITIS
This used to be the dreaded sequel to acute haematogenous osteomyelitis; nowadays it more frequently follows an open fracture or operation. The usual organisms (and with time there is always a mixed infection) are Staphylococcus aureus, Escherichia coli, Streptococcus pyogenes, Proteus mirabilis and Pseudomonas aeruginosa; in the presence of foreign implants Staphylococcus epidermidis, which is normally non-pathogenic, is the commonest of all.

ACUTE SUPPURATIVE ARTHRITIS
A joint can become infected by:
(1) direct invasion through a penetrating wound, intra-articular injection or arthroscopy;
(2) direct spread from an adjacent bone abscess; or
(3) blood spread from a distant site.
In infants it is often difficult to tell whether the infection started in the metaphyseal bone and spread to the joint or vice versa. In practice it hardly matters and in advanced cases it should be assumed that the entire joint and the adjacent bone ends are involved.
The causal organism is usually Staphylococcus aureus; however, in children between 1 and 4 years old, Haemophilus influenzae is an important pathogen unless they have been vaccinated against this organism. Occasionally other microbes, such as Streptococcus, Escherichia coli and Proteus, are encountered.
Predisposing conditions are rheumatoid arthritis, chronic debilitating disorders, intravenous drug abuse, immunosuppressive drug therapy and acquired immune deficiency syndrome (AIDS).

Pathology
The usual trigger is a haematogenous infection which settles in the synovial membrane; there is an acute inflammatory reaction with a serous or seropurulent exudate and an increase in synovial fluid. As pus appears in the joint, articular cartilage is eroded and destroyed, partly by bacterial enzymes and partly by proteolytic enzymes released from synovial cells, inflammatory cells and pus. In infants the entire epiphysis, which is still largely cartilaginous, may be severely damaged; in older children, vascular occlusion may lead to necrosis of the epiphyseal bone. In adults the effects are usually confined to the articular cartilage, but in late cases there may be extensive erosion due to synovial proliferation and ingrowth. If the infection goes untreated, it will spread to the underlying bone or burst out of the joint to form abscesses and sinuses. With healing there may be: (1) complete resolution and a return to normal; (2) partial loss of articular cartilage and fibrosis of the joint; (3) loss of articular cartilage and bony ankylosis; or (4) bone destruction and permanent deformity of the joint.

Clinical features
The clinical features differ somewhat according to the age of the patient.
In new-born infants the emphasis is on septicaemia rather than joint pain. The baby is irritable and refuses to feed; there is a rapid pulse and sometimes a fever.
Infection is often suspected, but it could be anywhere!
The joints should be carefully felt and moved to elicit the local signs of warmth, tenderness and resistance to movement. The umbilical cord should be examined for a source of infection. An inflamed intravenous infusion site should always excite suspicion. The baby's chest, spine and abdomen should be carefully examined to exclude other sites of infection. Special care should be taken not to miss a concomitant osteomyelitis in an adjacent bone end.
In children the usual features are acute pain in a single large joint (commonly the hip or the knee) and reluctance to move the limb ('pseudoparesis'). The child is ill, with a rapid pulse and a swinging fever. The overlying skin looks red and in a superficial joint swelling may be obvious. There is local warmth and marked tenderness. All movements are restricted, and often completely abolished, by pain and spasm. It is essential to look for a source of infection – a septic toe, a boil or a discharge from the ear.
In adults it is often a superficial joint (knee, wrist, a finger, ankle or toe) that is painful, swollen and inflamed. There is warmth and marked local tenderness, and movements are restricted. The patient should be questioned and examined for evidence of gonococcal infection or drug abuse. Patients with rheumatoid arthritis, and especially those on corticosteroid treatment, may develop a 'silent' joint infection. Suspicion may be aroused by an unexplained deterioration in the patient's general condition; every joint should be carefully examined.

Imaging
Ultrasonography is the most reliable method for revealing a joint effusion in early cases. Both hips should be examined for comparison. Widening of the space between capsule and bone of more than 2 mm is indicative of an effusion, which may be echo-free (perhaps a transient synovitis) or positively echogenic (more likely septic arthritis).
X-ray examination is usually normal early on but signs to be watched for are soft-tissue swelling, loss of tissue planes, widening of the radiographic 'joint space' and slight subluxation (because of fluid in the joint). With E. coli infections there is sometimes gas in the joint. Narrowing and irregularity of the joint space are late features.
MRI and radionuclide imaging are helpful in diagnosing arthritis in obscure sites such as the sacroiliac and sternoclavicular joints.

Investigations
The white cell count and ESR are raised and blood culture may be positive. However, special investigations take time and it is much quicker (and usually more reliable) to aspirate the joint and examine the fluid. It may be frankly purulent but beware! – in early cases the fluid may look clear. A white cell count and Gram stain should be carried out immediately: the normal synovial fluid leucocyte count is under 300 per mL; it may be over 10 000 per mL in non-infective inflammatory disorders, but counts of over 50 000 per mL are highly suggestive of sepsis. Gram-positive cocci are probably S. aureus; Gram-negative cocci are either H. Influenzae or Kingella kingae (in children) or Gonococcus (in adults). Samples of fluid are also sent for full micro - biological examination and tests for antibiotic sensitivity.
Differential diagnosis
Acute osteomyelitis In young children, osteomyelitis may be indistinguishable from septic arthritis; often one must assume that both are present. Other types of infection Psoas abscess and local infection of the pelvis must be kept in mind. Systemic features will obviously be the same as those of septic arthritis.
Trauma Traumatic synovitis or haemarthrosis may be associated with acute pain and swelling. A history of injury does not exclude infection. Diagnosis may remain in doubt until the joint is aspirated. Irritable joint At the onset the joint is painful and lacks some movement, but the child is not really ill and there are no signs of infection. Ultrasonography may help to distinguish septic arthritis from transient synovitis.

Haemophilic bleed An acute haemarthrosis closely resembles septic arthritis. The history is usually conclusive, but aspiration will resolve any doubt.
Rheumatic fever Typically the pain flits from joint to joint, but at the onset one joint may be misleadingly inflamed. However, there are no signs of septicaemia.
Juvenile rheumatoid arthritis This may start with pain and swelling of a single joint, but the onset is usually more gradual and systemic symptoms less severe than in septic arthritis.
Sickle-cell disease The clinical picture may closely resemble that of septic arthritis – and indeed the bone nearby may actually be infected! – so this condition should always be excluded in communities where the disease is common.
Gaucher's disease In this rare condition acute joint pain and fever can occur without any organism being found ('pseudo-osteitis'). Because of the predisposition to true infection, antibiotics should be given.
Gout and pseudogout In adults, acute crystal-induced synovitis may closely resemble infection. On aspirationthe joint fluid is often turbid, with a high whitecell count; however, microscopic examination by polarized light will show the characteristic crystals.
Treatment
The first priority is to aspirate the joint and examine the fluid. Treatment is then started without further delay and follows the same lines as for acute osteomyelitis. Once the blood and tissue samples have been obtained, there is no need to wait for detailed results before giving antibiotics. If the aspirate looks purulent, the joint should be drained without waiting for laboratory results (see below).
GENERAL SUPPORTIVE CARE
Analgesics are given for pain and intravenous fluids for dehydration.
SPLINTAGE
The joint should be rested, and for neonates and infants this may mean light splintage; with hip infection,the joint should be held abducted and 30 degrees flexed, on traction to prevent dislocation.

ANTIBIOTICS

Antibiotic treatment follows the same guidelines as presented for acute haematogenous osteomyelitis (see page 35). The initial choice of antibiotics is based on judgement of the most likely pathogens.

Neonates and infants up to the age of 6 months should be protected against staphylococcus and Gram-negative streptococci with one of the penicillinase- resistant penicillins (e.g. flucloxacillin) plus a third-generation cephalosporin.

Children from 6 months to puberty can be treated similarly. Unless they had been immunized there is a risk of Haemophilus infection.

Older teenagers and adults can be started on flucloxacillin and fusidic acid. If the initial examination shows Gram-negative organisms a third-generation cephalosporin is added. More appropriate drugs can be substituted after full microbiological investigation. Antibiotics should be given intravenously for 4–7 days and then orally for another 3 weeks.

DRAINAGE

Under anaesthesia the joint is opened through a small incision, drained and washed out with physiological saline. A small catheter is left in place and the wound is closed; suction–irrigation is continued for another 2 or 3 days. This is the safest policy and is certainly advisable (1) in very young infants, (2) when the hip is involved and (3) if the aspirated pus is very thick. For the knee, arthroscopic debridement and copious irrigation may be equally effective.

Older children with early septic arthritis (symptoms for less than 3 days) involving any joint except the hip can often be treated successfully by repeated closed aspiration of the joint; however, if there is no improvement within 48 hours, open drainage will be necessary.

AFTERCARE

Once the patient's general condition is satisfactory and the joint is no longer painful or warm, further damage is unlikely. If articular cartilage has been preserved, gentle and gradually increasing active movements are encouraged. If articular cartilage has been destroyed the aim is to keep the joint immobile while ankylosis is awaited. Splintage in the optimum position is therefore continuously maintained, usually by plaster, until ankylosis is sound.

Complications

Infants under 6 months of age have the highest incidence of complications, most of which affect the hip.

The most obvious risk factors are a delay in diagnosis and treatment (more than 4 days) and concomitant osteomyelitis of the proximal femur. Subluxation and dislocation of the hip, or instability of the knee should be prevented by appropriate posturing or splintage.

Damage to the cartilaginous physis or the epiphysis in the growing child is the most serious complication. Sequelae include retarded growth, partial or complete destruction of the epiphysis, deformity of the joint, epiphyseal osteonecrosis, acetabular dysplasia and pseudarthrosis of the hip. Articular cartilage erosion (chondrolysis) is seen in older patients and this may result in restricted movement or complete ankylosis of the joint.

TUBERCULOSIS

Once common throughout the world, tuberculosis showed a steady decline in its prevalence in developed countries during the latter half of the twentieth century, due mainly to the effectiveness of public health programmes, a general improvement in nutritional status and advances in chemotherapy. In the last two decades, however, the annual incidence (particularly of extrapulmonary tuberculosis) has risen again, a phenomenon which has been

attributed variously to a general increase in the proportion of elderly people, changes in population movements, the spread of intravenous drug abuse and the emergence of AIDS. The skeletal manifestations of the disease are seen chiefly in the spine and the large joints, but the infection may appear in any bone or any synovial or bursal sheath. Predisposing conditions include chronic debilitating disorders, diabetes, drug abuse, prolonged corticosteroid medication, AIDS and other disorders resulting in reduced defence mechanisms.

3.Inflammatory rheumatic disorders

The term ‘inflammatory rheumatic disorders’ covers a number of diseases that cause chronic pain, stiffness and swelling around joints and tendons. In addition, they are commonly associated with extra-articular features including skin rashes and inflammatory eye disease. Individuals with these diseases tend to die younger than their peers as a result of the effects of chronic inflammation. Many – perhaps all – are due to a faulty immune reaction resulting from a combination of environmental exposures against a background of genetic predisposition.

RHEUMATOID ARTHRITIS

Rheumatoid arthritis (RA) is the most common cause of chronic inflammatory joint disease. The most typical features are a symmetrical polyarthritis and tenosynovitis, morning stiffness, elevation of the erythrocyte sedimentation rate (ESR) and the appearance of autoantibodies that target immunoglobulins (rheumatoid factors) in the serum. Rheumatoid arthritis is a systemic disease and changes can be widespread in a number of tissues of the body. Individuals with RA tend to die younger than their peers as a result of the effects of chronic inflammation on a number of organ systems. Chief among these is early ischaemic heart disease secondary to the effects of inflammation on the cardiovascular system. The reported prevalence of RA in most populations is 1–3 per cent, with a peak incidence in the fourth or fifth decades. Women are affected 3 or 4 times more commonly than men. Both the prevalence and the clinical expression vary between populations; the disease is more common (and generally more severe) in Caucasians living in the urban communities of Europe and North America than in the rural populations of Africa.

Cause

The cause of RA is still incompletely worked out. However, a great deal is now known about the circumstances in which RA develops, and hypotheses about its aetiology and pathogenesis have been suggested.

Important factors in the evolution of RA are:
(1) genetic susceptibility;
(2) an immunological reaction,
possibly involving a foreign antigen, preferentially
focussed on synovial tissue;
(3) an inflammatory reaction in joints and tendon sheaths;
(4) the appearance of rheumatoid factors (RF) in the blood and synovium;
(5) perpetuation of the inflammatory process;
(6) articular cartilage destruction.

Genetic susceptibility A genetic association is suggested by the fact that RA is more common in first degree relatives of patients than in the population at large; furthermore twin studies have revealed a concordance rate of around 30 per cent if one of the pair is affected. The human leucocyte antigen (HLA) DR4 occurs in about 70 per cent of people with RA, compared to a frequency of less than 30 per cent in normal controls. HLA-DR4 is encoded in the major histocompatibility complex (MHC) region on chromosome 6. There are strong associations between HLA-DR4 and RA. In particular a key structural conformation within

the HLA-DR4 binding groove called the ‘shared epitope’ seems important. This may suggest that a particular antigen that fits into this may be playing a part. HLA Class II molecules appear as surface antigens on cells of the immune system (B lymphocytes, macrophages, dendritic cells), which can act as antigen- presenting cells (APCs). In some T-cell immune reactions, the process is initiated only when the antigenic peptide is presented in association with a specific HLA allele. It has been suggested that this is the case in people who develop RA; the idea is even more attractive if one proposes that the putative antigen has a special affinity for synovial tissue. So far no such antigen has been discovered.

The inflammatory reaction Once the APC/T-cell interaction is initiated, various local factors come into play and lead to a progressive enhancement of the immune response. There is a marked proliferation of cells in the synovium, with the appearance of new blood vessel formation. Immune cells coordinate their action by the use of ‘short-range hormones’ (cyto - kines), which can activate inflammatory cells such as macrophages and B cells. Some cytokines called chemokines attract other inflammatory cells to the area.

Over recent years it has become clear that certain cytokines are important in RA. These include tumour necrosis factor (TNF), interleukin-1 (IL-1) and interleukin- 6 (IL-6). The resulting synovitis, both in joints and in tendon sheath linings, is the hallmark of early RA. Rheumatoid factor B-cell activation in RA leads tothe production of anti-IgG autoantibodies, which are detected in the blood as ‘rheumatoid factor’ (RF). Low levels of RF can be found in many ‘normal’ individuals but when the levels are high an inflammatory disease is likely. Other autoimmune conditions such as systemic lupus erythematosus (SLE) and Sjögren’s syndrome are also associated with the presence of RF. In recent years other autoantibodies associated with RA have been identified. The most important are anticyclic citrullinated peptide antibodies (anti-CCP). The presence of anti-CCP is very specific for RA. Patients with a positive RF test tend to be more severely affected than those with a negative test.

Chronic synovitis and joint destruction Chronic rheumatoid synovitis is associated with the production of proteolytic enzymes, prostaglandins and the cytokines TNF and IL-1. Immune complexes are deposited in the synovium and on the articular cartilage, where they appear to augment the inflammatory process. This combination of factors leads to depletion of the cartilage matrix and, eventually, damage to cartilage and underlying bone. Vascular proliferation and osteoclastic activity, most marked at the edges of the articular surface, may contribute further to cartilage destruction and peri-articular bone erosion.

Pathology

Rheumatoid arthritis is a systemic disease but the most characteristic lesions are seen in the synovium or within rheumatoid nodules. The synovium is engorged with new blood vessels and packed full of inflammatory cells.

JOINTS AND TENDONS

The pathological changes, if unchecked, proceed in four stages. Previously it was felt that having gone through these stages the disease activity could be ‘burnt out’. This does not appear to be the case. In any one joint features of different stages can be occurring simultaneously and even when joints are very badly destroyed the ongoing inflammation can continue to seriously damage systemic health by accelerating other disease processes such as ischaemic heart disease.

Stage 1 – pre-clinical Well before RA becomes clinically apparent the immune pathology is already beginning. Raised ESR, C-reactive protein (CRP) and RF may be detectable years before the first diagnosis.

Stage 2 – synovitis Early changes are vascular congestion with new blood vessel formation, proliferation of synoviocytes and infiltration of the subsynovial layers by polymorphs, lymphocytes and plasma cells. There is thickening of the capsular structures, villous

formation of the synovium and a cell-rich effusion into the joints and tendon sheaths. Although painful, swollen and tender, these structures are still intact and mobile, and the disorder is potentially reversible.
Stage 3 – destruction Persistent inflammation causes joint and tendon destruction. Articular cartilage is eroded, partly by proteolytic enzymes, partly by vascular tissue in the folds of the synovial reflections, and partly due to direct invasion of the cartilage by a pannus of granulation tissue creeping over the articular surface. At the margins of the joint, bone is eroded by granulation tissue invasion and osteoclastic resorption.
Similar changes occur in tendon sheaths, causing tenosynovitis, invasion of the collagen bundles and, eventually, partial or complete rupture of tendons. A synovial effusion, often containing copious amounts of fibrinoid material, produces swelling of the joints, tendons and bursae.
Stage 4 – deformity The combination of articular destruction, capsular stretching and tendon rupture leads to progressive instability and deformity of the joints. The inflammatory process usually continues but the mechanical and functional effects of joint and tendon disruption now become vital.

EXTRA-ARTICULAR TISSUES
Rheumatoid nodules The rheumatoid nodule is a small granulomatous lesion consisting of a central necrotic zone surrounded by a radially disposed palisade of local histiocytes, and beyond that by inflammatory granulation tissue. Nodules occur under the skin (especially over bony prominences), in the synovium, on tendons, in the sclera and in many of the viscera.
Lymphadenopathy Not only the nodes draining inflamed joints, but also those at a distance such as the mediastinal nodes, can be affected. This, as well as a mild splenomegaly, is due to hyperactivity of the reti - culoendothelial system. More severe splenomegaly can also be associated with neutropaenia as part of Felty's syndrome. Vasculitis This can be a serious and life-threatening complication of RA. Involvement of the skin, including nailfold infarcts, is common but organ infarction can occur.
Muscle weakness Muscle weakness is common. It may be due to a generalized myopathy or neuropathy, but it is important to exclude spinal cord disease or cord compression due to vertebral displacement (atlantoaxial subluxation). Sensory changes may be part of a neuropathy, but localized sensory and motor symptoms can also result from nerve compression by thickened synovium (e.g. carpal tunnel syndrome).
Visceral disease The lungs, heart, kidneys, gastrointestinal tract and brain are sometimes affected. Ischaemic heart disease and osteoporosis are common complications.

Clinical features

The onset of RA is usually insidious, with symptoms emerging over a period of months. Occasionally the disease starts quite suddenly. In the early stages the picture is mainly that of a polysynovitis, with soft-tissue swelling and stiffness. Typically, a woman of 30–40 years complains of pain, swelling and loss of mobility in the proximal joints of the fingers. There may be a previous history of 'muscle pain', tiredness, loss of weight and a general lack of well-being. As time passes, the symptoms 'spread' to other joints – the wrists, feet, knees and shoulders in order of frequency. Another classic feature is generalized stiffness after periods of inactivity, and especially after rising from bed in the early morning. This early morning

stiffness typically lasts longer than 30 minutes. Physical signs may be minimal, but usually there is symmetrically distributed swelling and tenderness of the metacarpophalangeal joints, the proximal interphalangeal joints and the wrists. Tenosynovitis is common in the extensor compartments of the wrist and the flexor sheaths of the fingers; it is diagnosed by feeling thickening, tenderness and crepitation over the back of the wrist or the palm while passively moving the fingers. If the larger joints are involved, local warmth, synovial hypertrophy and intra-articular effusion may be more obvious. Movements are often limited but the joints are still stable and deformity is unusual. In the later stages joint deformity becomes increasingly apparent and the acute pain of synovitis is replaced by the more constant ache of progressive joint destruction. The combination of joint instability and tendon rupture produces the typical 'rheumatoid' deformities: ulnar deviation of the fingers, radial and volar displacement of the wrists, valgus knees, valgus feet and clawed toes. Joint movements are restricted and often very painful. About a third of all patients develop pain and stiffness in the cervical spine. Function is increasingly disturbed and patients may need help with grooming, dressing and eating.

Extra-articular features These often appear in patients with severe disease. The most characteristic is the appearance of nodules. They are usually found as small subcutaneous lumps, rubbery in consistency, at the back of the elbows, but they also develop in tendons (where they may cause 'triggering' or rupture), in the viscera and the eye. They are pathognomonic of RA, but occur in only 25% of patients.

Less specific features include muscle wasting, lymphadenopathy, scleritis, nerve entrapment syndromes, skin atrophy or ulceration, vasculitis and peripheral sensory neuropathy. Marked visceral disease, such as pulmonary fibrosis, is rare.

Imaging

X-rays Early on, x-rays show only the features of synovitis: soft-tissue swelling and peri-articular osteoporosis. The later stages are marked by the appearance of marginal bony erosions and narrowing of the articular space, especially in the proximal joints of the hands and feet. However, most individuals have evidence of erosions within 2 years. In advanced disease, articular destruction and joint deformity are obvious. Flexion and extension views of the cervical spine often show subluxation at the atlanto-axial or mid-cervical levels; surprisingly, this causes few symptoms in the majority of cases.

Ultrasound scanning and MRI The use of other imaging techniques to look at soft-tissue changes and early erosions within joints has become more common. Ultrasound can be particularly useful in defining the presence of synovitis and early erosions. Additional information on vascularity can be obtained if Doppler techniques are used.

Blood investigations

Normocytic, hypochromic anaemia is common and is a reflection of abnormal erythropoiesis due to disease activity. It may be aggravated by chronic gastrointestinal blood loss caused by non-steroidal anti-inflammatory drugs. In active phases the ESR and CRP concentration are usually raised. Serological tests for rheumatoid factor are positive in about 80 per cent of patients and antinuclear factors are present in 30 per cent. Neither of these tests is specific and neither is required for a diagnosis of rheumatoid arthritis. Newer tests such as those for anti-CCP antibodies have added much greater specificity but at the expense of sensitivity.

Synovial biopsy

Synovial tissue may be obtained by needle biopsy, via the arthroscope, or by open operation. Unfortunately, most of the histological features of rheumatoid arthritis are non-specific.

Diagnosis

The usual criteria for diagnosing rheumatoid arthritis are the presence of a bilateral, symmetrical polyarthritis involving the proximal joints of the hands or feet, and persisting for at least 6 weeks. If there are subcutaneous nodules or x-ray signs of peri-articular erosions, the diagnosis is certain. A positive test for rheumatoid factor in the absence of the above features is not sufficient evidence of rheumatoid arthritis, nor does a negative test exclude the diagnosis if the other features are all present. The chief value of the rheumatoid factor tests is in the assessment of prognosis: persistently high titres herald more serious disease including extra-articular features.

Atypical forms of presentation are not uncommon. The early stages may be punctuated by spells of quiescence, during which the diagnosis is doubted, but sooner or later the more characteristic features appear. Occasionally, in older people, the onset is explosive, with the rapid appearance of severe joint pain and stiffness; paradoxically these patients have a relatively good prognosis. Now and then (more so in young women) the disease starts with chronic pain and swelling of a single large joint and it may take months or years before other joints are involved. The presence of tenderness on squeezing across all metacarpophalangeal or metatarsophalangeal joints, early morning stiffness of at least 30 minutes and a raised ESR are highly suggestive of a diagnosis of rheumatoid arthritis. A rapid diagnosis is vital so that early treatment can be started with disease-modifying antirheumatic drugs. In the differential diagnosis of polyarthritis several disorders must be considered.

Seronegative inflammatory polyarthritis Polyarthritis is a feature of a number of conditions including psoriatic arthritis, adult Still's disease, systemic lupus erythematosus and other connective-tissue diseases. These are considered in later sections.

Ankylosing spondylitis This is primarily an inflammatory disease of the sacroiliac and intervertebral joints, causing back pain and progressive stiffness; however, it may also involve the peripheral joints.

Reiter's disease The larger joints and the lumbosacral spine are the main targets. There is usually a history of urethritis or colitis and often also conjunctivitis.

Polyarticular gout Tophaceous gout affecting multiple joints can, at first sight, be mistaken for rheumatoid arthritis. On x-ray the erosions are quite different from those of rheumatoid arthritis; the diagnosis is clinched by identifying typical birefringent urate crystals in the joint fluid or a nodular tophus. It is a curious fact that, although both gout and RA are fairly common, the two conditions are rarely seen in the same patient. The reason for this is unknown.

Calcium pyrophosphate deposition disease This condition is usually seen in older people. Typically it affects large joints, but it may occur in the wrist and metacarpophalangeal joints as well. X-ray signs are fairly characteristic and crystals may be identified in synovial fluid or synovium.

Sarcoidosis Sarcoid disease sometimes presents with a symmetrical small-joint polyarthritis and no bone involvement; in other cases a large joint such as the knee or ankle may be involved. Erythema nodosum and hilar lymphadenopathy on chest x-ray are clues to the diagnosis. Acute sarcoidosis usually subsides spontaneously within 6 months. Chronic sarcoidosis produces granulomatous infiltration of lungs, bone, synovium and other organs and is more common in Afro-Caribbean than Caucasian peoples. In addition to polyarthritis and tenosynovitis, there are usually x-ray features of punched-out 'cysts' and cortical erosions in the bones of the hands and feet. The ESR and serum angiotensin converting enzyme (SACE) may be raised. Biopsy of affected tissue shows typical noncaseating granulomas.

Treatment with non-steroidal anti-inflammatory drugs (NSAIDs) may be adequate but in more intractable cases corticosteroids or other immunosuppressive preparations are necessary.
Lyme disease This tick-borne spirochaetal infection usually starts with a skin lesion and flu-like symptoms and then spreads to multiple organs. If the initial lesions are missed or left untreated, patients may present with an asymmetrical inflammatory polyarthritis affecting mainly the larger joints. It is most likely to be encountered in known endemic areas in North America, Europe and Asia. In late cases serological tests may be positive. Treatment with doxycycline or one of the newer cephalosporins is usually effective for the arthritic features.
Viral arthritis Viral infections are often associated with a transient polyarthralgia; flu-like illness and a rash will suggest the diagnosis. However, some infections – most typically parvovirus B19 – occasionally cause a symmetrical polysynovitis (including the finger joints) and early morning stiffness, symptoms which may last for several months or may recur over a few years. The absence of 'rheumatoid' x-ray features and subcutaneous nodules will raise suspicions about the diagnosis.
Polymyalgia rheumatica This condition, which is seen mainly in the middle-aged or elderly, is characterized by aching discomfort around the pectoral and pelvic girdles, post-inactivity stiffness and muscular weakness. The joints are not tender but the muscles may be. The ESR and CRP are almost always elevated. Corticosteroids (as little as 10 mg a day) provide rapid and dramatic relief of all symptoms, and this response is often used as a diagnostic test. The condition may be associated with, and certainly carries the risk of, temporal arteritis resulting in blindness.
Osteoarthritis Polyarticular osteoarthritis (OA), which typically involves the finger joints, is often mistaken for RA. A moment's reflection will usually dispel any doubt: OA always involves the distal interphalangeal joints and causes a nodular arthritis with radiologically obvious osteophytes, whereas RA affects the proximal joints of the hand and causes predominantly erosive features.
Some confusion may arise from the fact that RA, in its later stages, is associated with loss of articular cartilage and secondary OA. Enquiry into the early history will usually untangle the diagnosis. Sometimes, however, RA atypically affects only a few of the larger joints and it is then very difficult to distinguish from OA; x-ray features such as loss of articular cartilage throughout the entire joint and lack of hypertrophic bone changes (sclerosis and osteophytes) should suggest an inflammatory arthritis.

Treatment

There is no cure for rheumatoid arthritis. However, advances in therapy have revolutionized the treatment approach with associated major improvements in outcome (Kennedy et al., 2005). Medical treatment is guided by the principle that inflammation should be reduced rapidly and aggressively. A multidisciplinary approach is needed from the beginning: ideally the therapeutic team should include a rheumatologist, orthopaedic surgeon, physiotherapist, occupational therapist, orthotist and social worker. Their deployment and priorities will vary according to the individual and stage of the disease. At the onset of the disease both the patient and the doctor will be uncertain about the likely rate of progress. An attempt should be made to determine the likely prognosis. Poor prognosis is associated with female sex, multiple joint involvement, high ESR and CRP, positive RF and anti-CCP, younger age and the presence of erosions at diagnosis.

PRINCIPLES OF MEDICAL MANAGEMENT
Treatment should be aimed at controlling inflammation as rapidly as possible. This is likely to require the use of corticosteroids for their rapid onset (initially oral doses of 30 mg of

prednisolone or 120 mg i.m. methylprednisolone may be used). Steroids should be rapidly tapered to prevent significant side effects. In addition, disease-modifying antirheumatic drugs (DMARDs) should be started at this time. The first choice is now methotrexate at doses of 10–25 mg/week. This may be used initially alone or in combination with sulfasalazine and hydroxychloroquine. Leflunomide can also be considered if methotrexate is not tolerated. Gold and penicillamine are now used rarely.
Control of pain and stiffness with non-steroidal anti-inflammatory drugs (NSAIDs) may be needed, maintaining muscle tone and joint mobility by a balanced programme of exercise, and general advice on coping with the activities of daily living.
If there is no satisfactory response to DMARDs, it is wise to progress rapidly to biological therapies such as the TNF inhibitors infliximab, etanercept and adalimumab (Scott & Kingsley, 2006; Deighton et al.,2006). Additional measures include the injection of longacting corticosteroid preparations into inflamed joints and tendon sheaths. It is sometimes feared that such injections may themselves cause damage to articular cartilage or tendons. However, there is little evidence that they are harmful, provided they are used sparingly and with full precautions against infection. Prolonged rest and immobility is likely to weaken muscles and lead to a worse prognosis. However, some splinting can be helpful at any stage of the disease.

KEY ELEMENTS IN MEDICAL TREATMENT

-Identify patients with RA as early as possible
-Start disease-modifying antirheumatic drugs (DMARDs) immediately
-Consider combination therapy with multiple DMARDs
-If DMARDs fail, progress rapidly to biological therapies such as the TNF inhibitors infliximab, etanercept and adalimumab

PHYSIOTHERAPY AND OCCUPATIONAL THERAPY
Preventative splinting and orthotic devices may delay the march of events; however, it is important to encourage activity. If these fail to restore and maintain function, operative treatment is indicated.

SURGICAL MANAGEMENT
At first this consists mainly of soft-tissue procedures (synovectomy, tendon repair or replacement and joint stabilization); in some cases osteotomy may be more appropriate.
In late rheumatoid disease, severe joint destruction, fixed deformity and loss of function are clear indications for reconstructive surgery. Arthrodesis, osteotomy and arthroplasty all have their place and are considered in the appropriate chapters. However, it should be recognized that patients who are no longer suffering the pain of active synovitis and who are contented with a limited pattern of life may not want or need heroic surgery merely to improve their anatomy. Careful assessment for occupational therapy, the provision of mechanical aids and adjustments to their home environment may be much more useful. It appears safe to continue methotrexate during elective orthopaedic surgery. However, doses of corticosteroids should be as low as possible and biological therapies such as the TNF inhibitors should be stopped prior to surgery where possible.

Complications

Fixed deformities The perils of rheumatoid arthritis are often the commonplace ones resulting from ignorance and neglect. Early assessment and planning should prevent postural deformities, which will result in joint contractures.

Muscle weakness Even mild degrees of myopathy or neuropathy, when combined with prolonged inactivity, may lead to profound muscle wasting and weakness. This should be prevented by control of inflammation, physiotherapy and pain control, if possible; if not, the surgeon must be forewarned of the difficulty of postoperative rehabilitation.

Joint rupture Occasionally the joint lining ruptures and synovial contents spill into the soft tissues. Treatment is directed at the underlying synovitis, i.e. splintage and injection of the joint, with synovectomy as a second resort.

Infection Patients with rheumatoid arthritis – and even more so those on corticosteroid therapy – are susceptible to infection. Sudden clinical deterioration, or increased pain in a single joint, should alert one to the possibility of septic arthritis and the need for joint aspiration.

Spinal cord compression This is a rare complication of cervical spine (atlanto-axial) instability. The onset of weakness and upper motor neuron signs in the lower limbs is suspicious. If they occur, immobilization of the neck is essential and spinal fusion should be carried out as soon as possible.

Systemic vasculitis Vasculitis is a rare but potentially serious complication. Corticosteroids and immunosuppressives such as intravenous cyclophosphamide may be required.

Amyloidosis This is another rare but potentially lethal complication of longstanding rheumatoid arthritis. The patient presents with proteinuria and progressive renal failure. Finding amyloid in a rectal or renal biopsy makes the diagnosis. Aggressive control of inflammation has reduced this complication significantly.

Prognosis

Rheumatoid arthritis runs a variable course. When the patient is first seen it is difficult to predict the outcome, but high titres of rheumatoid factor, peri-articular articular erosions, rheumatoid nodules, severe muscle wasting, joint contractures and evidence of vasculitis are bad prognostic signs. Women, on the whole, fare somewhat worse than men. Without effective treatment about 10 per cent of patients improve steadily after the first attack of active synovitis; 60 per cent have intermittent phases of disease activity and remission, but with a slow downhill course over many years; 20 per cent have severe joint erosion, which is usually evident within the first 5 years; and 10 per cent end up completely disabled. In addition, a reduction in life expectancy by 5–10 years is common and is often due to premature ischaemic heart disease. However, early aggressive medical treatment appears to reduce the morbidity and mortality.

SERONEGATIVE SPONDYLO ARTHROPATHIES

ANKYLOSING SPONDYLITIS

Like rheumatoid arthritis, this is a generalized chronicinflammatory disease, but its effects are seen mainly in the spine and sacroiliac joints. It is characterized by pain and stiffness of the back, with variable involvement of the hips and shoulders and (more rarely) the peripheral joints. Its reported prevalence is 0.1 to 0.2 per cent in western Europe and North America, but ismuch lower in Japanese and African peoples. Males are affected more frequently than females (estimates vary from 2:1 to 10:1) and the usual age at onset is between 15 and 25

years. There is a strong tendency to familial aggregation and association with the genetic marker HLA-B27.

REITER'S SYNDROME AND REACTIVE ARTHRITIS

The syndrome described by Hans Reiter in 1916 (and 100 years before that by Benjamin Brodie) is a clinical triad of urethritis, arthritis and conjunctivitis occurring some weeks after either dysentery or genitourinary infection. It is now recognized that this is one of the classic forms of reactive arthritis, i.e. an aseptic inflammatory arthritis associated with non-specific infection (often urogenital or bowel). Its prevalence is difficult to assess, but it is probably the commonest type of large-joint polyarthritis in young men. It is thought to occur in 1–3 per cent of all people who develop either non-specific urogenital infection or Shigella dysentery, but its incidence may be as high as 25 per cent in those who are HLA-B27 positive. Men are affected more often than women (the ratio is about 10:1), but this may simply reflect the difficulty of diagnosing the genitourinary infection in women. The usual age at onset is between 20 and 40 years, but children are affected too – perhaps after an episode of diarrhoea

PSORIATIC ARTHRITIS

Polyarthritis and psoriasis are often seen together. Usually this is simply a chance concurrence of two fairly common disorders. In some cases, however, the patient has a true psoriatic arthritis – a distinct entity characterized by seronegative polysynovitis, erosive (sometimes very destructive) arthritis, and a significant incidence of sacroiliitis and spondylitis. The prevalence of psoriasis is 1–2 per cent, but only about 5 per cent of those affected will develop psoriatic arthritis. The usual age at onset is 30–50 years (often later than the skin lesions).

JUVENILE IDIOPATHIC ARTHRITIS

Juvenile idiopathic arthritis (JIA) is the preferred term for non-infective inflammatory joint disease of more than 3 months' duration in children under 16 years of age. It embraces a group of disorders in all of which pain, swelling and stiffness of the joints are common features. The prevalence is about 1 per 1000 children, and boys and girls are affected with equal frequency. The cause is similar to that of rheumatoid arthritis: an abnormal immune response to some antigen in children with a particular genetic predisposition. However, rheumatoid factor is usually absent. The pathology, too, may be like that of rheumatoid arthritis: primarily a synovial inflammation leading to fibrosis and ankylosis. Stiffening tends to occur in whatever position the joint is allowed to assume; thus flexion deformities are a common and characteristic feature. Chronic inflammation and alterations in the local blood supply may affect the epiphyseal growth plates, leading to both local bone deformities and an overall retardation of growth. However, cartilage erosion is less marked than in rheumatoid arthritis and severe joint instability is uncommon.

Clinical features

Children with JIA present in several characteristicways. About 15 per cent have a systemic illness, and arthritis only develops somewhat later; the majority (60–70 per cent) have a pauciarticular arthritis affecting a few of the larger joints; about 10 per cent present with polyarticular arthritis, sometimes closely resembling RA; the remaining 5–10 per cent develop a seronegative spondyloarthritis

SYSTEMIC JIA

This, the classic Still's disease, is usually seen below the age of 3 years and affects boys and girls equally. It starts with intermittent fever, rashes and malaise; during these episodes, which occur almost daily, the child appears to be quite ill but after a few hours the clinical condition improves again. Less constant features are lymphadenopathy, splenomegaly and hepato megaly. Joint swelling occurs some weeks or months after the onset; fortunately, it usually resolves when the systemic illness subsides but it may go on to progressive seronegative polyarthritis, leading to permanent deformity of the larger joints and fusion of the cervical apophyseal joints. By puberty there may be stunting of growth, often abetted by the earlier use of corticosteroids.

PAUCIARTICULAR JIA

This is by far the commonest form of JIA. It usually occurs below the age of 6 years and is much more common in girls; occasionally older children are affected. Only a few joints are involved and there is no systemic illness. The child presents with pain and swelling of medium-sized joints (knees, ankles, elbows and wrists); sometimes only one joint is affected. Rheumatoid factor tests are negative but antinuclear antibodies (ANA) may be positive. A serious complication is chronic iridocyclitis, which occurs in about 50 per cent of patients. The arthritis often goes into remission after a few years but by then the child is left with asymmetrical deformities and growth defects that may be permanent.

POLYARTICULAR JIA

Polyarticular arthritis, typically with involvement of the temporomandibular joints and the cervical spine, is usually seen in older children, mainly girls. The hands and wrists are often affected, but the classic deformities of rheumatoid arthritis are uncommon and rheumatoid factor is usually absent. In some cases, however, the condition is indistinguishable from adult rheumatoid arthritis, with a positive rheumatoid factor test; these probably warrant the designation 'juvenile rheumatoid arthritis'.

SERONEGATIVE SPONDYLOARTHROPATHY

In older children – usually boys – the condition may take the form of sacroiliitis and spondylitis; hips and knees are sometimes involved as well. Tests for HLAB27 are often positive and this should probably be regarded as 'juvenile ankylosing spondylitis'. X-rays In early disease non-specific changes such as softtissue swelling may be seen, but x-ray is mainly useful to exclude other painful disorders. Later there may be signs of progressive joint erosion and deformity

Investigations

The white cell count and ESR are markedly raised in systemic JIA, less so in the other forms. Rheumatoid factor tests are positive only in juvenile RA. Joint aspiration and synovial fluid examination may be essential to exclude infection or haemarthrosis.

Diagnosis

In the early stages, before chronic arthritis is fully established, diagnosis may be difficult. Systemic JIA may start with an illness resembling a viral infection. Pauciarticular JIA, especially if only one joint is involved, is indistinguishable from Reiter's disease or septic arthritis (if the signs are acute) or tuberculous synovitis (if they are more subdued). Other conditions that need to be excluded are rheumatic fever, one of the bleeding disorders and leukaemia. In most cases the problem is resolved once the full pattern of joint involvement is established, but blood investigations, joint aspiration and synovial biopsy may be required to clinch the diagnosis.

Treatment

General treatment Systemic treatment is similar to that of rheumatoid arthritis, including the use of second-line drugs such as hydroxychloroquine, sulfasalazine or low-dose methotrexate

for those with seropositive juvenile RA. Corticosteroids should be used only for severe systemic disease and for chronic iridocyclitis unresponsive to topical therapy. Severe inflammatory disease may need to be treated with cytokine inhibitors such as anti-TNF therapies. Children and parents alike need sympathetic counselling to help them cope with the difficulties of social adjustments, education and training.
Local treatment The priorities are to prevent stiffness and deformity. Night splints may be useful for the wrists, hands, knees and ankles; prone lying for some period of each day may prevent flexion contracture of the hips. Between periods of splinting, active exercises are encouraged; these are started by the physiotherapist but the parents must be taught how to continue the programme. Fixed deformities may need correction by serial plasters or by a spell in hospital on a continuous passive motion (CPM) machine; when progress is no longer being made, joint capsulotomy may help. For painful eroded joints, useful procedures include custom-designed arthroplasties of the hip and knee (even in children), and arthrodesis of the wrist or ankle.

Complications
Ankylosis While most patients recover good function, some loss of movement is common. Hips, knees and elbows may be unable to extend fully, and in the spondylitic form of JIA the spine, hips and knees may be almost rigid. Temporomandibular ankylosis and stiffness of the cervical spine can make general anaesthesia difficult and dangerous.
Growth defects There is a general retardation of growth, aggravated by prolonged corticosteroid therapy. In addition, epiphyseal disturbances lead to characteristic deformities: external torsion of the tibia, dysplasia of the distal ulna, underdevelopment of the mandible, shortness of the neck and scoliosis.
Fractures Children with chronic joint disease may suffer osteoporosis and they are prone to fractures.
Iridocyclitis This is most common in ANA-positive pauciarticular disease; untreated it may lead to blindness.
Amyloidosis In children with longstanding active disease there is a serious risk of amyloidosis, which may be fatal.

Prognosis
Fortunately, most children with JIA recover from the arthritis and are left with only moderate deformity and limitation of function. However, 5–10 per cent (and especially those with juvenile rheumatoid arthritis) are severely crippled and require treatment throughout life.
A significant number of children with JIA (about 3 per cent) still die – usually as a result of renal failure due to amyloidosis, or following overwhelming infection.

4.Crystal deposition disorders

The crystal deposition disorders are a group of conditions characterized by the presence of crystals in and around joints, bursae and tendons. Although many different crystals are found, three clinical conditions in particular are associated with this phenomenon:
• gout
• calcium pyrophosphate dihydrate (CPPD) deposition
disease
• calcium hydroxyapatite (HA) deposition disorders.
Characteristically, in each of the three conditions, crystal deposition has three distinct consequences:
(1)it may be totally inert and asymptomatic;
(2) it mayinduce an acute inflammatory reaction; or
(3) it may result in slow destruction of the affected tissues.

GOUT

Gout is a disorder of purine metabolism characterized by hyperuricaemia, deposition of monosodium urate monohydrate crystals in joints and peri-articular tissues and recurrent attacks of acute synovitis. Late changes include cartilage degeneration, renal dysfunction and uric acid urolithiasis. The clinical disorder was known to Hippocrates and its association with hyperuricaemia was recognized well over 100 years ago. The prevalence of symptomatic gout varies from 1 to over 10 per 1000, depending on the race, sex and age of the population studied: it is much commoner in Caucasian than in Negroid peoples; it is more widespread in men than in women (the ratio may be as high as 20:1); and it is rarely seen before the menopause in females. Although the risk of developing clinical features of gout increases with increasing levels of serum uric acid, only a fraction of those with hyperuricaemia develop symptoms. However, ‘hyperuricaemia’ and ‘gout’ are generally regarded as part and parcel of the same disorder.

Pathology

Hyperuricaemia Nucleic acid and purine metabolism normally proceeds, through complex pathways, to the production of hypoxanthine and xanthine; the final breakdown to uric acid is catalysed by the enzyme xanthine oxidase. Monosodium urate appears in ionic form in all the body fluids; about 70 per cent is derived from endogenous purine metabolism and 30 per cent from purine-rich foods in the diet. It is excreted (as uric acid) mainly by the kidneys and partly in the gut. Urate is poorly soluble, with a plasma saturation value of only 7 mg/dL (0.42 mmol/L). This concentration is commonly exceeded in normal individuals and epidemiological studies have identified entire populations (for example the Maoris of New Zealand) who have unusually high levels of serum uric acid. The term ‘hyperuricaemia’ is therefore generally reserved for individuals with a serum urate concentration which is significantly higher than that of the population to which they belong (more than two standard deviations above the mean); this is about +0.42 mmol/L for men and 0.35 mmol/L for women in western Caucasian peoples. By this definition, about 5 per cent of men and less

than 1 per cent of women have hyperuricaemia; the majority suffer no pathological consequences and they remain asymptomatic throughout life.
Gout Urate crystals are deposited in minute clumps in connective tissue, including articular cartilage; the commonest sites are the small joints of the hands and feet. For months, perhaps years, they remain inert. Then, possibly as a result of local trauma, the needlelike crystals are dispersed into the joint and the surrounding tissues where they excite an acute inflammatory reaction. Individual crystals may be phagocytosed by synovial cells and polymorphs or may float free in the synovial fluid.With the passage of time, urate deposits may build up in joints, peri-articular tissues, tendons and bursae; common sites are around the metatarsophalangeal joints of the big toes, the Achilles tendons, the olecranon bursae and the pinnae of the ears. These clumps of chalky material, or tophi (L. tophus = porous stone), vary in size from less than 1 mm to several centimetres in diameter. They may ulcerate through the skin or destroy cartilage and peri-articular bone.

Some factors predisposing to hyperuricaemia
-Older age, male gender
-Genetic enzyme defects, hyperparathyroidism
-Haemolytic disorders, myeloproliferative disorders
-Obesity, diabetes, hypertension
-High consumption of red meat, hyperlipidaemia
-Chronic inflammatory diseases
-Long-term use of aspirin or diuretics
-Alcohol abuse

Classification
Gout is often classified into ‘primary’ and ‘secondary’ forms. Primary gout (95 per cent) occurs in the absence of any obvious cause and may be due to constitutional under-excretion (the vast majority) or overproduction of urate. Secondary gout (5 per cent) results from prolonged hyperuricaemia due to acquired disorders such as myeloproliferative diseases, administration of diuretics or renal failure. This division is somewhat artificial; people with an initial tendency to ‘primary’ hyperuricaemia may develop gout only when secondary factors are introduced – for example obesity, alcohol abuse, or treatment with diuretics or salicylates which increase tubular reabsorption of uric acid.

Clinical features
Patients are usually men over the age of 30 years; women are seldom affected until after the menopause. Often there is a family history of gout. The gouty stereotype is obese, rubicund, hypertensive and fond of alcohol. However, many patients have none of these attributes and some are nudged into an attack by the uncontrolled administration of diuretics or aspirin.

THE ACUTE ATTACK
The sudden onset of severe joint pain which lasts for a week or two before resolving completely is typical of acute gout. The attack usually comes out of the blue but may be precipitated by minor local trauma, operation, intercurrent illness, unaccustomed exercise or alcohol consumption. The commonest sites are the metatarsophalangeal joint of the big toe, the ankle and finger joints, and the olecranon bursa. Occasionally,more than one site is involved. The skin looks red and shiny and there is considerable swelling. The joint feels hot and extremely tender, suggesting a cellulitis or septic arthritis. Sometimes the only feature is acute pain and tenderness in the heel or the sole. Hyperuricaemia is present at some stage, though not necessarily during an acute attack. However, while a low serum uric acid makes

gout unlikely, hyperuricaemia is not ‘diagnostic’ and is often seen in normal middle-aged men. The true diagnosis can be established beyond doubtvby finding the characteristic negatively birefringent uratevcrystals in the synovial fluid. A drop of fluid on a glass slide is examined by polarizing microscopy. Crystals mayvbe sparse but if the fluid specimen is centrifuged a concentratedvpellet may be obtained for examination.
CHRONIC GOUT
Recurrent acute attacks may eventually merge intovpolyarticular gout. Joint erosion causes chronic pain,vstiffness and deformity; if the finger joints arevaffected, this may be mistaken for rheumatoid arthritis.vTophi may appear around joints over the olecranon,vin the pinna of the ear and – less frequently – in almost any other tissue. A large tophus can ulcerate through the skin and discharge its chalky material. Renal lesions include calculi, due to uric acid precipitation in the urine, and parenchymal disease due to deposition of monosodium urate from the blood.

X-rays
During the acute attack x-rays show only soft-tissue swelling. Chronic gout may result in joint space narrowing and secondary osteoarthritis. Tophi appear as characteristic punched-out ‘cysts’ or deep erosions in the para-articular bone ends; these excavations are larger and slightly further from the joint margin than the typical rheumatoid erosions. Occasionally, bone destruction is more marked and may resemble neoplastic disease.

Differential diagnosis

Infection Cellulitis, septic bursitis, an infected bunion or septic arthritis must all be excluded, if necessary by immediate joint aspiration. Remember that crystals and sepsis may coexist, so always send fluid for both culture and crystal analysis.
Reiter’s disease This may present with acute pain and swelling of a knee or ankle, but the history is more protracted and the response to anti-inflammatory drugs less dramatic.
Pseudogout Pyrophosphate crystal deposition may cause an acute arthritis indistinguishable from gout – except that it tends to affect large rather than small joints and is somewhat more common in women than in men. Articular calcification may show on x-ray.
Demonstrating the crystals in synovial fluid establishes the diagnosis.
Rheumatoid arthritis (RA) Polyarticular gout affecting the fingers may be mistaken for rheumatoid arthritis, and elbow tophi for rheumatoid nodules. In difficult cases biopsy will establish the diagnosis. RA and gout seldom occur together.

Treatment

The acute attack The acute attack should be treated by resting the joint, applying ice packs if pain is severe,and giving full doses of a non-steroidal anti-inflammatory drug (NSAID). Colchicine, one of the oldest of medications, is less effective and may cause diarrhoea, nausea and vomiting. A tense joint effusion may require aspiration and intra-articular injection of corticosteroids. Oral corticosteroids are sometimes used for patients who cannot tolerate NSAIDs or in whom NSAIDs are contraindicated. The sooner treatment is started the sooner is the attack likely to end. Interval therapy Between attacks, attention should be given to simple measures such as losing weight, cutting out alcohol and eliminating diuretics. Urate-lowering drug therapy is indicated if acute attacks recur at frequent intervals, if there are tophi or if renal function is impaired. It should also be considered for asymptomatic hyperuricaemia if the plasma urate concentration is persistently above 6 mg/dL (0.36 mmol/L). However, one must remember that this starts a life-long commitment and many clinicians feel that people

who have never had an attack of gout and are free of tophi or urinary calculi do not need treatment.Uricosuric drugs (probenecid or sulfinpyrazone) can be used if renal function is normal. However, allopurinol, a xanthine oxidase inhibitor, is usuallypreferred, and for patients with renal complications or chronic tophaceous gout allopurinol is definitely the drug of choice. Urate-lowering drugs should never be started before the acute attack has completely subsided, and they should always be covered by an anti-inflammatory preparation or colchicine, otherwise they may actually prolong or precipitate an acute attack. Patients who suffer an acute attack of gout while already on a constant dose of urate-lowering treatment should be advised to continue taking the drug at the usual dosage while the acute episode is being treated. Surgery With prolonged urate-lowering therapy, adjusted to maintain a normal serum uric acid level (less than 0.36 mmol/L), tophi may gradually dissolve. However, ulcerating tophi that fail to heal with conservative treatment can be evacuated by curettage; the wound is left open and dressings are applied until it heals.

CALCIUM PYROPHOSPHATE DIHYDRATE ARTHROPATHY (PSEUDOGOUT)

'CPPD deposition' encompasses three overlapping conditions:
(1) chondrocalcinosis – the appearance of calcific material in articular cartilage and menisci;
(2) pseudogout – a crystal-induced synovitis; and
(3) chronic pyrophosphate arthropathy – a type of degenerative joint disease.

Any one of these conditions may occur on its own or in any combination with the others (Dieppe et al., 1982). In contrast to classic gout, serum biochemistry shows no consistent abnormality. CPPD crystal deposition is known to occur in certain metabolic disorders (e.g. hyperparathyroidism and haemochromatosis) that cause a critical change in ionic calcium and pyrophosphate equilibrium in cartilage. The rare familial forms of chondrocalcinosis are probably due to a similar biochemical defect. However, in the vast majority of cases chondrocalcinosis follows some local change in the cartilage due to ageing, degeneration, enzymatic degradation or trauma.

GOUT AND PSEUDOGOUT

GOUT	**PSEUDOGOUT**
Smaller joints	Large joints
Pain intense	Pain moderate
Joint inflamed	Joint swollen
Hyperuricaemia	Chondrocalcinosis
Uric acid crystals	Ca pyrophosphate crystals

Treatment
The treatment of pseudogout is the same as that of acute gout: rest and high-dosage anti-inflammatory therapy. In elderly patients, joint aspiration and intra-articular corticosteroid injection is the treatment of choice as these patients are more vulnerable to the side effects of non-steroidal anti-inflammatory drugs. Chronic chondrocalcinosis appears to be irreversible.

Fortunately it usually causes few symptoms and little disability. When it is associated with progressive joint degeneration the treatment is essentially that of advanced osteoarthritis.

BASIC CALCIUM PHOSPHATE CRYSTAL DEPOSITION DISEASE

Basic calcium phosphate (BCP) is a normal component of bone mineral, in the form of calcium hydroxyapatite crystals. It also occurs abnormally in dead or damaged tissue. Minute deposits in joints and periarticular tissues can give rise to either an acute reaction (synovitis or tendinitis) or a chronic, destructive arthropathy. Prolonged hypercalcaemia or hyperphosphataemia, of whatever cause, may result in widespread metastatic calcification. However, by far the most common cause of BCP crystal deposition in and around joints is local tissue damage – strained or torn ligaments, tendon attrition and cartilage damage or degeneration.

Clinical features

Two clinical syndromes are associated with BCP crystal deposition: (1) an acute or subacute peri-arthritis; and (2) a chronic rapidly destructive arthritis.

ACUTE OR SUBACUTE PERI-ARTHRITIS

This is by far the commonest form of BCP crystal deposition disorder affecting joints. The patient, usually an adult between 30 and 50 years, complains of pain close to one of the larger joints – most commonly the shoulder or the knee. Symptoms may start suddenly, perhaps after minor trauma, and rise to a crescendo during which the tissues around the joint are swollen, warm and exquisitely tender – but tender near the joint in relation to a tendon or ligament, rather than in the joint. At other times the onset is more gradual and it is easier to localize the area of tenderness to one of the periarticular structures. Both forms of the condition are seen most commonly in rotator cuff lesions of the shoulder. Symptoms usually subside after a few weeks or months; sometimes they are aborted only when the calcific deposit is removed or the surrounding tissues are decompressed. In acute cases, operation may disclose a tense globule of creamy material oozing from between the frayed fibres of tendon or ligament.

CHRONIC DESTRUCTIVE ARTHRITIS

BCP crystals are sometimes found in association with a chronic erosive arthritis; whether they cause the arthritis or modify a pre-existing disorder remains uncertain. A more dramatic type of rapidly destructive arthritis of the shoulder is occasionally seen in elderly patients with rotator cuff lesions. This was described in 1981 by McCarty and his colleagues from Milwaukee and acquired the sobriquet ‘Milwaukee shoulder’. Similar conditions affect the hip and knee. They have been attributed to BCP crystal (or mixed BCP and CPPD crystal) shedding into the joint.

Treatment

Acute peri-arthritis should be treated by rest and nonsteroidal anti-inflammatory drugs. Resistant cases may respond to local injection of corticosteroids; this treatment should be used only to weather the acute storm – repeated injections for lesser pain may dampen the repair process in damaged tendons or ligaments and thus predispose to recurrent attacks. Persistent pain and tenderness may call for operative removal of the calcific deposit or ‘decompression’ of the affected tendon or ligament. Erosive arthritis is treated like osteoarthritis. However,

rapidly progressive bone destruction calls for early operation: in the case of the shoulder, synovectomyand soft-tissue repair; for the hip, usually total joint replacement.

5.Osteoarthritis

THE PHYSIOLOGY OF SYNOVIAL JOINTS

ARTICULAR CARTILAGE

Hyaline cartilage, the pearly gristle which covers the bone ends in every diarthrodeal joint, is supremely adapted to transmit load and movement from one skeletal segment to another. It increases the area of the articular surfaces and helps to improve their adaptability and stability; it changes its shape under load and distributes compressive forces widely to the subarticular bone; and, covered by a film of synovial fluid, it is more slippery than any man-made material, offering very little frictional resistance to movement and surface gliding.
This specialized connective tissue has a gel-like matrix consisting of a proteoglycan ground substance in which are embedded an architecturally structured collagen network and arelatively sparse scattering of specialized cells, the chondrocytes, which are responsible for producing all the structural components of the tissue. It has a high water content (60–80 per cent), most of which is exchangeable with the synovial fluid. Chondrocytes of adult hyaline cartilage have little capacity for cell division in vivo and direct damage to the articular surface is poorly repaired, or repaired only with fibrocartilage. The fact that the normal wear of daily joint activity does not result in degradation of the articular surface is due to the highly effective lubricating mechanisms bestowed by synovial fluid. In another sense, though, chondrocytes do undertake repair: in the early stages of cartilage degradation, matrix molecular constituents will be replenished by increased chondrocyte activity. The proteoglycans exist mainly in the form of aggrecan, a large aggregating molecule with a protein core along which are arranged up to 100 chondroitin sulphate and keratan sulphate glycosaminoglycans (GAGs), rather like the bristles on a bottlebrush. Hundreds of aggrecan molecules are linked, in turn, to a long unbranched hyalurinate chain (hyaluronan), to form an even larger molecule with a molecular weight of over 100 million daltons. These negatively charged macromolecules are responsible for the stiffness and springiness of articular cartilage.
The fibrillar component of articular cartilage is mainly type II collagen. The collagen bundles are arranged in structured patterns, parallel to the articular surface in the superficial zones and perpendicular to the surface in the deeper layers where they anchor the articular cartilage to the subchondral bone. The main functions of aggrecan are to absorb changes in load and mitigate deformation, while the collagen network copes with tensile forces. There is considerable interaction between the molecules of each component and between the molecules of the different components of cartilage: if these links are degraded or broken, the cartilage will tend to unravel. This happens to some degree with ageing, but much more so in pathological states leading to osteoarthritis. Proteoglycan has a strong affinity for water, resulting in the collagen network being subjected to considerable tensile stresses. With loading, the cartilage deforms and water is slowly squeezed onto the surface where it helps to form a lubricating film. When loading ceases, the surface fluid seeps back into the cartilage up to the point where the swelling pressure in the cartilage is balanced by the tensile force of the collagennetwork. As long as the network holds and the proteoglycans remain intact,

cartilage retains its compressibility and elasticity. If the collagen network is degraded or disrupted, the matrix becomes waterlogged and soft; this, in turn, is followed by loss of proteoglycans, cellular damage and splitting ('fibrillation') of the articular cartilage. Trouble mounts up further as the damaged chondrocytes begin to release matrix-degrading enzymes.

THREATS TO CARTILAGE INTEGRITY

-Loss of joint stability
-Localized increase in loading stress
-Increased stiffness of the cartilage
-Inflammatory (enzymatic) degradation
-Restriction of free joint movement
-Sclerosis in the subchondral bone

CAPSULE AND LIGAMENTS

The soft tissues enclosing the joint consist of a fibrous capsule with tough condensations on its surface – the ligaments – which, together with the overlying muscles, help to provide stability. The ligaments running from one bone to another are inelastic and have a fixed length. Not surprisingly, therefore, they are under different degrees of tension in different positions of the joint. When the joint assumes a position where the ligaments are fully taut, they provide maximum stability and may keep the joint 'locked' even without the assistance of muscles; when less taut they permit a certain degree of laxity in the joint; and when they are overstretched or torn the joint becomes unstable. Non-pathological ligamentous laxity is a fairly common heritable trait which is employed to astonishing (and sometimes bizarre) effect by acrobatic performers; stability is maintained by highly developed muscle power and the articular cartilage is not necessarily damaged. Inflamed or injured joints that need splinting should always be held in the position where the ligaments are fully taut; if the ligaments are allowed to fibrose and shorten in the 'relaxed' position it may take months (or be impossible) to regain full passive movement afterwards.

Synovium and synovial fluid

The interior surface of the capsule is lined by a thin membrane, the synovium, which is richly supplied with blood vessels, lymphatics and nerves. It provides a nonadherent covering for the articular surfaces and it produces synovial fluid, a viscous plasma dialysate laced with hyaluronan. This fluid nourishes the avascular articular cartilage, plays an important part in reducing friction during movement and has slight adhesive properties which assist in maintaining joint stability. In normal life the volume of synovial fluid in any particular joint remains fairly constant, regardless of movement. When a joint is injured fluid increases (as in any bruised or oedematous connective tissue) and this appears as a joint effusion. Synovium is also the target tissue in joint infections and autoimmune disorders such as rheumatoid arthritis.

MECHANISMS FOR MAINTAINING JOINT STABILITY

-Alignment of joint components
-Shape and fit of articular surfaces
-Adhesive property of synovial fluid
-Integrity of capsule and ligaments
-Muscle tone and power

-Neurological control of balance

Joint lubrication
The coefficient of friction in the normal joint is extremely low – one reason why, barring trauma or disease, there is little difference in the amount of wear on articular surfaces between young adults and old people. This extraordinary slipperiness of cartilage surfaces is produced by a highly efficient combination of lubricating systems. Boundary layer lubrication at the bearing surfaces is mediated by a large, water soluble glycoprotein fraction, lubricin, in the viscous synovial fluid. A single layer of molecules attaches to each articular surface and these glide upon each other in a manner that has been likened to surfaces rolling on miniscule ballbearings. This is most effective at points of direct contact. Fluid film lubrication is provided by the hydrodynamic mechanism described earlier (see under Articular cartilage). During movement and loading fluid is squeezed out of the proteoglycan-rich cartilage and forms a thin 'cushion' where contact is uneven, then seeps back into the cartilage when loading ceases. Lubrication between synovial folds is provided by hyalurinate molecules in the synovial fluid.

OSTEOARTHRITIS

Osteoarthritis (OA) is a chronic disorder of synovial joints in which there is progressive softening and disintegration of articular cartilage accompanied by new growth of cartilage and bone at the joint margins (osteophytes), cyst formation and sclerosis in the subchondral bone, mild synovitis and capsular fibrosis. It differs from simple wear and tear in that it is asymmetrically distributed, often localized to only one part of a joint and often associated with abnormal loading rather than frictional wear. In its most common form, it is unaccompanied by any systemic illness and, although there are sometimes local signs of inflammation, it is not primarily an inflammatory disorder. It is also not a purely degenerative disorder, and the term 'degenerative arthritis' – which is often used as a synonym for OA – is a misnomer. Osteoarthritis is a dynamic phenomenon; it shows features of both destruction and repair. Cartilage softening and disintegration are accompanied from the very outset by hyperactive new bone formation, osteophytosis and remodelling. The final picture is determined by the relative vigour of these opposing processes. In addition, there are various secondary factors which influence the progress of the disorder: the appearance of calcium containing crystals in the joint; ischaemic changes (especially in elderly people) which result in areas of osteonecrosis in the subchondral bone; the appearance of joint instability; and the effects of prolonged anti-inflammatory medication.

Aetiology
The most obvious thing about OA is that it increases in frequency with age. This does not mean that OA is simply an expression of senescence. Cartilage does 'age', showing diminished cellularity, reduced proteoglycan concentration, loss of elasticity and a decrease in breaking strength with advancing years. These factors may well predispose to OA, but it is significant that the progressive changes which are associated with clinical and radiological deterioration are restricted to certain joints, and to specific areas of those joints, while other areas show little or no progression with age (Byers et al., 1970).
Primary changes in cartilage matrix might (theoretically) weaken its structure and thus predispose to cartilage breakdown; crystal deposition disease and ochronosis are well-known

examples. ‘Inheritance’ has for many years been thought to play a role in the development of OA. A number of studies have demonstrated a significant increase in the prevalence of generalized OA in first-degree relatives of patients with OA as compared with controls (Kellgren, 1963) and others have published similar observations for OA of the hip (Lanyon et al., 2000). However, one should bear in mind that OA of large joints is often attributable to anatomical variations, e.g. acetabular dysplasia and other forms of epiphyseal dysplasia, and it is these that are inherited rather than any tendency to develop OA as a primary abnormality. At the molecular level, genetic defects in type II collagen have been demonstrated in some cases (Palotie et al., 1989; Knowlton et al., 1990), but it is unlikely that this is a major aetiological factor in the majority of cases. Articular cartilage may be damaged by trauma or previous inflammatory disorders. Enzymes released by synovial cells and leucocytes can cause leaching of proteoglycans from the matrix, and synovial-derived interleukin-1 (IL-1) may suppress proteoglycan synthesis. This could explain the appearance of ‘secondary’ OA in patients with rheumatoid diseases; whether similar processes operate in ‘primary’ (‘idiopathic’) OA is unknown. In most cases the precipitating cause of OA is increased mechanical stress in some part of the articular surface. This may be due to increased load (e.g. in deformities that affect the lever system around a joint) or to a reduction of the articular contact area (e.g. with joint incongruity or instability). Both factors operate in varus deformity of the knee and in acetabular dysplasia – common precursors of OA. Changes in the subchondral bone may also increase stress concentrationin the overlying cartilage, either by altering the shape of the articular surface or by an increase in bone density (e.g. following fracture healing) which reduces the shock-absorbing effect of the supporting cancellous bone. From the foregoing outline it should be apparent that the division of osteoarthritis into ‘primary’ (when there is no obvious antecedent factor) and ‘secondary’ (when it follows a demonstrable abnormality) is somewhat artificial. This is borne out in clinical practice: patients with ‘secondary’ OA of the knee following meniscectomy have been found also to have a higher than usual incidence of ‘primary’ OA in other joints (Doherty et al., 1983). Perhaps primary, generalized factors (genetic, metabolic or endocrine) alter the physical properties of cartilage and thereby determineis likely to develop OA, while secondary factors such as anatomical defects or trauma specifywhen and where it will occur. OA is, ultimately, more process than disease, occurring in any condition which causes a disparity between the mechanical stress to which articular cartilage is exposed and the ability of the cartilage to withstand that stress.

Pathogenesis

The initial stages of OA have been studied in animal models with induced joint instability and may not be representative of all types of OA. The earliest changes, while the cartilage is still morphologically intact, are an increase in water content of the cartilage and easier extractability of the matrix proteoglycans; similar findings in human cartilage have been ascribed to failure of the internal collagen network that normally restrains the matrix gel. At a slightly later stage there is loss of proteoglycans and defects appear in the cartilage. As the cartilage becomes less stiff, secondary damage to chondrocytes may cause release of cell enzymes and further matrix breakdown. Cartilage deformation may also add to the stress on the collagen network, thus amplifying the changes in a cycle that leads to tissue breakdown. Articular cartilage has an important role in distributing and dissipating the forces associated with joint loading. When it loses its integrity these forces are increasingly concentrated in the subchondral bone.

The result: focal trabecular degeneration and cyst formation, as well as increased vascularity and reactive sclerosis in the zone of maximal loading. What cartilage remains is still capable

of regeneration, repair and remodelling. As the articular surfaces become increasingly malapposed and the joint unstable, cartilage at the edges of the joint reverts to the more youthful activities of growth and endochondral ossification, giving rise to the bony excrescences, or osteophytes, that so clearly distinguish osteoarthritis (once called hypertrophic arthritis') from 'atrophic' disorders such as rheumatoid disease.

Patology

The cardinal features are:
(1) progressive cartilage destruction;
(2) subarticular cyst formation, with
(3) sclerosis of the surrounding bone;
(4) osteophyte formation;
(5) capsular fibrosis.

Initially the cartilaginous and bony changes are confined to one part of the joint – the most heavily loaded part. There is softening and fraying, or fibrillation, of the normally smooth and glistening cartilage.

The term *chondromalacia* (Gr = cartilage softening) seems apt for this stage of the disease, but it is used only of the patellar articular surfaces where it features as one of the causes of anterior knee pain in young people.

With progressive disintegration of cartilage, the underlying bone becomes exposed and some areas may be polished, or burnished, to ivory-like smoothness (eburnation). Sometimes small tufts of fibrocartilage may be seen growing out of the bony surface. At a distance from the damaged area the articular cartilage looks relatively normal, but at the edges of the joint there is remodelling and growth of osteophytes covered by thin, bluish cartilage. Beneath the damaged cartilage the bone is dense and sclerotic. Often within this area of subchondral sclerosis, and immediately subjacent to the surface, are one or more cysts containing thick, gelatinous material. The joint capsule usually shows thickening and fibrosis, sometimes of extraordinary degree. The synovial lining, as a rule, looks only mildly inflamed; sometimes, however, it is thick and red and covered by villi. The histological appearances vary considerably, according to the degree of destruction. Early on, the cartilage shows small irregularities or splits in the surface, while in the deeper layers there is patchy loss of metachromasia (obviously corresponding to the depletion of matrix proteoglycans). Most striking, however, is the increased cellularity, and the appearance of clusters, or clones, of chondrocytes – 20 or more to a batch. In later stages, the clefts become more extensive and in some areas cartilage is lost to the point where the underlying bone is completely denuded. The biochemical abnormalities corresponding to these changes were described by Mankin et al.(1971). The subchondral bone shows marked osteoblastic activity, especially on the deep aspect of any cyst. The cyst itself contains amorphous material; its origin is mysterious – it could arise from stress disintegration of small trabeculae, from local areas of osteonecrosis or from the forceful pumping of synovial fluid through cracks in the subchondral bone plate. As in all types of arthritis, small areas of osteonecrosis are quite common. The osteophytes appear to arise from cartilage hyperplasia and ossification at the edge of the articular surface. The capsule and synovium are often thickened but cellular activity is slight; however, sometimes there is marked inflammation or fibrosis of the capsular tissues. A feature of OA that is difficult to appreciate from the morbid anatomy is the marked vascularity and venous congestion of the subchondral bone. This can be shown by angiographic studies and the demonstration of increased intraosseous pressure. It is also apparent from the intense activity around osteoarthritic joints on radionuclide scanning.

Prevalence

Osteoarthritis is the commonest of all joint diseases. It is a truly universal disorder, affecting both sexes and all races; everyone who lives long enough will have it somewhere, in some degree. However, there are significant differences in its rate of occurrence in different ethnic groups, in the different sexes within any group, and in the different joints.
Reports of prevalence rates vary, depending on the method of evaluation. Autopsy studies show OA changes in everyone over the age of 65 years. Radiographic surveys suggest that the prevalence rises from 1 per cent below the age of 30 years to over 50 per cent in people above the age of 60. Osteoarthritis of the finger joints is particularly common in elderly women, affecting more than 70 per cent of those over 70 years. Men and women are equally likely to develop OA, but more joints are affected in women than in men.Osteoarthritis is much more common in some joints (the fingers, hip, knee and spine) than in others (the elbow, wrist and ankle). This may simply reflect the fact that some joints are more prone to predisposing abnormalities than others.
A similar explanation may account for certain geographical and ethnic differences in prevalence. For example, the female-to-male ratio for OA of the hip is about 1:1 in northern Europe but is nearer 2:1 in southern Europe where there is a high incidence of acetabular dysplasia in girls. Even more striking is the virtual absence of hip OA in southern Chinese and African blacks (Hoagland et al., 1973; Solomon, 1976); this may simply be because predisposing disorders such as developmental displacement of the hip, Perthes' disease and slipped femoral epiphysis are uncommon in these populations. That they have no inherent resistance to OA is shown by the fact that they often develop the condition in other joints, for example the knee.

Risk factors
Joint dysplasia Disorders such as congenital acetabular dysplasia and Perthes' disease presage a greater than normal risk of OA in later life. It is not always easy to spot minor degrees of dysplasia and careful studies may have to be undertaken if these are not to bemissed.
Trauma Fractures involving the articular surface are obvious precursors of secondary OA, so too lesser injuries which result in joint instability. What is less certain is whether malunion of a long-bone fracture predisposes to OA by causing segmental overload in a joint above or below the healed fracture (for example, in the knee or ankle after a tibial fracture). Contrary to popular belief, research has shown that moderate angular deformities of the tibia (up to 15 degrees) are not associated with an increased risk of OA (Merchant and Dietz, 1989). This applies to mid-shaft fractures; malunion close to a joint may well predispose to secondary OA.
Occupation There is good evidence of an association between OA and certain occupations which cause repetitive stress, for example OA of the knees in workers engaged in knee-bending activities (Felson, 1991), OA in the upper limbs in people working with heavy vibrating tools (Schumacher et al., 1972) and OA of the hands in cotton mill workers (Lawrence, 1961). More controversial is the relationship of OA to sporting activity. Boxers are certainly prone to developing OA of the hands but this may be due to trauma. The same applies to footballers with OA of the knees and baseball pitchers with OA of the shoulder. More convincingevidence of a causative relationship comes from recent studies which have shown a significant increase in the risk of hip and knee OA in athletes (Harris et al., 1994; Kulkala et al., 1994).
Bone density It has long been known that women with femoral neck fractures seldom have OA of the hip. This negative association between OA and osteoporosis is reflected in more recent studies which have demonstrated a significant increase in bone mineral density in people with OA compared to those without (Hannan et al., 1992; Hart et al., 1994). However, this may not be simple cause and effect: bone density is determined by a variety of

genetic, hormonal and metabolic factors which may also influence cartilage metabolism independently of any effect due to bone density.
Obesity The simple idea that obesity causes increased joint loading and therefore predisposes to OA may be correct – at least in part. The association is closer in women than in men and therefore (as with bone density) it may reflect other endocrine or metabolic factors in the pathogenesis of OA.
Family history Women whose mothers had generalized OA are more likely to develop the same condition. The particular trait responsible for this is not known .

Symptoms
Patients usually present after middle age. Joint involve -
ment follows several different patterns: symptoms
centre either on one or two of the weightbearing joints(hip or knee), on the interphalangeal joints (especially in women) or on any joint that has suffered a previous affliction (e.g. congenital dysplasia, osteonecrosis or intra-articular fracture). A family history is common in patients with polyarticular OA. Pain is the usual presenting symptom. It is often quite widespread, or it may be referred to a distant site – for example, pain in the knee from OA of the hip. It starts insidiously and increases slowly over months or years. It is aggravated by exertion and relieved by rest, although with time relief is less and less complete. In the late stage the patient may have pain in bed at night. There are several possible causes of pain: mild synovial inflammation, capsular fibrosis with pain on stretching the shrunken tissue; muscular fatigue; and, perhaps most important of all, bone pressure due to vascular congestion and intraosseous hypertension. Stiffness is common; characteristically it occurs after periods of inactivity, but with time it becomes constant and progressive. Swelling may be intermittent (suggesting an effusion) or continuous (with capsular thickening or large osteophytes). Deformity may result from capsular contracture or joint instability, but be aware that the deformity may actually have preceded and contributed to the onset of OA.
Loss of function, though not the most dramatic, is often the most distressing symptom. A limp, difficulty in climbing stairs, restriction of walking distance, or progressive inability to perform everyday tasks or enjoy recreation may eventually drive the patient to seek help. Typically, the symptoms of OA follow an intermittent course, with periods of remission sometimes lasting for months.

Signs
Joint swelling may be the first thing one notices in peripheral joints (especially the fingers, wrists, knees and toes). This may be due to an effusion, but hard (‘knobbly’) ridges around the margins of the distal interphalangeal, the first metatarsophalangeal or knee joints can be just as obvious. Tell-tale scars denote previous abnormalities, and muscle wasting suggests longstanding dysfunction. Deformity is easily spotted in exposed joints (the knee or the large-toe metatarsophalangeal joint), but deformity of the hip can be masked by postural adjustments of the pelvis and spine.
Local tenderness is common, and in superficial joints fluid, synovial thickening or osteophytes may be felt. Limited movement in some directions but not others is usually a feature, and is sometimes associated with pain at the extremes of motion. Crepitus may be felt over the joint (most obvious in the knee) during passive movements. Instability is common in the late stages of articular destruction, but it may be detected much earlier by special testing. Instability can be due to loss of cartilage and bone, asymmetrical capsular contracture and/or muscle weakness.
Other joints should always be examined; they may show signs of a more generalized disorder. It is also helpful to know whether problems in other joints add to the difficulties in the one

complained of (e.g. a stiff lumbar spine or an unstable knee making it more difficult to cope with restricted movement in an osteoarthritic hip). Function in everyday activities must be assessed. X-ray appearances do not always correlate with either the degree of pain or the patient's actual functional capacity. Can the patient with an arthritic knee walk up and down stairs, or rise easily from a chair? Does he or she limp? Or use a walking stick?

Imaging
X-rays X-ray appearances are so characteristic that other forms of imaging are seldom necessary for ordinary clinical assessment. The cardinal signs are asymmetrical loss of cartilage (narrowing of the 'joint space'), sclerosis of the subchondral bone under the area of cartilage loss, cysts close to the articular surface, osteophytes at the margins of the joint and remodelling of the bone ends on either side of the joint. Late features may include joint displacement and bone destruction. Look carefully for signs of previous disorders (e.g. congenital defects, old fractures, Perthes' disease or rheumatoid arthritis). Such cases are usually designated as 'secondary osteoarthritis', though in a certain sense OA is always secondary to some previous abnormality if only we could discover what it was!
Radionuclide scanning Scanning with 99mTc-HDP shows increased activity during the bone phase in the subchondral regions of affected joints. This is due to increased vascularity and new bone formation. CT and MRI Advanced imaging is sometimes needed to elucidate a specific problem, e.g. early detection of an osteocartilaginous fracture, bone oedema or avascular necrosis. These methods are also used for severity grading in clinical trials.

Arthroscopy
Arthroscopy may show cartilage damage before x-ray changes appear. The problem is that it reveals too much, and the patient's symptoms may be ascribed to chondromalacia or OA when they are, in fact, due to some other disorder.

Natural history
Osteoarthritis usually evolves as a slowly progressive disorder. However, symptoms characteristically wax and wane in intensity, sometimes disappearing for several months. The x-rays show no such fluctuation. However, there is considerable variation between patients in the degrees of destruction and repair. Most of the men and half of the women have a hypertrophic reaction, with marked sclerosis and large osteophytes. In about 20 per cent of cases – most of them women – reactive changes are more subdued, inviting descriptions such as atrophic or osteopaenic OA. Occasionally OA takesthe form of a rapidly progressive disorder (Solomon, 1976; Solomon, 1984).

Complications
Capsular herniation Osteoarthritis of the knee is sometimes associated with a marked effusion and herniation of the posterior capsule (Baker's cyst). Loose bodies Cartilage and bone fragments may give rise to loose bodies, resulting in episodes of locking. Rotator cuff dysfunction Osteoarthritis of the acromioclavicular joint may cause rotator cuff impingement, tendinitis or cuff tears. Spinal stenosis Longstanding hypertrophic OA of the lumbar apophyseal joints may give rise to acquired spinal stenosis. The abnormality is best demonstrated by CT and MRI. Spondylolisthesis In patients over 60 years of age, destructive OA of the apophyseal joints may result in severe segmental instability and spondylolisthesis (socalled 'degenerative' spondylolisthesis, which almost
always occurs at L4/5).
Clinical variants of osteoarthritis

Although the features of OA in any particular joint are fairly consistent, the overall clinical picture shows variations which define a number of subgroups.

MONARTICULAR AND PAUCIARTICULAR OSTEOARTHRITIS

In its 'classic' form, OA presents with pain and dysfunction in one or two of the large weightbearing joints. There may be an obvious underlying abnormality: multiple epiphyseal dysplasia, localized acetabular dysplasia, old Perthes' disease, previous slipped epiphysis, inflammatory joint disease, avascular necrosis, a previous fracture or damage to ligaments or menisci. In the majority, however, the abnormality is more subtle and may come to light only with special imaging techniques.

POLYARTICULAR (GENERALIZED) OSTEOARTHRITIS

This is by far the most common form of OA, though most of the patients never consult an orthopaedic surgeon. The patient is usually a middle-aged woman who presents with pain, swelling and stiffness of the finger joints. The first carpometacarpal and the big toe metatarsophalangeal joints, the knees and the lumbar facet joints may be affected at more or less the same time. The changes are most obvious in the hands. The interphalangeal joints become swollen and tender, and in the early stages they often appear to be inflamed. Over a period of years osteophytes and softtissue swelling produce a characteristic knobbly appearance of the distal interphalangeal joints (Herberden's nodes) and, less often, the proximal interphalangeal joints (Bouchard's nodes); pain may later disappear but stiffness and deformity persist. Some patients present with painful knees or backache and the knobbly fingers are noticed only in passing. There is a strong association with carpal tunnel syndrome and isolated tenovaginitis. X-rays show the characteristic features of OA, usually maximal in the distal interphalangeal joints of the fingers.

OSTEOARTHRITIS IN UNUSUAL SITES

Osteoarthritis is uncommon in the shoulder, elbow, wrist and ankle. If any of these joints is affected one should suspect a previous abnormality – congenital or traumatic – or an associated generalized disease such as a crystal arthropathy.

RAPIDLY DESTRUCTIVE OSTEOARTHRITIS (see also page 84)

Every so often a patient with apparently straightforward OA shows rapid and startling progression of bone destruction. The condition was at one time thought to be due to the dampening of pain impulses by powerful anti-inflammatory drugs – a notional type of 'analgesic arthropathy'. It is now recognized that it occurs mainly in elderly women and that it is associated with the deposition of calcium pyrophosphate dihydrate crystals, though whether this is the cause of the condition or a consequence thereof is still undecided.

Differential diagnosis of osteoarthritis

A number of conditions may mimic OA, some presenting as a monarthritis and some as a polyarthritis affecting the finger joint.

Avascular necrosis 'Idiopathic' osteonecrosis causes joint pain and local effusion. Early on the diagnosis is made by MRI. Later x-ray appearances are usually pathognomonic; however, once bone destruction occurs the x-ray changes can be mistaken for those of OA. The cardinal distinguishing feature is that in osteonecrosis the 'joint space' (articular cartilage) is preserved in the face of progressive bone collapse and deformity, whereas in OA articular cartilage loss precedesbone destruction.

Inflammatory arthropathies Rheumatoid arthritis, ankylosing spondylitis and Reiter's disease may start in one or two large joints. The history is short and there are local signs of

inflammation. X-rays show a predominantly atrophic or erosive arthritis. Sooner or later other joints are affected and systemic features appear. Polyarthritis of the fingers Polyarticular OA may be confused with other disorders which affect the finger joints (see Fig. 5.10). Close observation shows several distinguishing features. Nodal OA affects predominantly the distal joints, rheumatoid arthritis the proximal joints. Psoriatic arthritis is a purely destructive arthropathy and there are no interphalangeal 'nodes'. Tophaceous gout may cause knobbly fingers, but the knobs are tophi, not osteophytes. X-rays will show the difference.
Diffuse idiopathic skeletal hyperostosis (DISH) This is a fairly common disorder of middle-aged people, characterized by bone proliferation at the ligament and tendon insertions around peripheral joints and the intervertebral discs (Resnick et al., 1975). On x-ray examination the large bony spurs are easily mistaken for osteophytes. DISH and OA often appear together, but DISH is not OA: the bone spurs are symmetrically distributed, especially along the pelvic apophyses and throughout the vertebral column. When DISH occurs by itself it is usually asymptomatic.
Multiple diagnosis Osteoarthritis is so common after middle age that it is often found in patients with other conditions that cause pain in or around a joint. Before jumping to the conclusion that the symptoms are due to the OA features seen on x-ray, be sure to exclude peri-articular disorders as well as more distant abnormalities giving rise to referred pain.

Management
The management of OA depends on the joint (or joints) involved, the stage of the disorder, the severity of the symptoms, the age of the patient and his or her functional needs. Three observations should be borne in mind: (1) symptoms characteristically wax and wane, and pain may subside spontaneously for long periods; (2) some forms of OA actually become less painful with the passage of time and the patient may need no more than reassurance and a prescription for pain killers; (3) at the other extreme, the recognition (from serial x-rays) that the patient has a rapidly progressive type of OA may warrant an early move to reconstructive surgery before bone loss compromises the outcome of any operation.

EARLY TREATMENT
There is, as yet, no drug that can modify the effects of OA. Treatment is, therefore, symptomatic. The principles are:
(1) maintain movement and muscle strength;
(2) protect the joint from 'overload';
(3)relieve pain;
(4) modify daily activities.
Physical therapy The mainstay of treatment in the early case is physical therapy, which should be directed at maintaining joint mobility and improving muscle strength. The programme can include aerobic exercise, but care should be taken to avoid activities which increase impact loading. Other measures, such as massage and the application of warmth, may reduce pain but improvement is short-lived and the treatment has to be repeated.
Load reduction Protecting the joint from excessive load may slow down the rate of cartilage loss. It is also effective in relieving pain. Common sense measures such as weight reduction for obese patients, wearing shock-absorbing shoes, avoiding activities like climbing stairs and using a walking stick are worthwhile.

Analgesic medication Pain relief is important, but not all patients require drug therapy and those who do may not need it all the time. If other measures do not provide symptomatic improvement, patients may respond to a simple analgesic such as paracetamol. If this fails to control pain, a non-steroidal anti-inflammatory preparation may be better.

INTERMEDIATE TREATMENT

Joint debridement (removal of loose bodies, cartilage tags, interfering osteophytes or a torn or impinging acetabular or glenoid labrum) may give some improvement. This may be done either by arthroscopy or by open operation. If appropriate radiographic images suggest that symptoms are due to localized articular overload arising from joint malalignment (e.g. varus deformity of the knee) or incongruity (e.g. acetabular and femoral head dysplasia), a corrective osteotomy may prevent or delay progression of the cartilage damage.

LATE TREATMENT

Progressive joint destruction, with increasing pain, instability and deformity (particularly of one of the weightbearing joints), usually requires reconstructive surgery. Three types of operation have, at different times, held the field: realignment osteotomy, arthroplasty and arthrodesis.

Realignment osteotomy Until the development of joint replacement surgery in the 1970s, realignment osteotomy was widely employed. Refinements in techniques, fixation devices and instrumentation led to acceptable results from operations on the hip and knee, ensuring that this approach has not been completely abandoned. High tibial osteotomy is still considered to be a viable alternative to partial joint replacement for unicompartmental OA of the knee, and intertrochanteric femoral osteotomy is sometimes preferred for young patients with localized destructive OA of the hip. These operations should be done while the joint is still stable and mobile and x-rays show that a major part of the articular surface (the radiographic 'joint space') is preserved. Pain relief is often dramatic and is ascribed to (1) vascular decompression of the subchondral bone, and (2) redistribution of loading forces towards less damaged parts of the joint. After load redistribution, fibrocartilage may grow to cover exposed bone.

Joint replacement Joint replacement, in one form or another, is nowadays the procedure of choice for OA in patients with intolerable symptoms, marked loss of function and severe restriction of daily activities. For OA of the hip and knee in middle-aged and older patients, total joint replacement by modern techniques promises improvement lasting for 15 years or longer. Similar operations for the shoulder, elbow and ankle are less successful but techniques are improving year by year. However, joint replacement operations are highly dependent on technical skills, implant design, appropriate instrumentation and postoperative care – requirements that cannot always be met, or may not be cost-effective, in all parts of the world.

Arthrodesis Arthrodesis is still a reasonable choice if the stiffness is acceptable and neighbouring joints are not likely to be prejudiced. This is most likely to apply to small joints that are prone to OA, e.g. the carpal and tarsal joints and the large toe metatarsophalangeal joint.

NEUROPATHIC JOINT DISEASE

Charcot, in 1868, described a type of destructive arthropathy associated with disease of the central nervous system. Almost all his patients had tabes dorsalis,but the name ‘Charcot’s joint disease’ came to be applied to any destructive arthropathy arising from loss of pain sensibility and position sense. Nowadays the most common cause is diabetic neuropathy, which occurs in 0.2–0.5 per cent of patients with diabetes mellitus; other causes are tabes dorsalis, leprosy (affecting mainly the lower limb joints), syringomyelia (upper and lower limbs), multiple sclerosis, myelomeningocele, spinal cord compression and congenital indifference to pain. The term is also applied (less accurately) to rapidly destructive forms of osteoarthritis where there is no neurological lesion.

HAEMOPHILIC ARTHROPATHY

Recurrent intra-articular bleeding may lead to chronic synovitis and progressive articular destruction. Clinically this is seen only in classic haemophilia, in which there is a deficiency of clotting factor VIII, and Christmas disease, due to deficiency of factor IX. Both are X-linked recessive disorders manifesting in males but carried by females. Their incidence is about 1 per 10 000 male births. Plasma clotting factor levels above 40 per cent of the normal are compatible with normal control of haemorrhage. Patients with clotting factor levels above 5 per cent (‘mild haemophilia’) may have prolonged bleeding after injury or operation; those with levels below 1 per cent (‘severe haemophilia’) have frequent spontaneous joint and muscle haemorrhages.

Metabolic and endocrine disorders 7

Metabolic bone disorders are associated with critical alterations in the regulation of bone formation, bone resorption and distribution of minerals in bone. Clinical features arise from both systemic responses to changes in mineral exchange and local effects of abnormal bone structure and composition. Orthopaedic surgeons deal mainly with the bone abnormalities (e.g. rickety deformities in growing bones or insufficiency fractures in the elderly) but it is important also to be aware of the systemic disorders that may lie behind apparently straightforward 'orthopaedic' defects and to understand the unseen metabolic changes that influence the outcome of many of our surgical interventions.

OSTEOPOROSIS

Osteoporosis as a clinical disorder is characterized by an abnormally low bone mass and defects in bone structure, a combination which renders the bone unusually fragile and at greater than normal risk of fracture in a person of that age, sex and race. Although the cancellous regions are more porous and the cortices thinner than normal, the existing bone is fully mineralized. Bone depletion may be brought about by predominant bone resorption, decreased bone formation or a combination of the two. It seems self-evident that the main reason for the loss of bone strength is the reduction in bone mass; however, in the remaining trabecular bone there may also be a loss of structural connectivity between bone plates, and this so alters the mechanical properties that the loss of strength is out of proportion to the diminution in bone mass. As a consequence, the bone – particularly around the diaphyseo-metaphyseal junctions in tubular bones and in the mainly cancellous vertebral bodies – eventually reaches a state in which a comparatively modest stress or strain causes a fracture. For reasons that are not fully understood, black African peoples are considerably less prone to these effects and have a low incidence of 'osteoprotic fractures' (Solomon, 1968). This section deals with generalized osteoporosis, but it should not be forgotten that osteoporosis is sometimes confined to a particular bone or group of bones – regional osteoporosis (for example due to disuse, immobilization or inflammation) – which is usually reversible once the local cause is addressed.

X-rays and bone densitometry

The term osteopaenia is sometimes used to describe bone which appears to be less 'dense' than normal on x-ray, without defining whether the loss of density is due to osteoporosis or osteomalacia, or indeed whether it is sufficiently marked to be regarded as at all pathological. More characteristic signs of osteoporosis are loss of trabecular definition, thinning of the cortices and insufficiency fractures. Compression fractures of the vertebral bodies, wedging at multiple levels or biconcave distortion of the vertebral end-plates due to bulging of intact

intervertebral discs are typical of severe postmenopausal osteoporosis. The clinical and radiographic diagnosis should be backed up by assessment of BMD as measured by DXA of the spine and hips, using the lower value of the two. In otherwise 'normal' women over the age of 50 years, anything more than 2 standard deviations below the average for the relevant population group may be taken as indicative of osteoporosis.

POSTMENOPAUSAL OSTEOPOROSIS

Symptomatic postmenopausal osteoporosis is an exaggerated form of the physiological bone depletion that normally accompanies ageing and loss of gonadal activity. Two overlapping phases are recognized: anearly postmenopausal syndrome characterized by rapid bone loss due predominantly to increased osteoclastic resorption (high-turnover osteoporosis) and a less well-defined syndrome which emerges in elderly people and is due to a gradual slow-down in osteoblastic activity and the increasing effects of dietary insufficiencies, chronic ill health and reduced mobility (low-turnover osteoporosis). Around the menopause, and for the next 10 years, bone loss normally accelerates to about 3 per cent per year compared with 0.3 per cent during the preceding two decades. This is due mainly to increased bone resorption, the withdrawal of oestrogen having removed one of the normal restraints on osteoclastic activity. Genetic influences play an important part in determining when and how this process becomes exaggerated, but a number of other risk factors have been identified .

RISK FACTORS FOR POSTMENOPAUSAL OSTEOPOROSIS

Caucasoid (white) or Asiatic ethnicity
Family history of osteoporosis
History of anorexia nervosa and/or amenorrhoea
Low peak bone mass in the third decade
Early onset of menopause
Unusually slim or emaciated build
Oophorectomy
Early hysterectomy
Nutritional insufficiency
Chronic lack of exercise
Cigarette smoking
Alcohol abuse

Clinical features and diagnosis

A woman at or near the menopause develops back pain and increased thoracic kyphosis; she, or someone in the family, may have noticed that her height has diminished. X-rays of the spine may show wedging or compression of one or more vertebral bodies and often the lateral view also shows calcification of the aorta. This is the typical picture, but sometimes the first clinical event is a low-energy fracture of the distal radius (Colles' fracture), the hip or the ankle. Women who have had one low-energy fracture have twice the normal risk of developing another.

DXA may show significantly reduced bone density in the vertebral bodies or femoral neck. The rate of bone turnover is either normal or slightly increased; measurement of excreted collagen cross-link products and telopeptides may suggest a high-turnover type of bone loss.

Once the clinical diagnosis has been established, screening tests should be performed to rule out other causes of osteoporosis (e.g. hyperparathyroidism, malignant disease or hypercortisonism).

Prevention and treatment

Bone densitometry can be used to identify women who are at more than usual risk of suffering a fracture at the menopause, and prophylactic treatment of this group is sensible. However, routine DXA screening (even in countries where it is available) is still not universally employed; for practical purposes, it is usually reserved for women with multiple risk factors and particularly those with suspected oestrogen deficiency (premature or surgically induced menopause) or some other bone-losing disorder, and those who have already suffered previous low-energy fractures at the menopause. Women approaching the menopause should be advised to maintain adequate levels of dietary calcium and vitamin D, to keep up a high level of physical activity and to avoid smoking and excessive consumption of alcohol. If necessary, the recommended daily requirements should be met by taking calcium and vitamin D supplements; these measures have been shown to reduce the risk of low-energy fractures in elderly women (Chapuy et al., 1994).

Hormone replacement therapy (HRT) Until the beginning of the twenty-first century HRT was the most widely used medication for postmenopausal osteoporosis. Taking oestrogen (or a combination of oestrogen and progesterone) for 5–10 years was shown convincingly to reduce the risk of osteoporotic fractures, though after stopping the medication the BMD gradually falls to the usual low level. Moreover there was growing concern about the apparent increased risks of thromboembolism, stroke, breast cancer and uterine cancer. As more experience has been gained with other antiresorptive drugs, the preference for HRT has waned.

Bisphosphonates Bisphosphonates are now regarded as the preferred medication for postmenopausal osteoporosis. They act by reducing osteoclastic bone resorption and the general rate of bone turnover. The newer preparations have been shown to prevent bone loss and to reduce the risk of vertebral and hip fractures. Alendronate can be administered by mouth in once-weekly doses for both prevention and treatment of osteoporosis. Gastrointestinal side effects are a bother and suitable precautions should be taken; for patients who cannot tolerate the drug, pamidronate has been given intravenously at 3-monthly intervals.

Parathyroid hormone Trials of parathyroid hormone, either by itself or in combination with alendronate, have shown good results in obtaining a rise in BMD in patients with postmenopausal osteoporosis (Black et al., 2005). This could be a way of managing patients with severe osteoporosis who do not respond to bisphosphonates alone.

Recent advances in treatment A novel way of reducing osteoclastic activity and bone resorption is to interrupt the RANKL–RANK interaction which is essential for prompting osteoclastogenesis (see page 122). Phase 3 trials are now being conducted using denosumab, an antibody to RANKL, which holds out the promise of an effective new line of treatment for postmenopausal osteoporosis (McClung et al., 2006).

Management of fractures Femoral neck and other long-bone fractures may need operative treatment.Methods are described in the relevant chapters in Section 3.

Vertebral fractures are painful and patients will need analgesic treatment, partial rest and assistance with personal care for about 6 weeks. Physiotherapy should initially be aimed at maintaining muscle tone and movement in all unaffected areas; if pain is adequately controlled, patients should be encouraged to walk and when symptoms allow they can be introduced to postural training. Spinal orthoses may be needed for support and pain relief, but

they cannot be expected to correct any structural deformity. Operative measures are occasionally called for to treat severe compression fractures.

INVOLUTIONAL OSTEOPOROSIS

In advanced age the rate of bone loss slowly decreases but the incidence of femoral neck and vertebral fractures rises steadily; by around 75 years of age almost a third of white women will have at least one vertebral fracture. For reasons that are not completely known, age-related fractures are much less common in black people. BMD measurements in this age group show that there is considerable overlap between those who suffer fractures and those who do not; the assumption is that qualitative changes contribute increasingly to bone fragility in old age. Causes include a rising incidence of chronic illness, mild urinary insufficiency, dietary deficiency, lack of exposure to sunlight, muscular atrophy, loss of balance and an increased tendency to fall. Many old people suffer from vitamin D deficiency and develop some degree of osteomalacia on top of the postmenopausal osteoporosis (Solomon, 1973).

Treatment Initially, treatment is directed at management of the fracture. This will often require internal fixation; the sooner these patients are mobilized and rehabilitated the better. Patients with muscle weakness and/or poor balance may benefit from gait training and, if necessary, the use of walking aids and rail fittings in the home. Thereafter the question of general treatment must be considered. Obvious factors such as concurrent illness, dietary deficiencies, lack of exposure to sunlight and lack of exercise will need attention. If the patient is not already on vitamin D and calcium as well as antiresorptive medication, this should be prescribed; although bone mass will not be restored, at least further loss may be slowed.

POST-CLIMACTERIC OSTEOPOROSIS IN MEN

With the gradual depletion in androgenic hormones, men eventually suffer the same bone changes as postmenopausal women, only this occurs about 15 years later unless there is some specific cause for testicular failure. Osteoporotic fractures in men under 60 years of age should arouse the suspicion of some underlying disorder– notably hypogonadism, metastatic bone disease, multiple myeloma, liver disease, renal hypercalciuria, alcohol abuse, malabsorption disorder, malnutrition, glucocorticoid medication or anti-gonadal hormone treatment for prostate cancer. Other causes of secondary osteoporosis are shown in Table 7.4. Treatment is much the same as for postmenopausal osteoporosis. Vitamin D and calcium supplementation is important; alendronate is the antiresorptive drug of choice. If testosterone levels are unusually low, hormone treatment should be considered.

SECONDARY OSTEOPOROSIS

Among the numerous causes of secondary osteoporosis, hypercortisonism, gonadal hormone deficiency, hyperthyroidism, multiple myeloma, chronic alcoholism and immobilization will be considered further.

Hypercortisonism

Glucocorticoid overload occurs in endogenous Cushing's disease or after prolonged treatment with corticosteroids. This often results in severe osteoporosis, especially if the condition for which the drug is administered is itself associated with bone loss – for example, rheumatoid arthritis. Glucocorticoids have a complex mode of action. The deleterious effect on bone is mainly by suppression of osteoblast function, but it also causes reduced calcium absorption, increased calcium excretion and stimulation of PTH secretion (Hahn, 1980). There is now

evidence that it also depresses OPG expression and this would have an enhancing effect on osteoclastogenesis and bone resorption.
Treatment presents a problem, because the drug may be essential for the control of some generalized disease.
However, forewarned is forearmed: corticosteroid dosage should be kept to a minimum, and it should not be forgotten that intra-articular preparations and cortisone ointments are absorbed and may have systemic effects if given in high dosage or for prolonged periods. Patients on long-term glucocorticoid treatment should, ideally, be monitored for bone density. Preventive measures include the use of calcium supplements (at least 1500 mg per day) and vitamin D metabolites. In postmenopausal women and elderly men bisphosphonates may be effective in reducing bone resorption.In late cases general measures to control bone pain may be required. Fractures are treated as and when they occur.

Gonadal hormone insufficiency

Oestrogen lack is an important factor in postmenopausal osteoporosis. It also accounts for osteoporosis in younger women who have undergone oophorectomy, and in pubertal girls with ovarian agenesis and primary amenorrhoea (Turner's syndrome). Treatment is the same as for postmenopausal osteoporosis. Amenorrhoeic female athletes, and adolescents with anorexia nervosa, may become osteoporotic; fortunately these conditions are usually self-limiting. A decline in testicular function probably contributes to the continuing bone loss and rising fracture rate in men over 70 years of age. A more obvious relationship is found in young men with overt hypogonadism; this may require long-term treatment with testosterone.

Hyperthyroidism

Thyroxine speeds up the rate of bone turnover, but resorption exceeds formation. Osteoporosis is quite common in hyperthyroidism, but fractures usually occur only in older people who suffer the cumulative effects of the menopause and thyroid overload. In the worst cases osteoporosis may be severe with spontaneous fractures, a marked rise in serum alkaline phosphatase, hypercalcaemia and hypercalciuria. Treat ment is needed for both the osteoporosis and the thyro toxicosis.

Multiple myeloma and carcinomatosis

Generalized osteoporosis, anaemia and a high ESR are characteristic features of myelomatosis and metastatic bone disease. Bone loss is due to overproduction of local osteoclast-activating factors. Treatment with bisphosphonates may reduce the risk of fracture.

Alcohol abuse

This is a common (and often neglected) cause of osteoporosis at all ages, with the added factor of an increased tendency to falls and other injuries. Bone changes are due to a combination of decreased calcium absorption, liver failure and a toxic effect on osteoblast function. Alcohol also has a mild glucocorticoid effect.

Immobilization

The worst effects of stress reduction are seen in states of weightlessness; bone resorption, unbalanced by formation, leads to hypercalcaemia, hypercalciuria and severe osteoporosis. Lesser degrees of osteoporosis are seen in bedridden patients, and regional osteoporosis is common after immobilization of a limb. The effects can be mitigated by encouraging mobility, exercise and weightbearing.

Other conditions

There are many other causes of secondary osteoporosis, including hyperparathyroidism (which is considered below), rheumatoid arthritis, ankylosing spondylitis and subclinical forms of osteogenesis imperfecta. The associated clinical features usually point to the diagnosis.

Causes of secondary osteoporosis

Nutritional
Malabsorption
Malnutrition
Scurvy
Inflammatory disorders
Rheumatoid disease
Ankylosing spondylitis
Tuberculosis
Drug induced
Corticosteroids
Excessive alcohol consumption
Anticonvulsants
Heparin
Immunosuppressives

Endocrine disorders
Gonadal insufficiency
Hyperparathyroidism
Thyrotoxicosis
Cushing's disease

Malignant disease
Carcinomatosis
Multiple myeloma
Leukaemia

Other
Smoking
Chronic obstructive pulmonary disease
Osteogenesis imperfecta
Chronic renal disease

RICKETS AND OSTEOMALACIA

Rickets and osteomalacia are different expressions of the same disease: inadequate mineralization of bone. Osteoid throughout the skeleton is incompletely calcified, and the bone is therefore 'softened' (osteomalacia). In children there are additional effects on physeal growth and ossification, resulting in deformities of the endochondral skeleton (rickets).
The inadequacy may be due to defects anywhere along the metabolic pathway for vitamin D: nutritional lack, underexposure to sunlight, intestinal malabsorption, decreased 25-hydroxylation (liver disease, anticonvulsants) and reduced 1α-hydroxylation (renal disease, nephrectomy, 1α-hydroxylase deficiency). The pathological changes may also be caused by calcium deficiency or hypophosphataemia.

Pathology

The characteristic pathological changes in rickets arise from the inability to calcify the intercellular matrix in the deeper layers of the physis. The proliferative zone is as active as ever, but the cells, instead of arranging themselves in orderly columns, pile up irregularly; the entire physeal plate increases in thickness, the zone of calcification is poorly mineralized and bone formation is sparse in the zone of ossification. The new trabeculae are thin and weak, and with joint loading the juxta-epiphyseal metaphysis becomes broad and cupshaped. Away from the physis the changes are essentially those of osteomalacia. Sparse islands of bone are lined by wide osteoid seams, producing unmineralized ghost trabeculae that are not very strong. The cortices also are thinner than normal and may show signs of new or older stress fractures. If the condition has been present for a long time there may be stress deformities of the bones: indentation of the pelvis, bending of the femoral neck (coxa vara) and bowing of the femora and tibiae. Remember that even mild osteomalacia can increase the risk of fracture if it is superimposed on postmenopausal or senile osteoporosis.

Clinical features of rickets and osteomalacia

In the past the vast majority of cases of rickets and osteomalacia were due to dietary vitamin D deficiency and/or insufficient exposure to sunlight. These patients still embody the classical picture of the disorder.

Children The infant with rickets may present with tetany or convulsions. Later the parents may notice that there is a failure to thrive, listlessness and muscular flaccidity. Early bone changes are deformity of the skull (craniotabes) and thickening of the knees, ankles and wrists from physeal overgrowth. Enlargement of the costochondral junctions ('rickety rosary') and lateral indentation of the chest (Harrison's sulcus) may also appear. Distal tibial bowing has been attributed to sitting or lying cross-legged. Once the child stands, lower limb deformities increase, and stunting of growth may be obvious. In severe rickets there may be spinal curvature, coxa vara and bending or fractures of the long bones.

Adults Osteomalacia has a much more insidious course and patients may complain of bone pain, backache and muscle weakness for many years before the diagnosis is made. Vertebral collapse causes loss of height, and existing deformities such as mild kyphosis or knock knees – themselves perhaps due to childhood rickets – may increase in later life. Unexplained pain in the hip or one of the long bones may presage a stress fracture.

X-rays

Children In active rickets there is thickening and widening of the growth plate, cupping of the metaphysis and, sometimes, bowing of the diaphysis. The metaphysis may remain abnormally wide even after healing has occurred. If the serum calcium remains persistently low there may be signs of secondary hyperparathyroidism: sub-periosteal erosions are at the sites of maximal remodelling (medial borders of the proximal humerus, femoral neck, distal femur and proximal tibia, lateral borders of the distal radius and ulna).

Adults The classical lesion of osteomalacia is the Looser zone, a thin transverse band of rarefaction in an otherwise normal-looking bone. These zones, seen especially in the shafts of long bones and theaxillary edge of the scapula, are due to incomplete stress fractures which heal with callus lacking in calcium. More often, however, there is simply a slow fading of skeletal structure, resulting in biconcave vertebrae (from disc pressure), lateral indentation of the acetabula ('trefoil' pelvis) and spontaneous fractures of the ribs, pubic rami, femoral neck or the metaphyses above and below the knee. Features of secondary hyperparathyroidism characteristically appear in the middle phalanges of the fingers, and in severe cases so-called 'brown tumours' are seen in the long bones.

Biochemistry

Changes common to almost all types of vitamin D related rickets and osteomalacia are diminished levels of serum calcium and phosphate, increased alkaline phosphatase and diminished urinary excretion of calcium. In vitamin D deficiency 25-OH D levels also are low. The 'calcium phosphate product' (derived by multiplying calcium and phosphorus levels expressed in mmol/L), normally about 3, is diminished in rickets and osteomalacia, and values of less than 2.4 are diagnostic.

Bone biopsy

With clearcut clinical and x-ray features the diagnosis is obvious. In less typical cases a bone biopsy will provide the answer. Osteoid seams are both wider and more extensive, and tetracycline labelling shows that mineralization is defective.

Treatment

Dietary lack of vitamin D (less than 100 IU per day) is common in strict vegetarians, in old people who often eat very little and even in entire populations whose traditional foods contain very little vitamin D. If there is also reduced exposure to sunlight, rickets or osteomalacia may result. The use of sun-blocking lotions, or overall cover by clothing, may seriously reduce exposure to ultraviolet light. Some of these problems can be corrected by simple social adjustments. Treatment with vitamin D (400–1000 IU per day) and calcium supplements is usually effective; however, elderly people often require larger doses of vitamin D (up to 2000 IU per day). Intestinal malabsorption – especially fat malabsorption – can cause vitamin D deficiency (fat and vitamin D absorption go hand in hand). If vitamin D supplements are administered they have to be given in large doses (50 000 IU per day). Surgery Established long-bone deformities may need bracing or operative correction once the metabolic disorder has been treated.

VITAMIN D RESISTANT RICKETS AND OSTEOMALACIA

There are several types of rickets and osteomalacia that do not respond to physiological doses of vitamin D. Although some are uncommon, they should be borne in mind in dealing with resistant cases. Inadequacy of hepatic 25-OHD Defective conversion to (or too-rapid breakdown of) 25-OHD in the liver may result from long-term administration of anticonvulsants or rifampicin, and if these drugs are prescribed it is wise to give adequate amounts of vitamin D at the same time. Occasionally the condition is also seen in severe liver failure. Treatment in these cases requires vitamin D in very large doses. Abnormalities of 1,25-(OH)2D metabolism

Renal failure Patients with early renal failure sometimes develop osteomalacia; this is thought to be due to reduced 1α-hydroxylase activity resulting in deficiency of 1,25-(OH)2D. The condition can be treated with 1,25-(OH)2D (or else with very large doses of vitamin D). Patients with advanced renal disease treated by haemodialysis develop a more complex syndrome – renal osteodystrophy. This is considered on page 141.

Vitamin D dependent rickets and osteomalacia Rare causes of 1,25-(OH)2D failure are two heritable (autosomal recessive) disorders.

Type I (pseudo vitamin D deficient rickets) is due to deficiency of 1α-hydroxylase; children develop very severe rickets and secondary hyperparathyroidism causing multiple fractures and generalized myopathy, as well as dental enamel hypoplasia. They need lifelong treatment with 1-(OH) D.

Type II vitamin D dependent rickets and osteomalacia is resistant to treatment with both vitamin D and calcitriol (1,25-(OH)2D). Plasma 1,25-(OH)2D levels are elevated but vitamin D receptors at the target organs (intestine and bone) are defective. Bone changes usually appear during childhood but adults also are affected. There is hypocalcaemia and secondary hyperparathyroidism. Neither vitamin D nor any of its metabolites is curative and patients may need long-term parenteral calcium.

NB: Patients treated with supra-physiological doses of calcitriol run the risk of developing hypercalcaemia, hypercalciuria and nephrocalcinosis; plasma calcium concentration should be measured regularly and ideally treatment should be conducted under the supervision of a specialist in this field.

PAGET'S DISEASE (OSTEITIS DEFORMANS)

Paget's disease is characterized by increased bone turnover and enlargement and thickening of the bone, but the internal architecture is abnormal and the bone is unusually brittle. The condition has a curious ethnic and geographical distribution, being relatively common (a prevalence of more than 3 per cent in people aged over 40) in North America, Britain, western Europe and Australia but rare in Asia, Africa and the Middle East. There is a tendency to familial aggregation. The cause is unknown, although the discovery of inclusion bodies in the osteoclasts has suggested a viral infection (Rebel et al., 1980).

HYPERVITAMINOSIS

Hypervitaminosis A occurs in children following excessive dosage; in adults it seldom occurs except in explorers who eat polar bear livers. There may be bone pain, and headache and vomiting due to raised intracranial pressure. X-ray shows increased density in the metaphyseal region and sub-periosteal calcification.

Hypervitaminosis D occurs if too much vitamin D is given. It exerts a PTH-like effect and so, as in the underlying rickets, calcium is withdrawn from bones; but metastatic calcification occurs. In treatment the dose of vitamin D must be properly regulated and the infant given a low-calcium diet but plentiful fluids.

SCURVY

Vitamin C (ascorbic acid) deficiency causes failure of collagen synthesis and osteoid formation. The result is osteoporosis, which in infants is most marked in the juxta-epiphyseal bone. Spontaneous bleeding is common. The infant is irritable and anaemic. The gums may be spongy and bleeding. Sub-periosteal haemorrhage causes excruciating pain and tenderness near the large joints. Fractures or epiphyseal separations may occur. X-rays show generalized bone rarefaction, most marked in the long-bone metaphyses. The normal calcification in growing cartilage produces dense transverse bands at the juxta-epiphyseal zones and around the ossific centres of the epiphyses (the 'ring sign'). The metaphyses may be deformed or fractured. Sub-periosteal haematomas show as soft-tissue swellings or peri-osseous calcification.

Treatment is with large doses of vitamin C.

ENDOCRINE DISORDERS

The endocrine system plays an important part in skeletal growth and maturation, as well as the maintenance of bone turnover. The anterior lobe of the pituitary gland directly affects growth; it also controls the activities of the thyroid, the gonads and the adrenal cortex, each of which has its own influence on bone; and the pituitary itself is subject to feedback stimuli from the other glands. The various mechanisms are, in fact, part of an interactive system in which balance is more important than individual activity. For example: pituitary growth hormone stimulates cell proliferation and growth at the physes. Gonadal hormone promotes growth plate maturation and fusion. While pituitary activity is in the ascendant, the bones elongate; after sexual maturation, the rise in gonadal hormone activity simultaneously 'feeds

back' on the pituitary and also directly closes down further physeal growth. When the system goes out of balance abnormalities occur. They are often complex, with several levels of dysfunction, due to (a) the local effects of the lesion which upsets the endocrine gland (e.g. pressure on cranial nerves from a pituitary adenoma); (b) oversecretion or undersecretion by the gland affected; and (c) over- or under-activity of other glands that are dependent on the primary dysfunctional gland. The descriptions which follow have been somewhat simplified.

Genetic disorders, skeletal dysplasias and malformations 8

There can be few diseases in which genetic factors do not play a role – if only in creating a background favourable to the operation of some more proximate pathogen. Sometimes, however, a genetic defect is the major – or the only – determinant of an abnormality that is either present at birth (e.g. achondroplasia) orevolves over time (e.g. Huntington's chorea). Such conditions can be broadly divided into three categories: chromosome disorders, single gene disorders and polygenic or multifactorial disorders. Various anomalies may also result from injury to the formed embryo. Many of these conditions affect the musculoskeletal system, producing cartilage and bone dysplasia (abnormal bone growth and/or modelling), malformations (e.g. absence or duplication of certain parts) or structural defects of connective tissue. In some a specific metabolic abnormality has been identified.
Genetic influences also contribute to the development of many acquired disorders. Osteoporosis, for example, is the result of a multiplicity of endocrine, dietary and environmental factors, yet twin studies have shown a significantly closer concordance in bone mass between identical twins than between non-identical twins. Before considering the vast range of developmental disorders, it may be helpful to review certain general aspects of genetic abnormalities.

DIAGNOSIS IN CHILDHOOD

Clinical features

Tell-tale features suggesting skeletal dysplasia are:
- retarded growth and shortness of stature
- disproportionate length of trunk and limbs
- localized malformations (dysmorphism)
- soft-tissue contractures
- childhood deformity.

All the skeletal dysplasias affect growth, although this may not be obvious at birth. Children should be measured at regular intervals and a record kept of height, length of lower segment (top of pubic symphysis to heel), upper segment (pubis to cranium), span, head circumference and chest circumference. Failure to reach the expected height for the local population group should be noted, and marked shortness of stature is highly suspicious. Bodily proportion is as important as overall height. The normal upper segment:lower segment ratio changes gradually from about 1.5:1 at the end of the first year to about 1:1 at puberty. Shortness of stature with normal proportions is not necessarily abnormal, but it is also seen in endocrine disorders

which affect the different parts of the skeleton more or less equally (e.g. hypopituitarism). By contrast, small stature with disproportionate shortness of the limbs is characteristic of skeletal dysplasia, the long bones being more markedly affected than the axial skeleton. The different segments of the limbs also may be disproportionately affected. The subtleties of dysplastic growth are reflected in terms such as rhizomelia – unusually short proximal segments (humeri and femora), mesomelia – short middle segments (forearms and legs) and acromelia – stubby hands and feet. Dysmorphism (a misshapen part of the body) is most obvious in the face and hands. There is a remarkable consistency about these changes, which makes for a disturbing similarity of appearance in members of a particular group.
Local deformities – such as kyphosis, valgus or varus knees, bowed forearms and ulnar deviated wrists – result from disturbed bone growth.

X-rays

The presence of any of the above features calls for a limited radiographic survey: a posteroanterior view of the chest, anteroposterior views of the pelvis, knees and hands, additional views of one arm and one leg, a lateral view of the thoracolumbar spine and standard views of the skull. Fractures, bent bones, exostoses, epiphyseal dysplasia and spinal deformities may be obvious, especially in the older child. Sometimes a complete survey is needed and it is important to note which portion of the long bones (epiphysis, metaphysis or diaphysis) is affected. With severe and varied changes in the metaphyses, periosteal new bone formation or epiphyseal separation, always consider the possibility of non-accidental injuries – the 'battered baby' syndrome.

Special investigations

In many cases the diagnosis can be made without laboratory tests; however, routine blood and urine analysis may be helpful in excluding metabolic and endocrine disorders such as rickets and pituitary or thyroid dysfunction. Special tests are also available to identify specific excretory metabolites in the storage disorders, and specific enzyme activity can be measured in serum, blood cells or cultured fibroblasts. Bone biopsy is occasionally helpful in disorders of bone density. Direct testing for gene mutations is already available for a number of conditions and is rapidly being extended to others. It is a useful adjunct to clinical diagnosis. Still somewhat controversial is its application to pre-clinical diagnosis of late-onset disorders and neonatal screening for potentially dangerous conditions such as sickle-cell disease.

DIAGNOSIS IN ADULTHOOD

It is unusual for a patient to present in adulthood with a condition that has been present since birth but in milder cases the abnormality may not have been recognized, particularly when several members of the family are similarly affected. In the worst of the genetic disorders the fetus is still-born or survives for only a short time. Individuals who reach adulthood, though recognizably abnormal, may lead active lives, marry and have children of their own. Nevertheless, they often seek medical advice for several reasons:

- short stature – especially disproportionate shortness of the lower limbs
- local bone deformities or exostoses
- spinal stenosis
- repeated fractures
- secondary osteoarthritis (e.g. due to epiphyseal dysplasia)
- joint laxity or instability.

The clinical approach is similar to that employed with children.

Table 8.1 A practical grouping of generalized developmental disorders

1 Disorders of cartilage and bone growth

1.1 Dysplasias with predominantly physeal and metaphyseal changes

1.1.1 Hereditary multiple exostosis
1.1.2 Achondroplasia
1.1.3 Hypochondroplasia
1.1.4 Metaphyseal chondrodysplasia
1.1.5 Dyschondroplasia (enchodromatosis, Ollier's disease)

1.2 Dysplasias with predominantly epiphyseal changes

1.2.1 Multiple epiphyseal dysplasia
1.2.2 Spondyloepiphyseal dysplasia
1.2.3 Dysplasia epiphysealis hemimlica (Trevor's disease)
1.2.4 Chondrodysplasia punctata (stippled epiphysis)

1.3 Dysplasias with predominantly metaphyseal and diaphyseal changes

1.3.1 Metaphyseal dysplasia (Pyle's disease)
1.3.2 Craniometaphyseal dysplasia
1.3.3 Diaphyseal dysplasia (Engelmann's disease, Cumurati's disease)
1.3.4 Craniodiaphyseal dysplasia
1.3.5 Osteopetrosis (marble bones, Albers–Shönbert disease)
1.3.6 Pyknodysostosis
1.3.7 Candle bones, spotted bones and striped bones

1.4 Combined and mixed dysplasias

1.4.1 Spondylometaphyseal
1.4.2 Pseudoachondroplasia
1.4.3 Diastrophic dysplasia
1.4.4 Cleidocranial dysplasia
1.4.5 Nail–patella syndrome
1.4.6 Craniofacial dysplasia

2 Connective tissue disorders
2.1 Generalized joint laxity
2.2 Ehlers–Danlos syndrome
2.3 Larsen's syndrome
2.4 Osteogenesis imperfecta (brittle bones)
2.4.1 Mild
2.4.2 Lethal
2.4.3 Severe
2.4.4 Moderate
2.5 Fibrodysplasia ossificans progressive

3 Storage disorders and other metabolic defects
3.1 Mucopolysaccharidoses
3.1.1 Hurler's syndrome (MPS I)
3.1.2 Hunter's syndrome (MPS II)
3.1.3 Morquio–Brailsford syndrome (MPS IV)
3.2 Gaucher's disease
3.3 Homocystinuria
3.4 Alkaptonuria
3.5 Congenital hyperuricaemia
4 Chromosome disorders
4.1 Down's syndrome
4.2 Thoracospinal anomalies
4.3 Elevation of the scapula (Sprengel's deformity)
4.4 Limb anomalies

HEREDITARY MULTIPLE EXOSTOSIS
(DIAPHYSEAL ACLASIS)
Multiple exostosis is the most common, and least disfiguring, of the skeletal dysplasias.
Clinical Features The condition is usually discovered in childhood; hard lumps appear at the ends of the long bones and along the apophyseal borders of the scapula and pelvis. As the child grows, these lumps enlarge and some may become hugely visible, especially around the knee. The more severely affected bones are abnormally short; this is seldom very marked but on measurement the lower body segment is shorter than the upper and span is less than height (Solomon, 1963). In the forearm and leg, the thinner of the two bones (the ulna or fibula) is usually the more defective, resulting in typical deformities: ulnar deviation of the wrist, bowing of the radius, subluxation of the radial head, valgus knees and valgus ankles. Bony lumps may cause pressure on nerves or vessels. Occasionally one of the cartilagecapped exostoses goes on growing into adult life and transforms to a chondrosarcoma; this is said to occur in 1–2 per cent of patients.

EHLERS–DANLOS SYNDROME
This syndrome comprises a collection of 6 major but heterogenous subtypes with a common phenotype of unusual skin elasticity, joint hypermobility and vascular fragility, expressions of underlying abnormalities of elastin and collagen formation. Sub-grouping is based on clinical findings, genetic cause and inheritance pattern. Of the many types of EDS so far described over 90 per cent show autosomal dominant inheritance.
Clinical Features
Babies may show marked hypotonia and joint laxity. Hypermobility persists and older patients are often capable of bizarre feats of contortion. The skin is soft and hyperextensible;

it is easily damaged and vascular fragility may give rise to ‘spontaneous’ bruising. Joint laxity, recurrent dislocations and scoliosis are common.

Management

Complications (e.g. recurrent dislocation or scoliosis) may need treatment. However, if joint laxity is marked, soft-tissue reconstruction usually fails to cure the tendency to dislocation. Beware! Blood vessel fragility may cause severe bleeding at operation or afterwards. Wound healing is often poor, leaving ‘cigarette paper’ scars. Joint instability may lead to osteoarthritis in later life.

OSTEOGENESIS IMPERFECTA (BRITTLE BONES)

Osteogenesis imperfecta (OI) is one of the commonest of the genetic disorders of bone, with an estimated incidence of 1 in 20 000. Abnormal synthesis and structural defects of type I collagen result in abnormalities of the bones, teeth, ligaments, sclerae and skin. The defining clinical features are (1) osteopenia, (2) liability to fracture, (3) laxity of ligaments, (4) blue coloration of the sclerae and (5) dentinogenesis imperfecta (‘crumbling teeth’). However, there are considerable variations in the severity of expression of these features and in the pattern of inheritance and it is now recognized that the condition embraces a heterogeneous group of collagen abnormalities resulting from many different genetic mutational defects (Kocher and Shapiro, 1998).

Clinical features

The clinical features vary considerably, according to the severity of the condition. The most striking abnormality is the propensity to fracture, generally after minor trauma and often without much pain or swelling. In the classic case fractures are discovered during infancy and they recur frequently throughout childhood. Callus formation is florid, so much so that the lump has occasionally been mistaken for an osteosarcoma; however, the new bone is also abnormal and it remains ‘pliable’ for a long time, thus predisposing to malunion and an increased risk of further fracture. By the age of 6 years there may be severe deformities of the long bones, and vertebral compression fractures often lead to kyphoscoliosis. After puberty fractures occur less frequently. The skin is thin and somewhat loose and the joints are hypermobile. Blue or grey sclerae, when they occur, are due to uveal pigment showing through the hypertranslucent cornea. The teeth may be discoloured and carious. In milder cases fractures develop a year or two after birth – perhaps when the child starts to walk; they are also less frequent and deformity is not a marked feature. In the most severe types of OI, fractures are present before birth and the infant is either stillborn or lives only for a few weeks, death being due to respiratory failure, basilar indentation or intracranial haemorrhage following injury.

Diagnosis

In most cases the clinical and radiological features are so distinctive that the diagnosis is not in doubt. However, mistakes have been made and rare disorders causing multiple fractures may have to be excluded by laboratory tests. In hypophosphatasia, for example, the serum alkaline phosphatase level is very low. In older children with atypical features it is essential to look for evidence of physical abuse.

Management

There is no medical treatment which will counteract the effects of this abnormality, and genetic manipulation is no more than a promise for the future. Conservative treatment is

directed at preventing fractures – if necessary by using lightweight orthoses during physical activity – and treating fractures when they occur. However, splintage should not be overdone as this may contribute further to the prevailing osteopenia. General measures to prevent recurrent trauma, maintain movement and encourage social adaptation are very important. Children with severe OI may be treated medically with cyclical bisphosphonates to increase bone mineral density and reduce the tendency to fracture. Most of the long-term orthopaedic problems are encountered in types III and IV. Fractures are treated conservatively, but immobilization must be kept to a minimum. Long-bone deformities are common, due either to malunion of complete fractures or breaking of recurrent incomplete fractures; these may require operative correction, usually by 4 or 5 years of age. Multiple osteotomies are performed and the bone fragments are then realigned on a straight intra medullary rod; the same effect can be achieved by closed osteoclasis. The problem of the bone outgrowing the rod has been addressed by using telescoping nails; however, these carry a fairly high complication rate. Spinal deformity is also common and is particularly difficult to treat. Bracing is ineffectual and progressive curves require operative instrumentation and spinal fusion. After adolescence, fractures are much less common and patients may pursue a reasonably comfortable and useful life.

NEUROFIBROMATOSIS

Neurofibromatosis is one of the commonest single gene disorders affecting the skeleton. Two types are recognized:

Type 1 (NF-1) – also known as **von Recklinghausen's disease** – has an incidence of about 1 in 3500 live births. The abnormality is located in the gene which codes for neurofibromin, on chromososme 17. It is transmitted as autosomal dominant, with almost 100 per cent penetrance, but more than 50 per cent of cases are due to new mutation. The most characteristic lesions are neurofibromata (Schwann cell tumours) and patches of skin pigmentation (café au lait spots), but other features are remarkably protean and musculoskeletal abnormalities are seen in almost half of those affected.

Type 2 (NF-2) is much less common, with an incidence of 1 in 50 000 births. It is associated with the gene which codes for schwannomin, located on chromosome 22. Like NF-1, it is transmitted as autosomal dominant. Unlike NF-1, intracranial lesions (e.g. acoustic neuromas and meningiomas) are usual while musculoskeletal manifestations are rare.

ELEVATION OF THE SCAPULA (SPRENGEL'S DEFORMITY)

Mild degrees of congenital elevation of the scapula are common. In the full-blown Sprengel deformity the child has obvious asymmetry of the shoulders, with elevation and underdevelopment of the affected side. The scapula is abnormally small and too high. Sometimes the clavicle is affected as well. Shoulder movements may be restricted and on abduction or elevation the scapula moves very little or not at all. Occasionally both sides are involved. Sprengel's deformity may be associated with other defects of the cervical spine (e.g. Klippel–Feil syndrome), and high thoracic kyphosis or scoliosis is quite common. This condition, which usually occurs sporadically, represents a failure of scapular descent from the cervical spine. The high scapula may still be attached to the spine by a tough fibrous band or a cartilaginous bar (the omovertebral bar). Associated vertebral or rib anomalies are quite common.

Treatment is required only if shoulder movements are severely limited or if the deformity is particularly unsightly. Operation is best performed before the age of 6 years. The vertebroscapular muscles are released from the spine, the supraspinous part of the scapula is

excised together with the omovertebral bar and the scapula is repositioned by tightening the lower muscles. Great care is needed as there is a risk of injury to the accessory nerve or the brachial plexus.

Tumours 9

Tumours, tumour-like lesions and cysts are considered together, partly because their clinical presentation and management are similar and partly because the definitive classification of bone tumours is still evolving and some disorders may yet move from one category to another. Benign lesions are quite common, primary malignant ones rare; yet so often do they mimic each other, and so critical are the decisions on treatment, that a working knowledge of all the important conditions is necessary.

CLASSIFICATION

Most classifications of bone tumours are based on the recognition of the dominant tissue in the various lesions (Table 9.1). Knowing the cell line from which the tumour has sprung may help with both diagnosis and planning of treatment. There are, however, pitfalls in this approach:

- the most pervasive tissue is not necessarily the tissue of origin
- there is not necessarily any connection between conditions in one category
- there is often no relationship between benign and malignant lesions with similar tissue elements (e.g. osteoma and osteosarcoma)
- the commonest malignant lesions in bone – metastatic tumours – are not, strictly speaking, 'bone' tumours, i.e. not of mesenchymal origin

Table 9.1 A classification of bone tumours. Modified after Revised WHO Classification – Schajowicz (1994)

Predominant tissue	Benign	Malignant
Bone forming	Osteoma	Osteosarcoma:
	Osteoid osteoma	central
	Osteoblastoma	peripheral
		parosteal
Cartilage forming	Chondroma	Chondrosarcoma:
	Osteochondroma	central
	Chondroblastoma	peripheral
	?Chondromyxoid fibroma	juxtacortical
		clear-cell
		mesenchymal
Fibrous tissue	Fibroma	Fibrosarcoma
	Fibromatosis	
Mixed	?Chondromyxoid fibroma	
Giant-cell tumours	Benign osteoclastoma	Malignant osteoclastoma
Marrow tumours		Ewing's tumour
		Myeloma
Vascular tissue	Haemangioma	Angiosarcoma
	Haemangiopericytoma	Malignant haemangiopericytoma
	Haemangioendothelioma	
Other connective tissue	Fibroma	Fibrosarcoma
	Fibrous histiocytoma	Malignant fibrous histiocytoma
	Lipoma	Liposarcoma
Other tumours	Neurofibroma	Adamantinoma
	Neurilemmoma	Chordoma

CLINICAL PRESENTATION

HISTORY

The history is often prolonged, and this unfortunately results in a delay in obtaining treatment. Patients may be completely asymptomatic until the abnormality is discovered on x-ray. This is more likely with benign lesions; and, since some of these (e.g. non-ossifying fibroma) are common in children but rare after the age of 30, they must be capable of spontaneous resolution. Malignant tumours, too, may remain silent if they are slow-growing and situated where there is room for inconspicuous expansion (e.g. the cavity of the pelvis). Age may be a useful clue. Many benign lesions present during childhood and adolescence – but so do some primary malignant tumours, notably Ewing's tumour and osteosarcoma. Chondrosarcoma and fibrosarcoma typically occur in older people (fourth or sixth decades); and myeloma, the commonest of all primary malignant bone tumours, is seldom seen before the sixth decade. In patients over 70 years of age, metastatic bone lesions are more common than all primary tumours together. Pain is a common complaint and gives little indication of the nature of the lesion; however, progressive and unremitting pain is a sinister symptom. It may be caused by rapid expansion with stretching of surrounding tissues, central haemorrhage or degeneration

in the tumour, or an incipient pathological fracture. However, even a tiny lesion may be very painful if it is encapsulated in dense bone (e.g. an osteoid osteoma). Swelling, or the appearance of a lump, may be alarming. Often, though, patients seek advice only when a mass becomes painful or continues to grow. A history of trauma is offered so frequently that it cannot be dismissed as having no significance. Yet, whether the injury initiates a pathological change or merely draws attention to what is already there remains unanswered. Neurological symptoms (paraesthesiae or numbness) may be caused by pressure upon or stretching of a peripheral nerve. Progressive dysfunction is more ominous and suggests invasion by an aggressive tumour. Pathological fracture may be the first (and only) clinical signal. Suspicion is aroused if the injury was slight; in elderly people, whose bones usually fracture at the cortico-cancellous junctions, any break in the mid-shaft should be regarded as pathological until proved otherwise.

EXAMINATION

If there is a lump, where does it arise? Is it discrete or ill-defined? Is it soft or hard, or pulsatile? And is it tender? Swelling is sometimes diffuse, and the overlying skin warm and inflamed; it can be difficult to distinguish a tumour from infection or a haematoma. If the tumour is near a joint there may be an effusion and/or limitation of movement. Spinal lesions, whether benign or malignant, often cause muscle spasm and back stiffness, or a painful scoliosis. The examination will focus on the symptomatic part, but it should include the area of lymphatic drainage and, often, the pelvis, abdomen, chest and spine.

IMAGING

X-RAYS

Plain x-rays are still the most useful of all imaging techniques. There may be an obvious abnormality in the bone – cortical thickening, a discrete lump, a ‘cyst’ or ill-defined destruction. Where is the lesion: in the metaphysis or the diaphysis? Is it solitary or are there multiple lesions? Are the margins well-defined or ill-defined? Remember that ‘cystic’ lesions are not necessarily hollow cavities: any radiolucent material (e.g. a fibroma or a chondroma) may look like a cyst. If the boundary of the ‘cyst’ is sharply defined it is probably benign; if it is hazy and diffuse it suggests an invasive tumour. Stippled calcification inside a cystic area is characteristic of cartilage tumours. Look carefully at the bone surfaces: periosteal newbone formation and extension of the tumour into the soft tissues are suggestive of malignant change. Look also at the soft tissues: Are the muscle planes distorted by swelling? Is there calcification? For all its informative detail, the x-ray alone can seldom be relied on for a definitive diagnosis. With some notable exceptions, in which the appearances are pathognomonic (osteochondroma, non-ossifying fibroma, osteoid osteoma), further investigations will be needed. If other forms of imaging are planned (bone scans, CT or MRI), they should be done before undertaking a biopsy, which itself may distort the appearances.

RADIONUCLIDE SCANNING

Scanning with 99mTc-methyl diphosphonate (99mTc- MDP) shows non-specific reactive changes in bone; this can be helpful in revealing the site of a small tumour (e.g. an osteoid osteoma) that does not show up clearly on x-ray. Skeletal scintigraphy is also useful for detecting skip lesions or ‘silent’ secondary deposits.

COMPUTED TOMOGRAPHY

CT extends the range of x-ray diagnosis; it shows more accurately both intraosseous and extraosseous extension of the tumour and the relationship to surrounding structures. It may also reveal suspected lesions in inaccessible sites, like the spine or pelvis; and it is a reliable method of detecting pulmonary metastases.

MAGNETIC RESONANCE IMAGING

MRI provides further information. Its greatest value is in the assessment of tumour spread: (a) within the bone, (b) into a nearby joint and (c) into the soft tissues. Blood vessels and the relationship of the tumour to the perivascular space are well defined. MRI is also useful in assessing soft-tissue tumours and cartilaginous lesions.

LABORATORY INVESTIGATIONS

Blood tests are often necessary to exclude other conditions, e.g. infection or metabolic bone disorders, or a 'brown tumour' in hyperparathyroidism. Anaemia, increased ESR and elevated serum alkaline phosphatase levels are non-specific findings, but if other causes are excluded they may help in differentiating between benign and malignant bone lesions. Serum protein electrophoresis may reveal an abnormal globulin fraction and the urine may contain Bence Jones protein in patients with myeloma. A raised serum acid phosphatase suggests prostatic carcinoma.

BIOPSY

Needle biopsy Needle biopsy should be performed either by the surgeon planning definitive treatment or by an experienced radiologist. Often it is carried out with the help of ultrasound or CT guidance (Stoker et al., 1991; Saifuddin et al., 2000). A large bore biopsy needle, such as a Jamshidi or a Trucut needle, is used. It is important to ensure that a representative sample of the tumour is taken and that it is adequate to make a histological diagnosis; a frozen section can be used in order to confirm this. If infection is suspected then a sample should be sent for microbiology. It is also essential that the biopsy is carried out in the line of any further surgical incision so that the tract can be excised at the time of definitive surgery.

Open biopsy This is a more reliable way of obtaining a representative sample, however it is associated with significant morbidity (Mankin et al., 1982). It is often performed if a needle biopsy would place the neurovascular structures at risk or if a diagnosis has not been made after needle biopsy. The site is selected so that it can be included in any subsequent operation. As little as possible of the tumour is exposed and a block of tissue is removed – ideally in the boundary zone, so as to include normal tissue, pseudocapsule and abnormal tissue. If bone is removed the raw area is covered with bone wax or methylmethacrylate cement. If a tourniquet is used, it should be released and full haemostasis achieved before closing the wound. Drains should be avoided, so as to minimize the risk of tumour contamination. An experienced histopathologist should be on hand and the specimens should be delivered fresh, unfixed and uncrushed. For tumours that are almost certainly benign, an excisional biopsy is permissible (the entire lesion is removed); with cysts that need operations, representative tissue can be obtained by careful curettage. In either case, histological confirmation of the diagnosis is essential. Biopsy should never be regarded as a 'minor' procedure. Complications include haemorrhage, wound breakdown, infection and pathological fracture (Mankin at al., 1982, 1996; Springfield and Rosenberg, 1996). The person doing the biopsy should have a clear idea of what may be done next and where operative incisions or skin flaps will be placed. Errors and complications are far less likely if the procedure is performed in a specializing centre. A last word of warning: When dealing with tumours that could be malignant, there is a strong temptation to perform the biopsy as soon as possible; as this may alter the CT and MRI appearances, it is important to delay the procedure until all the imaging studies have been completed.

DIFFERENTIAL DIAGNOSIS

A number of conditions may mimic a tumour, either clinically or radiologically, and the histopathology may be difficult to interpret. It is important not to be misled by the common dissemblers.

Soft-tissue haematoma A large, clotted sub-periosteal or soft-tissue haematoma may present as a painful lump in the arm or lower limb. Sometimes the x-ray shows an irregular surface on the underlying bone. Important clues are the history and the rapid onset of symptoms.
Myositis ossificans Although rare, this may be a source of confusion. Following an injury the patient develops a tender swelling in the vicinity of a joint; the x-ray shows fluffy density in the soft tissue adjacent to bone. Unlike a malignant tumour, however, the condition soon becomes less painful and the new bone better defined and well demarcated.
Stress fracture Some of the worst mistakes have been made in misdiagnosing a stress fracture. The patient is often a young adult with localized pain near a large joint; x-rays show a dubious area of cortical 'destruction' and overlying periosteal new bone; if a biopsy is performed the healing callus may show histological features resembling those of osteosarcoma. If the pitfall is recognized, and there is adequate consultation between surgeon, radiologist and pathologist, a serious error can be prevented.
Tendon avulsion injuries Children and adolescents – especially those engaged in vigorous sports – are prone to avulsion injuries at sites of tendon insertion, particularly around the hip and knee (Donnelly et al., 1999). The best known example is the tibial apophyseal stress lesion of Osgood–Schlatter's disease (see page 565), but lesions at less familiar sites (the iliac crest, the ischial tuberosity, the lesser trochanter of the femur, the hamstring insertions, the attachments of adductor magnus and longus and the distal humeral apophyses) may escape immediate recognition.
Bone infection Osteomyelitis typically causes pain and swelling near one of the larger joints; as with primary bone tumours, the patients are usually children or young adults. X-rays may show an area of destruction in the metaphysis, with periosteal new bone. Systemic features, especially if the patient has been treated with antibiotics, may be mild. If the area is explored, tissue should be submitted for both bacteriological and histological examination.
Gout Occasionally a large gouty tophus causes a painful swelling at one of the bone ends, and x-ray shows a large, poorly defined excavation. If it is kept in mind the diagnosis will be easily confirmed – if necessary by obtaining a biopsy from the lump.
Other bone lesions Non-neoplastic bone lesions such as fibrous cortical defects, medullary infarcts and 'bone islands' are occasionally mistaken for tumours.

STAGING OF BONE TUMOURS

In treating tumours we strive to reconcile two conflicting principles: the lesion must be removed widely enough to ensure that it does not recur, but damage must be kept to a minimum. The balance between these objectives depends on knowing (a) how the tumour usually behaves (i.e. how aggressive it is), and (b) how far it has spread. The answers to these two questions are embodied in the staging system developed by Enneking (1986).

Table 9.2 Staging of benign bone tumours as described by Enneking

Latent	Well-defined margin. Grows slowly and then stops Remains static/heals spontaneously E.g. Osteoid osteoma
Active	Progressive growth limited by natural barriers Not self-limiting. Tendency to recur E.g. Aneurysmal bone cyst
Aggressive	Growth not limited by natural barriers (e.g. giant cell tumour)

AGGRESSIVENESS

Tumours are graded not only on their cytological characteristics but also on their clinical behaviour, i.e. the likelihood of recurrence and spread after surgical removal.
Benign lesions, by definition, occupy the lowest grade, though even in this group there are important differences in behaviour calling for further subdivision into latent, active and aggressive lesions (Table 9.2). The least aggressive tumours may disappear spontaneously (e.g. non-osteogenic fibroma); the most aggressive are difficult to distinguish from a low-grade sarcoma and sometimes undergo malignant change (e.g. aggressive osteoblastoma). Most are amenable to local (marginal) excision with little risk of recurrence.
Malignant tumours are divided into 'low-grade' and 'high-grade': the former are only moderately aggressive and take a long time to metastasize (e.g. secondary chondrosarcoma or parosteal osteosarcoma), while the latter are usually very aggressive and metastasize early (e.g. osteosarcoma or fibrosarcoma).

SPREAD

Assuming that there are no metastases, the local extent of the tumour is the most important factor in deciding how much tissue has to be removed. Lesions that are confined to an enclosed tissue space (e.g. a bone, a joint cavity or a muscle group within its fascial envelope) are called 'intracompartmental'. Those that extend into interfascial or extrafascial planes with no natural barrier to proximal or distal spread (e.g. perivascular sheaths, pelvis, axilla) are designated 'extracompartmental'. The extent of the tumour and adjacent 'contaminated' tissue are best shown by CT and MRI; skip lesions can be detected by scintigraphy.

SURGICAL STAGE

'Staging' the tumour is an important step towards selecting the operation best suited to that particular patient, and carrying a low risk of recurrence. Locally recurrent sarcomas tend to be more aggressive, more often extracompartmental and more likely to metastasize than the original tumour.
Bone sarcomas are broadly divided as follows:
• Stage I All low-grade sarcomas.
• Stage II Histologically high-grade lesions.
• Stage III Sarcomas which have metastasized.
Following Enneking's original classification, each category is further subdivided into Type A (intracompartmental) and Type B (extracompartmental) (Fig. 9.3). Thus, a localized chondrosarcoma arising in a cartilage-capped exostosis would be designated IA, suitable for wide excision without exposing the tumour. An osteosarcoma confined to bone would be IIA – operable by wide excision or amputation with a low risk of local recurrence; if it has spread into the soft tissues it would be IIB – less suitable for wide excision and preferably treated by radical resection or disarticulation through the proximal joint. If there are pulmonary metastases it would be classified as stage III.

Table 9.3 Surgical stages as described by Enneking

Stage	Grade	Site	Metastases
IA	Low	Intracompartmental	No
IB	Low	Extracompartmental	No
IIA	High	Intracompartmental	No
IIB	High	Extracompartmental	No
IIIA	Low	Intra- or extracompartmental	Yes
IIIA	High	Intra- or extracompartmental	Yes

STAGING OF SOFT-TISSUE TUMOURS

Soft-tissue tumours are staged using the American Joint Committee for Cancer Staging System, according to their histological grade (G), size (T), lymph node involvement (N) and whether they have metastasized (M) (Russell et al., 1977). The main differences between this and the Enneking system are the increased number of histological grades (from low and high to 1, 2 and 3) and use of the size of the tumour (less than or greater than 5 cm), rather than whether it is intra- or extracompartmental.

PRINCIPLES OF MANAGEMENT

For all but the simplest and most obvious of benign tumours, management calls for a multidisciplinary team approach and is best conducted in a tertiary centre specializing in the treatment of bone and softtissue tumours. Consultation and cooperation between the orthopaedic surgeon, radiologist, patho -logist and (certainly in the case of malignant tumours) the oncologist is essential in the initial management. In many cases physiotherapists, occupational therapists and prosthetists will also be involved. Once clinical and radiological examination have suggested the most likely diagnosis, further management proceeds as follows. Benign, asymptomatic lesions If the diagnosis is beyond doubt (e.g. a non-ossifying fibroma or a small osteochondroma) one can afford to temporize; treatment may never be needed. However, if the appearances are not pathognomonic, a biopsy is advisable and this may take the form of excision or curettage of the lesion. Benign, symptomatic or enlarging tumours Painful lesions, or tumours that continue to enlarge after the end of normal bone growth, require biopsy and confirmation of the diagnosis. Unless they are unusually aggressive, they can generally be removed by local (marginal) excision or (in the case of benign cysts) by curettage.

Suspected malignant tumours If the lesion is thought to be a primary malignant tumour, the patient is admitted for more detailed examination, blood tests, chest x-ray, further imaging (including pulmonary CT) and biopsy. This should allow a firm diagnosis and staging to be established. The various treatment options can then be discussed with the patient (or the parents, in the case of a young child). A choice needs to be made between amputation, limb-sparing operations and different types of adjuvant therapy, and the patient must be fully informed about the pros and cons of each.

METHODS OF TREATMENT

TUMOUR EXCISION

The more aggressive the lesion the more widely does it need to be excised, in order to ensure that the tumour as well as any dubious marginal tissue is completely removed.

Intracapsular (intralesional) excision and curettage are incomplete forms of tumour ablation and therefore applicable only to benign lesions with a very low risk of recurrence, or to incurable tumours which need debulking to relieve local symptoms. Adjunctive treatment such as the use of acrylic cement after curettage decreases the risk of local recurrence.

Marginal excision goes beyond the tumour, but only just. If the dissection of a malignant lesion is carried through the reactive zone, there is a significant risk of recurrence (up to 50 per cent). For benign lesions, however, this is a suitable method; the resulting cavity can be filled with graft bone. Wide excision implies that the dissection is carried out well clear of the tumour, through normal tissue. This is appropriate for low-grade intracompartmental lesions (grade IA), providing a risk of local recurrence below 10 per cent. However, wide excision is also used in conjunction with chemotherapy for grade IIA lesions. Radical resection means that the entire compartment in which the tumour lies is removed en bloc without exposing the lesion. It may be possible to do this while still sparing the limb, but the surrounding muscles,

ligaments and connective tissues will have to be sacrificed; in some cases a true radical resection can be achieved only by amputating at a level above the compartment involved. This method is required for high-grade tumours (IIA or IIB).

LIMB SALVAGE

Amputation is no longer the automatic choice for grade II sarcomas. Improved methods of imaging and advances in chemotherapy have made limb salvage the treatment of choice for many patients. However, this option should be considered only if the local control of the tumour is likely to be as good as that obtained by amputation, if it is certain that there are no skip lesions and if a functional limb can be preserved. The ongoing debate around limb sparing versus amputation is addressed in an excellent paper by DiCaprio and Friedlaender (2003). Advanced surgical facilities for bone grafting and endoprosthetic replacement at various sites must be available. The first step consists of wide excision of the tumour with preservation of the neurovascular structures. The resulting defect is then dealt with in one of several ways. Short diaphyseal segments can be replaced by vascularized or non-vascularized bone grafts. Longer gaps may require custom-made implants. Osteo-articular segments can be replaced by large allografts, endoprostheses or allograft–prosthetic composites. It is recognized, however, that the use of large allografts carries a high risk of infection and fracture; this has led to them not being used as widely as in the past. Endoprostheses used to be custom-made but nowadays modular systems for tumour reconstruction are available.

In growing children, extendible implants have been used in order to avoid the need for repeated operations; however, they may need to be replaced at the end of growth. Other procedures, such as grafting and arthrodesis or distraction osteosynthesis, are suitable for some situations. Sarcomas around the hip and shoulder present special problems. Complete excision is difficult and reconstruction involves complex grafting and replacement procedures (O'Connor et al., 1996). Outcome Tumour replacement by massive endoprosthesis carries a high risk of complications such as wound breakdown and infection; the 10-year survival rate of these prostheses with mechanical failure as the end point is 75 per cent and for failure due to any cause is 58 per cent. The limb salvage rate at 20 years is 84 per cent (Jeys et al., 2008).

AMPUTATION

Considering the difficulties of limb-sparing surgery – particularly for high-grade tumours or if there is doubt about whether the lesion is intracompartmental – amputation and early rehabilitation may be the wisest option. Preoperative planning and the definitive operation are best carried out in a specialized unit, so as to minimize the risk of complications and permit early rehabilitation. Amputation may be curative but it is sometimes performed essentially to achieve local control of a tumour which is resistant to chemotherapy and radiation therapy.

MULTI-AGENT CHEMOTHERAPY

Multi-agent chemotherapy is now the preferred neoadjuvant and adjuvant treatment for malignant bone and soft-tissue tumours. There is good evidence to show that, for sensitive tumours, modern chemotherapy regimens effectively reduce the size of the primary lesion, prevent metastatic seeding and improve the chances of survival. When combined with surgery for osteosarcoma and Ewing's tumours, the long-term disease-free survival rate in the best series is now about 60 per cent. Drugs currently in use are methotrexate, doxorubicin (Adriamycin), cyclophosphamide, vincristine and cis-platinum. Treatment is started 8–12 weeks preoperatively and the effect is assessed by examining the resected tissue for tumour necrosis; greater than 90 per cent necrosis is taken as a good response. If there is little or no necrosis, a different drug may be selected for postoperative treatment. Maintenance chemotherapy is continued for another 6–12 months.

RADIOTHERAPY

High-energy irradiation has long been used to destroy radiosensitive tumours or as adjuvant therapy before operation. Nowadays the indications are more restricted. For highly sensitive tumours (such as Ewing's sarcoma) it offers an alternative to amputation; it is then combined with adjuvant chemotherapy. The same combination can be used as adjunctive treatment for high-grade tumours, for tumours in inaccessible sites, lesions that are inoperable because of their size, proximity to major blood vessels or advanced local spread, for marrow-cell tumours such as myeloma and malignant lymphoma, for metastatic deposits and for palliative local tumour control where no surgery is planned. Radiotherapy may also be employed postoperatively when a marginal or intralesional excision has occurred, so as to 'sterilize' the tumour bed. The main complications of this treatment are the occurrence of post-irradiation spindle-cell sarcoma and pathological fracture in weightbearing bones, particularly in the proximal half of the femur.

BENIGN BONE LESIONS

NON-OSSIFYING FIBROMA (FIBROUS CORTICAL DEFECT)

This, the commonest benign lesion of bone, is a developmental defect in which a nest of fibrous tissue appears within the bone and persists for some years before ossifying. It is asymptomatic and is almost always encountered in children as an incidental finding on x-ray. The commonest sites are the metaphyses of long bones; occasionally there are multiple lesions. The x-ray appearance is unmistakable. There is a more or less oval radiolucent area surrounded by a thin margin of dense bone; views in different planes may show that a lesion that appears to be 'central' is actually adjacent to or within the cortex, hence the alternative name 'fibrous cortical defect'.

Pathology Although it looks cystic on x-ray, it is a solid lesion consisting of unremarkable fibrous tissue with a few scattered giant cells. As the bone grows the defect becomes less obvious and it eventually heals spontaneously. However, it sometimes enlarges to several centimetres in diameter and there may be a pathological fracture. There is no risk of malignant change.

Treatment Treatment is usually unnecessary. If the defect is very large or has led to repeated fractures, it can be treated by curettage and bone grafting. Recurrence is rare.

FIBROUS DYSPLASIA

Fibrous dysplasia is a developmental disorder in which areas of trabecular bone are replaced by cellular fibrous tissue containing flecks of osteoid and woven bone. It may affect one bone (monostotic), one limb (monomelic) or many bones (polyostotic). If the lesions are large, the bone is considerably weakened and pathological fractures or progressive deformity may occur. The most common sites of occurrence are the proximal femur, tibia, humerus, ribs and cranio-facial bones. Small, single lesions are asymptomatic. Large, monostotic lesions may cause pain or may be discovered only when the patient develops a pathological fracture. Patients with polyostotic disease present in childhood or adolescence with pain, limp, bony enlargement, deformity or pathological fracture. Untreated, the characteristic deformities persist through adult life. Occasionally the bone disorder is associated with café-au-lait patches on the skin and (in girls) precocious sexual development (Albright's syndrome). X-rays show radiolucent 'cystic' areas in the metaphysis or shaft; because they contain fibrous tissue with diffuse spots of immature bone, the lucent patches typically have a slightly hazy or 'ground-glass' appearance. The weightbearing bones may be bent, and one of the classic features is the 'shepherd's crook' deformity of the proximal femur. Radioscintigraphy shows marked activity in the lesion.

Pathology At operation the lesional tissue has a coarse, gritty feel (due to the specks of immature bone). The histological picture is of loose, cellular fibrous tissue with widespread patches of woven bone and scattered giant cells. Both clinically and histologically the monostotic condition may resemble either a bone-forming tumour or hyperparathyroidism. However, detailed xray and laboratory studies will exclude these disorders. Malignant transformation to fibrosarcoma occurs in 0.5 per cent of patients with monostotic lesions and up to 5 per cent of patients with Albright's syndrome. Treatment Treatment depends on the extent of the defect and the presence or absence of deformities. Small lesions need no treatment. Those that are large and painful or threatening to fracture (or have fractured) can be curetted and grafted, but there is a strong tendency for the abnormality to recur. A mixture of cortical and cancellous bone grafts may provide added strength even if the lesion is not eradicated. For very large lesions, the grafts can be supplemented by methylmethacrylate cement. Deformities may need correction by suitably designed osteotomies. With large cysts, the bone often bleeds profusely at operation: forewarned is forearmed.

OSTEOID OSTEOMA

This tiny bone tumour (less than 1 cm in diameter) causes symptoms out of all proportion to its size. Patients are usually under 30 years of age and males predominate. Any bone except the skull may be affected, but over half the cases occur in the femur or tibia. The patient complains of persistent pain, sometimes well localized but sometimes referred over a wide area. Typically the pain is relieved by salicylates. If the diagnosis is delayed, other features appear: a limp or muscle wasting and weakness; spinal lesions may cause intense pain, muscle spasm and scoliosis. The important x-ray feature is a small radiolucent area, the so-called 'nidus'. Lesions in the diaphysis are surrounded by dense sclerosis and cortical thickening; this may be so marked that the nidus can be seen only in fine cut CT scans. Lesions in the metaphysis show less cortical thickening. Further away the bone may be osteoporotic. 99mTc-MDP scintigraphy reveals intense, localized activity. It is sometimes difficult to distinguish an osteoid osteoma from a small Brodie's abscess without biopsy. Ewing's sarcoma and chronic periostitis must also be excluded.

Pathology The excised lesion appears as a dark-brown or reddish 'nucleus' surrounded by dense bone; the central area consists of unorganized sheets of osteoid and bone cells. There is no risk of malignant transformation.

Treatment The only effective treatment is complete removal or destruction of the nidus. The lesion is carefully localized by x-ray and/or CT and then excised in a small block of bone or destroyed by Ctlocalized radio-ablation. The specimen should be xrayed immediately to confirm that it does contain the little tumour. If the excision is likely to weaken the host bone (especially in the vulnerable medial cortex of the femoral neck), prophylactic internal fixation may be needed.

OSTEOBLASTOMA (GIANT OSTEOID OSTEOMA)

This tumour is similar to an osteoid osteoma but it is larger (more than 1 cm in diameter), more cellular and sometimes more ominous in appearance. It is usually seen in young adults, more often in men than in women. It tends to occur in the spine and the flat bones; patients present with pain and local muscle spasm. X-ray shows a well-demarcated osteolytic lesion which may contain small flecks of ossification. There is surrounding sclerosis but this is not always easy to see, especially with lesions in the flat bones or the vertebral pedicle. A radioisotope scan will reveal the 'hot' area. Larger lesions may appear cystic, and sometimes a typical aneurysmal bone cyst appears to have arisen in an osteoblastoma.

Pathology When the tumour is exposed it has a somewhat fleshy appearance. Histologically it resembles an osteoid osteoma, but the cellularity is more striking. Occasionally the picture may suggest a low-grade osteosarcoma.
Treatment Treatment consists of excision and bone grafting. With lesions in the vertebral pedicle or the floor of the acetabulum, this is not always easy and removal may be incomplete; local recurrence is common and malignant transformation has been reported (McLeod et al., 1976).

OSTEOCHONDROMA (CARTILAGE-CAPPED EXOSTOSIS)

This, one of the commonest 'tumours' of bone, is a developmental lesion which starts as a small overgrowth of cartilage at the edge of the physeal plate and develops by endochondral ossification into a bony protuberance still covered by the cap of cartilage. Any bone that develops in cartilage may be involved; the commonest sites are the fast-growing ends of long bones and the crest of the ilium. In long bones, growth leaves the bump stranded further down the metaphysis. Here it may go on growing but at the end of the normal growth period for that bone it stops enlarging. Any further enlargement after the end of the growth period is suggestive of malignant transformation. The patient is usually a teenager or young adult when the lump is first discovered. Occasionally there is pain due to an overlying bursa or impingement on soft tissues, or, rarely, paraesthesia due to stretching of an adjacent nerve.
The x-ray appearance is pathognomonic. There is a well-defined exostosis emerging from the metaphysis, its base co-extensive with the parent bone. It looks smaller than it feels because the cartilage cap is usually invisible on x-ray; however, large lesions undergo cartilage degeneration and calcification and then the x-ray shows the bony exostosis surrounded by clouds of calcified material. Multiple lesions may develop as part of a heritable disorder – hereditary multiple exostosis – in which there are also features of abnormal bone growth resulting in characteristic deformities (see Chapter 8).
Pathology At operation the cartilage cap is seen surmounting a narrow base or pedicle of bone. The cap consists of simple hyaline cartilage; in a growing exostosis the deeper cartilage cells are arranged in columns, giving rise to the formation of endochondral new bone. Large lesions may have a 'cauliflower' appearance, with degeneration and calcification in the centre of the cartilage cap.
Complications The incidence of malignant transformation is difficult to assess because troublesome lesions are so often removed before they show histological features of malignancy. Figures usually quoted are 1 per cent for solitary lesions and 6 per cent for multiple.
Features suggestive of malignant change are: (1) enlargement of the cartilage cap in successive examinations; (2) a bulky cartilage cap (more than 1 cm in thickness); (3) irregularly scattered flecks of calcification within the cartilage cap; and (4) spread into the surrounding soft tissues. MRI may be needed to reveal these changes.
Treatment If the tumour causes symptoms it should be excised; if, in an adult, it has recently become bigger or painful then operation is urgent, for these features suggest malignancy. This is seen most often with pelvic exostoses – not because they are inherently different but because considerable enlargement may, for long periods, pass unnoticed. If there are suspicious features, further imaging and staging should be carried out before doing a biopsy. If the histology is that of 'benign' cartilage but the tumour is known for certain to be enlarging after the end of the growth period, it should be treated as a chondrosarcoma.

GIANT-CELL TUMOUR

Giant-cell tumour, which represents 5 per cent of all primary bone tumours, is a lesion of uncertain origin that appears in mature bone, most commonly in the distal femur, proximal tibia, proximal humerus and distal radius, though other bones also may be affected. It is hardly ever seen before closure of the nearby physis and characteristically it extends right up to the subarticular bone plate. Rarely, there are multiple lesions. The patient is usually a young adult who complains of pain at the end of a long bone; sometimes there is slight swelling. A history of trauma is not uncommon and pathological fracture occurs in 10–15 per cent of cases. On examination there may be a palpable mass with warmth of the overlying tissues. X-rays show a radiolucent area situated eccentrically at the end of a long bone and bounded by the subchondral bone plate. The endosteal margin may be quite obvious, but in aggressive lesions it is ill-defined. The centre sometimes has a soap-bubble appearance due to ridging of the surrounding bone. The cortex is thin and sometimes ballooned; aggressive lesions extend into the soft tissue. The appearance of a 'cystic' lesion in mature bone, extending right up to the subchondral plate, is so characteristic that the diagnosis is seldom in doubt. However, it is prudent to obtain estimations of blood calcium, phosphate and alkaline phosphatise concentrations so as exclude an unusual 'brown tumour' associated with hyperparathyroidism. Because of the tumour's potential for aggressive behaviour, detailed staging procedures are essential. CT scans and MRI will reveal the extent of the tumour, both within the bone and beyond. It is important to establish whether the articular surface has been breached.

Biopsy is essential. This can be done either as a frozen section before proceeding with operative treatment or (especially if a more extensive operation is contemplated) as a separate procedure.

Treatment Well-confined, slow-growing lesions with benign histology can safely be treated by thorough curettage and 'stripping' of the cavity with burrs and gouges, followed by swabbing with hydrogen peroxide or by the application of liquid nitrogen; the cavity is then packed with bone chips. More aggressive tumours, and recurrent lesions, should be treated by excision followed, if necessary, by bone grafting or prosthetic replacement. Tumours in awkward sites (e.g. the spine) may be difficult to eradicate; supplementary radiotherapy is sometimes recommended, but it carries a significant risk of causing malignant transformation.

PRIMARY MALIGNANT BONE TUMOURS

CHONDROSARCOMA

Chondrosarcoma is one of the commonest malignant tumours originating in bone. The highest incidence is in the fourth and fifth decades and men are affected more often than women. These tumours are slow-growing and are usually present for many months before being discovered. Patients may complain of a dull ache or a gradually enlarging lump. Medullary lesions may present as a pathological fracture. Although chondrosarcoma may develop in any of the bones that normally develop in cartilage, almost 50 per cent appear in the metaphysis of one of the long tubular bones, mostly in the lower limbs. The next most common sites are the pelvis and the ribs. Despite the relatively frequent occurrence of benign cartilage tumours in the small bones of the hands and feet, malignant lesions are rare at these sites.

Chondrosarcomas take various forms, usually designated according to:

(a) their location in the bone (central or peripheral);

(b) whether they develop without precedent (primary chondrosarcoma) or by malignant change in a pre-existing benign lesion (secondary chondrosarcoma); and

(c) the predominant cell type in the tumour.

By far the majority of chondrosarcomas fall into two well-defined categories: central tumours occupying the medullary cavity of the bone, and so-called 'peripheral tumours' growing out from the cortex. Less common varieties are juxtacortical chondrosarcoma, clear-cell chondrosarcoma and mesenchymal chondrosarcoma.

Central chondrosarcoma The tumour develops in the medullary cavity of either tubular or flat bones, most commonly at the proximal end of the femur or in the innominate bone of the pelvis. X-rays show an expanded, somewhat radiolucent area in the bone, with flecks of increased density due to calcification within the tumour. Aggressive lesions may take on a globular appearance with scalloping or destruction of the cortex. When a benign medullary chondroma (enchondroma) undergoes malignant transformation, it is difficult to be sure that the lesion was not a slowly evolving sarcoma from the outset.

Peripheral chondrosarcoma This tumour usually arises in the cartilage cap of an exostosis (osteochondroma) that has been present since childhood. Exostoses of the pelvis and scapula seem to be more susceptible than others to malignant change, but perhaps this is simply because the site allows a tumour to grow without being detected and removed at an early stage. Xrays show the bony exostosis, often surmounted by clouds of patchy calcification in the otherwise unseen lobulated cartilage cap. A tumour that is very large and calcification that is very fluffy and poorly outlined are suspicious features, but the clearest sign of malignant change is a demonstrable progressive enlargement of an osteoch ondroma after the end of normal bone growth. MRI is the best means of showing the size and internal features of the cartilage cap.

Juxtacortical (periosteal) chondrosarcoma Here the lesion appears as an excrescence on the surface of one of the tubular bones – usually the femur. It arises from the outermost layers of the cortex, deep to the periosteum. X-ray changes comprise features of both a chondrosarcoma and a periosteal osteosarcoma: an outgrowth from the bone surface, often containing flecks of calcification, as well as 'sunray' streaks and new-bone formation at the margins of the stripped periosteum. The dominant cell type is chondroblastic but there may also be sparse osteoid formation, leading one to doubt whether this is a cartilage tumour or a non-aggressive osteosarcoma.

Clear-cell chondrosarcoma There is some doubt as to whether this rare tumour is really a chondrosarcoma. In some respects the tumour resembles an aggressive chondroblastoma (e.g. its typical location in the head of the femur rather than the metaphysis). However, despite the fact that it is very slow-growing, it does eventually metastasize.

Mesenchymal chondrosarcoma This is an equally controversial entity. It tends to occur in younger individuals and in about 50 per cent of cases the tumour lies in the soft tissues outside an adjacent bone. The x-ray appearances are similar to those of the common types of chondrosarcoma but the clinical behaviour of the tumour is usually more aggressive. Histology shows a mixture of mesenchymal cells and chondroid tissue.

Treatment

Since most chondrosarcomas are slow-growing and metastasize late, they present the ideal case for wide excision and prosthetic replacement, provided it is certain that the lesion can be completely removed without exposing the tumour and without causing an unacceptable loss of function; in that case amputation may be preferable. In some cases isolated pulmonary metastases can be resected. The tumour does not respond to either radiotherapy or chemotherapy. Prognosis is determined largely by the cellular grade and the resection margin. There is a tendency for these tumours to recur late and the patient should therefore be followed up for 10 years or longer.

OSTEOSARCOMA

In its classic (intramedullary) form, osteosarcoma is a highly malignant tumour arising within the bone and spreading rapidly outwards to the periosteum and surrounding soft tissues. It is said to occur predominantly in children and adolescents, but epidemiological studies suggest that between 1972 and 1981 the age of presentation rose significantly (Stark et al., 1990). It may affect any bone but most commonly involves the long-bone metaphyses, especially around the knee and at the proximal end of the humerus.

Pain is usually the first symptom; it is constant, worse at night and gradually increases in severity. Sometimes the patient presents with a lump. Pathological fracture is rare. On examination there may be little to find except local tenderness. In later cases there is a palp able mass and the overlying tissues may appear swollen and inflamed. The ESR is usually raised and there may be an increase in serum alkaline phosphatase.

X-rays

The x-ray appearances are variable: hazy osteolytic areas may alternate with unusually dense osteoblastic areas. The endosteal margin is poorly defined. Often the cortex is breached and the tumour extends into the adjacent tissues; when this happens, streaks of new bone appear, radiating outwards from the cortex – the so-called 'sunburst' effect. Where the tumour emerges from the cortex, reactive new bone forms at the angles of periosteal elevation (Codman's triangle). While both the sunburst appearance and Codman's triangle are typical of osteosarcoma, they may occasionally be seen in other rapidly growing tumours.

Diagnosis and staging

In most cases the diagnosis can be made with confidence on the x-ray appearances. However, atypical lesions can cause confusion. Conditions to be excluded are post-traumatic swellings, infection, stress fracture and the more aggressive 'cystic' lesions. Other imaging studies are essential for staging purposes. Radioisotope scans may show up skip lesions, but a negative scan does not exclude them. CT and MRI reliably show the extent of the tumour. Chest xrays are done routinely, but pulmonary CT is a much more sensitive detector of lung metastases. About 10 per cent of patients have pulmonary metastases by the time they are first seen.

A biopsy should always be carried out before commencing treatment; it must be carefully planned to allow for complete removal of the tract when the tumour is excised.

Pathology

The tumour is usually situated in the metaphysis of a long bone, where it destroys and replaces normal bone. Areas of bone loss and cavitation alternate with dense patches of abnormal new bone. The tumour extends within the medulla and across the physeal plate. There may be obvious spread into the soft tissues with ossification at the periosteal margins and streaks of new bone extending into the extraosseous mass. The histological appearances show considerable variation: some areas may have the characteristic spindle cells with a pink-staining osteoid matrix; others may contain cartilage cells or fibroblastic tissue with little or no osteoid. Several samples may have to be examined; pathologists are reluctant to commit themselves to the diagnosis unless they see evidence of osteoid formation.

Treatment

The appalling prognosis that formerly attended this tumour has markedly improved, partly as a result of better diagnostic and staging procedures, and possibly because the average age of the patients has increased, but mainly because of advances in chemotherapy to control metastatic spread. However, it is still important to eradicate the primary lesion completely; the mortality rate after local recurrence is far worse than following effective ablation at the first encounter. The principles of treatment are outlined on page 192. After clinical assessment and advanced imaging, the patient is admitted to a special centre for biopsy. The lesion will probably be graded IIA or IIB. Multiagent neoadjuvant chemotherapy is given for 8–12 weeks and then, provided the tumour is resectable and there are no skip lesions, a wide resection is carried out. Depending on the site of the tumour, preparations would have been made to

replace that segment of bone with either a large bone graft or a custommade implant; in some cases an amputation may be more appropriate. The pathological specimen is examined to assess the response to preoperative chemotherapy. If tumour necrosis is marked (more than 90 per cent), chemotherapy is continued for another 6–12 months; if the response is poor, a different chemotherapeutic regime is substituted. Pulmonary metastases, especially if they are small and peripherally situated, may be completely resected with a wedge of lung tissue.

Outcome

Long-term survival after wide resection and chemotherapy has improved from around 50 per cent in 1980 (Rosen et al., 1982; Carter et al., 1991) to over 60 per cent in recent years (Smeland et al., 2004). Tumour-replacement implants usually function well. There is a fairly high complication rate (mainly wound breakdown and infection) but, in patients who survive, 10-year survival with mechanical failure as the end point is 75 per cent and for failure for any cause is 58 per cent. The limb salvage rate at 20 years is 84 per cent (Jeys et al., 2008) Aseptic loosening is more prevalent in younger patients.

VARIANTS OF OSTEOSARCOMA

PAROSTEAL OSTEOSARCOMA

This is a low-grade sarcoma situated on the surface of one of the tubular bones, usually at the distal femoral or proximal tibial metaphysis. The patient is a young adult who presents with a slowly enlarging mass near the bone end. X-ray shows a dense bony mass on the surface of the bone or encircling it; the cortex is not eroded and usually a thin gap remains between cortex and tumour. The picture is easily mistaken for that of a benign bone lesion and the diagnosis is often missed until the tumour recurs after local excision. CT and MRI will show the boundary between tumour and surrounding soft tissues. Although the lesion is outside the bone, it does not spread into the adjacent muscle compartment until fairly late. Staging, therefore, often defines it as a lowgrade intracompartmental tumour (stage IA).

Pathology At biopsy the tumour appears as a hard mass. On microscopic examination the lesion consists of well-formed bone but without any regular trabecular arrangement. The spaces between trabeculae are filled with cellular fibroblastic tissue; a few atypical cells and mitotic figures can usually be found. Occasionally the tumour has a much more aggressive appearance (dedifferentiated parosteal osteosarcoma).

Treatment For a low-grade parosteal osteosarcoma, wide excision without adjuvant therapy is sufficient to ensure a recurrence rate below 10 per cent. Dedifferentiated parosteal osteosarcoma should be treated in the same way as intramedullary sarcoma.

PERIOSTEAL OSTEOSARCOMA

This rare tumour is quite distinct from parosteal osteo - sarcoma. It is more like an intramedullary osteosarcoma, but situated on the surface of the bone. It occurs in young adults and causes local pain and swelling. X-ray shows a superficial defect of the cortex, but CT and MRI may reveal a larger soft-tissue mass. The appearances sometimes suggest a periosteal chondroma and the diagnosis may not be certain until a biopsy is performed.

Pathology Histologically this is a true osteosarcoma, but characteristically the sections show a prominent cartilaginous element.

Treatment Treatment is the same as that of classic osteosarcoma.

PAGET'S SARCOMA

Paget's disease affects about 2 per cent of western Europeans. Although malignant transformation is a rare complication of this disease, most osteosarcomas appearing after the age of 50 years fall into this category. Warning signs are the appearance of pain or swelling in a patient with longstanding Paget's disease. In late cases, pathological fracture may occur.

X-ray shows the usual features of Paget's disease, but with areas of bone destruction and soft-tissue invasion. This is a high-grade tumour – if anything even more malignant than classic osteosarcoma. Staging usually shows that extracompartmental spread has occurred; most patients have pulmonary metastases by the time the tumour is diagnosed.
Treatment Even with radical resection or amputation and chemotherapy the 5-year survival rate is low. If the lesion is definitely extracompartmental, palliative treatment by radiotherapy may be preferable; chemotherapy is usually difficult because of the patient's age and uncertainty about renal and cardiac function.

EWING'S SARCOMA

Ewing's sarcoma is believed to arise from endothelial cells in the bone marrow. It occurs most commonly between the ages of 10 and 20 years, usually in a tubular bone and especially in the tibia, fibula or clavicle. The patient presents with pain – often throbbing in character – and swelling. Generalized illness and pyrexia, together with a warm, tender swelling and a raised ESR, may suggest a diagnosis of osteomyelitis.

Imaging

X-rays usually show an area of bone destruction which, unlike that in osteosarcoma, is predominantly in the mid-diaphysis. New bone formation may extend along the shaft and sometimes it appears as fusiform layers of bone around the lesion – the socalled 'onion-peel' effect. Often the tumour extends into the surrounding soft tissues, with radiating streaks of ossification and reactive periosteal bone at the proximal and distal margins. These features (the 'sunray' appearance and Codman's triangles) are usually associated with osteosarcoma, but they are just as common in Ewing's sarcoma. CT and MRI reveal the large extraosseous component. Radioisotope scans may show multiple areas of activity in the skeleton.

Pathology

Macroscopically the tumour is lobulated and often fairly large. It may look grey (like brain) or red (like redcurrant jelly) if haemorrhage has occurred into it. Microscopically, sheets of small dark polyhedral cells with no regular arrangement and no ground substance are seen.

Diagnosis

The condition which should be excluded as rapidly as possible is bone infection. On biopsy the essential step is to recognize this as a malignant round-cell tumour, distinct from osteosarcoma. Other round-cell tumours that may resemble Ewing's are reticulum-cell sarcoma (see below) and metastatic neuroblastoma.

Treatment

The prognosis is always poor and surgery alone does little to improve it. Radiotherapy has a dramatic effect on the tumour but overall survival is not much enhanced. Chemotherapy is much more effective, offering a 5-year survival rate of about 50 per cent (Souhami and Craft, 1988; Damron et al., 2007). The best results are achieved by a combination of all three methods: a course of preoperative neoadjuvant chemotherapy; then wide excision if the tumour is in a favourable site, or radiotherapy followed by local excision if it is less accessible; and then a further course of chemotherapy for 1 year. Postoperative radiotherapy may be added if the resected specimen is found not to have a sufficiently wide margin of normal tissue.
The prognosis for these tumours has improved dramatically since the introduction of multi-agent chemotherapy – from an erstwhile 10 per cent survival rate to the current 70 per cent for patients with nonmetastatic Ewing's sarcoma.

METASTATIC BONE DISEASE

The skeleton is one of the commonest sites of secondary cancer; in patients over 50 years bone metastases are seen more frequently than all primary malignant bone tumours together.

The commonest source is carcinoma of the breast; next in frequency are carcinomas of the prostate, kidney, lung, thyroid, bladder and gastrointestinal tract. In about 10 per cent of cases no primary tumour is found. The commonest sites for bone metastases are the vertebrae, pelvis, the proximal half of the femur and the humerus. Spread is usually via the blood stream; occasionally, visceral tumours spread directly to adjacent bones (e.g. the pelvis or ribs). Metastases are usually osteolytic, and pathological fractures are common. Bone resorption is due either to the direct action of tumour cells or to tumour-derived factors that stimulate osteoclastic activity. Osteoblastic lesions are uncommon; they usually occur in prostatic carcinoma.

Clinical features

The patient is usually aged 50–70 years; with any destructive bone lesion in this age group, the differential diagnosis must include metastasis. Pain is the commonest – and often the only – clinical feature. The sudden appearance of backache or thigh pain in an elderly person (especially someone known to have been treated for carcinoma in the past) is always suspicious. If x-rays do not show anything, a radionuclide scan might. Some deposits remain clinically silent and are discovered incidentally on x-ray examination or bone scanning, or after a pathological fracture. Sudden collapse of a vertebral body or a fracture of the mid-shaft of a long bone in an elderly person are ominous signs; if there is no history and no clinical clue pointing to a primary carcinoma, a biopsy of the fracture area is essential. Symptoms of hypercalcaemia may occur (and are often missed) in patients with skeletal metastases. These include anorexia, nausea, thirst, polyuria, abdominal pain, general weakness and depression. In children under 6 years of age, metastatic lesions are most commonly from adrenal neuroblastoma. The child presents with bone pain and fever; examination reveals the abdominal mass.

Imaging

X-rays Most skeletal deposits are osteolytic and appear as rarified areas in the medulla or produce a moth-eaten appearance in the cortex; sometimes there is marked bone destruction, with or without a pathological fracture. Osteoblastic deposits suggest a prostatic carcinoma; the pelvis may show a mottled increase in density which has to be distinguished from Paget's disease or lymphoma.

Radioscintigraphy Bone scans with 99mTc-MDP are the most sensitive method of detecting 'silent' metastatic deposits in bone; areas of increased activity are selected for x-ray examination.

Special investigations

The ESR may be increased and the haemoglobin concentration is usually low. The serum alkaline phosphatase concentration is often increased, and in prostatic carcinoma the acid phosphatase also is elevated. Patients with breast cancer can be screened by measuring blood levels of tumour-associated antigen markers.

Treatment

By the time a patient has developed secondary deposits the prognosis for survival is poor. Occasionally, radical treatment (combined chemotherapy, radiotherapy and surgery) targeted at a solitary secondary deposit and the parent primary lesion may be rewarding and even apparently curative. This applies particularly to solitary renal cell, breast and thyroid tumour metastases; but in the great majority of cases, and certainly in those with multiple secondaries, treatment is entirely symptomatic. For that reason, elaborate witch-hunts to discover the source of an occult primary tumour are avoided, though it may be worthwhile investigating for tumours that are amenable to hormonal manipulation.

Prognosis

Bauer (1995) has suggested useful criteria for assessing prognosis (see Box). In his series of patients, survivorship at 1 year was as follows:
• of patients with 4 or 5 of Bauer's criteria 50 per cent were alive
• of patients with 2 or 3 criteria 25 per cent were alive
• of patients with only 1 or none of the criteria, the majority survived for less than 6 months and none were alive at 1 year.

BAUER'S POSITIVE CRITERIA FOR SURVIVAL
A solitary metastasis
No pathological fracture
No visceral metastases
Renal or breast primary
No lung cancer

Palliative care
Despite a poor prognosis, patients deserve to be made comfortable, to enjoy (as far as possible) their remaining months or years, and to die in a peaceful and dignified way. The active treatment of skeletal metastases contributes to this in no small measure. In addition, patients need sympathetic counselling and practical assistance with their material affairs.
Control of pain and metastatic activity Most patients require analgesics, but the more powerful narcotics should be reserved for the terminally ill. Unless specifically contraindicated, radiotherapy is used both to control pain and to reduce metastatic growth. This is often combined with other forms of treatment (e.g. internal fixation). Secondary deposits from breast or prostate can often be controlled by hormone therapy: stilboestrol for prostatic secondaries and androgenic drugs or oestrogens for breast carcinoma. Disseminated secondaries from breast carcinoma are sometimes treated by oophorectomy combined with adrenalectomy or by hypophyseal ablation.
Hypercalcaemia may have serious consequences, including renal acidosis, nephrocalcinosis, unconsciousness and coma. It should be treated by ensuring adequate hydration, reducing the calcium intake and, if necessary, administering bisphosphonates.

SOFT-TISSUE TUMOURS

Benign soft-tissue tumours are common, malignant ones rare. The distinction between these two groups is not always easy, and some lesions, treated confidently as 'benign', recur in more aggressive form after inadequate removal. Features suggestive of malignancy are: pain in a previously painless lump; a rapid increase in size; a lump deep to the fascia; size greater than 5 cm; poor demarcation; and attachment to the surrounding structures. As with bone tumours, special imaging and staging should be carried out before the field is disturbed by operation. Chest x-rays and blood investigations may also be necessary. If the imaging is conclusive then the lesion can be removed with either a marginal or wide excision biopsy, dependent on the diagnosis. Alternatively, a biopsy to confirm the diagnosis should be undertaken prior to excision. The role of chemotherapy for soft-tissue sarcomas is uncertain, except in the treatment of rhabdomyosarcoma and synovial sarcoma. Radiotherapy is indicated for all high-grade lesions and for tumours that are removed with poor margins or by intralesional excision. If margins are contaminated then re-operation with wide resection of that margin must be performed. The account that follows is intended as a summary of those soft-tissue tumours likely to be encountered in orthopaedics.

LIPOMA

A lipoma, one of the commonest of all tumours, may occur almost anywhere; sometimes there are multiple lesions. The tumour usually arises in the subcutaneous layer. It consists of lobules of fat with a surrounding capsule which may become tethered to neighbouring structures. The patient, usually aged over 50, complains of a painless swelling. The lump is soft and almost fluctuant; the well-defined edge and lobulated surface distinguish it from a chronic abscess. Fat is notably radiotranslucent, a feature that betrays the occasional subperiosteal lipoma.

LIPOSARCOMA

Liposarcoma is rare but should be suspected if a fatty tumour (especially in the buttock, the thigh or the popliteal fossa) goes on growing and becomes painful. The lump may feel quite firm and is usually not translucent. CT or MRI is essential to determine the extent of the tumour. Treatment depends on the degree of malignancy. Lowgrade lesions can be removed by wide excision; highgrade tumours need radical resection. For liposarcomas in inaccessible sites, radiation therapy is often effective.

SYNOVIAL TUMOURS

PIGMENTED VILLONODULAR SYNOVITIS AND GIANTCELL TUMOUR OF TENDON SHEATH

These are two forms of the same condition – a benign disorder that occurs wherever synovial membrane is found: in joints, tendon sheaths or bursae.

Pigmented villonodular synovitis (PVNS) presents as a longstanding boggy swelling of the joint – usually the hip, knee or ankle – in an adolescent or young adult. Xray may show excavations in the juxta- articular bone on either side of the joint. When the joint is opened, the synovium is swollen and hyperplastic, often covered with villi and golden-brown in colour – the effect of haemosiderin deposition. The juxta-articular excavations contain clumps of friable synovial material.

Tendon sheath lesions are seen mainly in the hands and feet, where they cause nodular thickening of the affected sheath. X-ray may show pressure erosion of an adjacent bone surface – for example, on one of the phalanges. At operation the boggy synovial tissue is often yellow; this type of lesion is sometimes called *xanthoma of tendon sheath.*

Pathology Histologically, joint and tendon sheath lesions are identical. There is proliferation and hypertrophy of the synovium, which contains fibroblastic tissue with foamy histiocytes and multinucleated giant cells. These features have engendered yet another name for the same condition: *giant-cell tumour of tendon sheath.*

Treatment The only effective treatment is synovectomy. Although the tumour does not undergo malignant change, the recurrence rate is high unless excision is complete. This may be unattainable and subtotal synovectomy is then sometimes combined with local radiotherapy. If, despite such aggressive treatment, there are repeated recurrences, it may be necessary to sacrifice the joint and carry out arthroplasty or arthrodesis.

SYNOVIAL SARCOMA

This malignant tumour usually develops near synovial joints in adolescents and young adults. However, only about 20 per cent involve the joint itself and the term ‘synovioma’ is a misnomer because this is not a tumour of synovium, though the histological appearance may resemble that of synovium. The patient usually complains of rapid enlargement of a lump around one of the larger joints – the hip, the knee or the shoulder. Occasionally the tumour

presents as a small swelling in the hand or foot and the histological diagnosis comes as a complete surprise. Pain is a common feature and many lesions are present for years before they are diagnosed. *X-rays* show a soft-tissue mass, sometimes with extensive calcification. *MRI* will help to outline the tumour.

Biopsy reveals a fleshy lesion composed of proliferative 'synovial' cells and fibroblastic tissue; characteristically the cellular areas are punctured by vacant slits that give the tissue an acinar appearance. Cellular abnormality and mitoses reflect the degree of malignancy. Small, well-defined lesions can be treated by wide excision. High-grade lesions, which usually have illdefined margins, require radical resection – and this may mean radical amputation. Resection may be combined with radiotherapy and occasionally chemotherapy.

MUSCLE TUMOURS

Tumours of muscle are rare; only those that occur in the striped muscle of the extremities are considered here.

RHABDOMYOMA

Rhabdomyoma is a rare cause of a lump in the muscle. It is occasionally confused with the 'lump' that appears after muscle rupture: both are in the line of a muscle, can be moved across but not along it, and harden with muscle contraction. However, with muscle rupture symptoms appear quite suddenly, there is a depression proximal or distal to the lump and the swelling does not grow any bigger. If a tumour is suspected, early exploration and biopsy are advisable because malignant change may occur. If the diagnosis is confirmed, the tumour should be excised.

RHABDOMYOSARCOMA

Malignant tumours are occasionally seen in the muscles around the shoulder or hip. The patient – usually a young adult – presents with ache and an enlarging, ill-defined lump that moves with the affected muscle.

CT and MRI show that the mass is in the muscle, but the edge may be poorly demarcated because the tumour tends to spread along the fascial planes. At biopsy the tissue looks and feels different from normal muscle and microscopic examination shows clusters of highly abnormal muscle cells. This is a high-grade lesion which requires radical resection of the affected muscle, i.e. from its origin to its insertion. If this cannot be assured or if the tumour has spread beyond the fascial sheath, amputation is advisable. Recurrent lesions are also treated by amputation. If complete removal is impossible, adjunctive radiotherapy may lessen the risk of recurrence.

Section 2

Regional Orthopaedics

The shoulder and pectoral girdle 13

CLINICAL ASSESSMENT
SYMPTOMS
Pain is the commonest symptom. However, ‘pain in the shoulder’ is not necessarily ‘shoulder pain’! If the patient points to the top of the shoulder, think of the acromioclavicular joint, or referred pain from the neck. Pain from the shoulder joint and the rotator cuff is felt, typically, over the front and outer aspect of the joint, often as far down as the middle of the arm.
The relationship to posture may be significant: pain which appears when the arm is in the ‘windowcleaning’ position is characteristic of rotator cuff impingement; pain which comes on suddenly when the arm is held high overhead suggests instability. Beware the trap of referred pain. Mediastinal disorders, including cardiac ischaemia, can present with aching in either shoulder. Weakness may appear as a true loss of power, suggesting a neurological disorder, or as a sudden and surprising inability to abduct the shoulder – perhaps due to a tendon rupture. Between these extremes there is weakness in performing only certain movements and weakness associated with pain. Instability symptoms may be gross and alarming (‘my shoulder jumps out of its socket when I raise my arm’); more often they are quite subtle: a click or jerk when the arm is held overhead, or the ‘dead arm’ sensation that overtakes the tennis player as he or she prepares to serve. Stiffness may be progressive and severe – so much so as to merit the term ‘frozen shoulder’. Swelling may be in the joint, the muscle or the bone; the patient will not know the difference. Deformity may consist of muscle wasting, prominence of the acromioclavicular joint, winging of the scapula or an abnormal position of the arm. Loss of function is usually expressed as difficulty with dressing and grooming, or inability to lift objects or work with the arm above shoulder height.
SIGNS
The patient should always be examined from in front and from behind. Both upper limbs, the neck, the outlineof the scapula and the upper chest must be visible.
Look
Skin Scars or sinuses are noted; do not forget the axilla!
Shape The two sides should be compared. Asymmetry of the shoulders, winging of the scapula, wasting of the deltoid, supraspinatus and infraspinatus muscles and

acromioclavicular dislocation are best seen from behind; swelling of the acromioclavicular or sternoclavicular joint or wasting of the pectoral muscles is more obvious from the front. A joint effusion causes swelling anteriorly and occasionally 'points' in the axilla. Wasting of the deltoid suggests a nerve lesion whereas wasting of the supraspinatus may be due to either a full-thickness tear or a suprascapular nerve lesion. The typical 'Popeye' bulge of a ruptured biceps is more easily seen if the elbow is flexed.

Position If the arm is held internally rotated, think of posterior dislocation of the shoulder.

Feel

Skin Because the joint is well covered, inflammation rarely influences skin temperature.

Bony points and soft tissues The deeper structures are carefully palpated, following a mental picture of the anatomy. Start with the sternoclavicular joint, then follow the clavicle laterally to the acromioclavicular joint, and so onto the anterior edge of the acromion and around the acromion. The anterior and posterior margins of the glenoid should be palpated. With the shoulder held in extension, the supraspinatus tendon can be pinpointed just under the anterior edge of the acromion; below this, the bony prominence bounding the bicipital groove is easily felt, especially if the arm is gently rotated so that the hard ridge slips medially and laterally under the palpating fingers. Crepitus over the supraspinatus tendon during movement suggests tendinitis or a tear.

Move

Active movements Movements are observed first from in front and then from behind, with the patient either standing or sitting. Sideways elevation of the arms normally occurs in the plane of the scapula, i.e. about 20 degrees anterior to the coronal plane, with the arm rising through an arc of 180 degrees. However, by convention, abduction is performed in the coronal plane and flexion–extension in the sagittal plane.

Abduction starts at 0 degrees; the early phase of movement takes place almost entirely at the glenohumeral joint, but as the arm rises the scapula begins to rotate on the thorax and in the last 60 degrees of movement is almost entirely scapulo-thoracic (hence sideways movement beyond 90 degrees is sometimes called 'elevation' rather than 'abduction'). The rhythmic transition from gleno-humeral to scapulothoracic movement is disturbed by disorders in the joint or by dysfunction of the stabilizing tendons around the joint. Thus, abduction may be (1) difficult to initiate, (2) diminished in range or (3) altered in rhythm, the scapula moving too early and creating a shrugging effect. If movement is painful, the arc of pain must be noted; pain in the mid-range of abduction suggests a minor rotator cuff tear or supraspinatus tendinitis; pain at the end of abduction is often due to acromioclavicular arthritis. Flexion and extension are examined by asking the patient to raise the arms forwards and then backwards. The normal range is 180 degrees of flexion and 40 degrees of extension.

Rotation is tested in two ways: The arms are held close to the body with the elbows flexed to 90 degrees; the hands are then separated as widely as possible (external rotation) and brought together again across the body (internal rotation). This is a rather unnatural movement and one learns more by simply asking the patient to clasp his (or her) fingers behind his neck (external rotation in abduction) and then to reach up his back with his fingers (internal rotation in adduction); the two sides are compared.

Passive movements To test the range of glenohumeral movement (as distinct from combined glenohumeral and scapular movement) the scapula must first be anchored; this is done by the examiner pressing firmly down on the top of the shoulder with one hand while the other hand moves the patient's arm. Grasping the angle of the scapula as a method of anchorage is less satisfactory.

Power The deltoid is examined for bulk and tautness while the patient abducts against resistance. To test serratus anterior (long thoracic nerve, C5, 6, 7) the patient is asked to push forcefully against a wall with both hands; if the muscle is weak, the scapula is not stabilized

on the thorax and stands out prominently (winged scapula). Pectoralis major is tested by having the patient thrust both hands firmly into the waist. Rotator power is tested by asking the patient to stand with his arms tucked into his side and the elbows flexed, then to externally rotate against resistance. Weakness may be associated with a rotator cuff lesion, instability or a neurological disorder.

Other systems Clinical assessment is completed by examining the cervical spine (as a common source of referred pain), testing for generalized joint laxity (a frequent accompaniment of shoulder instability) and performing a focussed neurological examination.

Special clinical tests

Special clinical tests have been developed for localizing more precisely the site of pain and tenderness, the source of muscle weakness and the presence of instability. These are described in the relevant sections that follow.

Examination after local anaesthetic injection

It is sometimes possible to localize the source of shoulder pain by injecting local anaesthetic into the target site (for example the supraspinatus tendon or the acromioclavicular joint) and thus to see whether there is a temporary reduction in pain on movement. Injection into the subacromial space may help to distinguish loss of movement due to pain from that due to a rotator cuff tear.

Diagnostic focus

Important as it is to adopt a systematic approach in the clinical examination, the practical exercise of working towards a diagnosis requires a sensible balance in the focus of attention. A young athletic person who develops pain and weakness on abduction and external rotation of the shoulder is more likely to be suffering from a rotator cuff disorder than an inflammatory arthritis of the shoulder and therefore the full panoply of special tests for localization of pain and weakness would be justified, whereas some of these tests would be quite inappropriate in an elderly person with the longstanding pain and swelling of an arthritic condition.

DISORDERS OF THE ROTATOR CUFF

The rotator cuff is made up of the lateral portions of the infraspinatus, supraspinatus and subscapularis muscles and their conjoint tendon which is inserted into the greater tuberosity of the humerus. The musculo tendinous cuff passes beneath the coracoacromial arch, from which it is separated by the subacromial bursa; during abduction of the arm the cuff slides outwards under the arch. The deep surface of the cuff is intimately related to the joint capsule and the tendon of the long head of the biceps. Although contraction of the individual muscles that make up the rotator cuff exerts a rotational pull on the proximal end of the humerus, the main function of the conjoint structure is to draw the head of the humerus firmly into the glenoid socket and stabilize it there when the deltoid muscle contracts and abducts the arm. Consequently, patients with rotator cuff tendinitis experience pain and weakness on active abduction and those with a severe tear of the cuff are unable to initiate abduction but can hold the arm abducted once it has been raised aloft by the examiner. The commonest cause of pain around the shoulder is a disorder of the rotator cuff. This is sometimes referred to rather loosely as *'rotator cuff syndrome'*, which comprises at least four conditions with distinct clinical features and natural history:

- supraspinatus impingement syndrome and tendinitis
- tears of the rotator cuff
- acute calcific tendinitis
- biceps tendinitis and/or rupture.

In all these conditions the patient is likely to complain of pain and/or weakness during certain movements of the shoulder. Pain may have started recently, sometimes quite suddenly, after a particular type of exertion; the patient may know precisely which movements now reignite the pain and which to avoid, providing a valuable clue to its origin. 'Rotator cuff' pain typically appears over the front and lateral aspect of the shoulder during activities with the arm abducted and medially rotated, but it may be present even with the arm at rest. Tenderness is felt at the anterior edge of the acromion. Pain and tenderness directly in front along the delto-pectoral boundary could be associated with the biceps tendon. Localized pain over the top of the shoulder is more likely to be due to acromioclavicular pathology, and pain at the back along the scapular border may come from the cervical spine. All these sites should be inspected for muscle wasting, carefully palpated for local tenderness and constantly compared with the opposite shoulder. If there is weakness with some movements but not with others, then one must rule out a partial or complete tendon rupture; here again, as with pain, localization to a specific site is the key to diagnosis. In both cases clinical examination should include a number of provocative tests to determine the source of the patient's symptoms.

Cuff disruption

The most advanced stage of the disorder is progressive fibrosis and disruption of the cuff, resulting in either a partial or full thickness tear. The patient is usually aged over 45 and gives a history of refractory shoulder pain with increasing stiffness and weakness.

Partial tears may occur within the substance or on the deep surface of the cuff and are not easily detected, even on direct inspection of the cuff. They are deceptive also in that continuity of the remaining cuff fibres permits active abduction with a painful arc, making it difficult to tell whether chronic tendinitis is complicated by a partial tear.

A *full thickness tear* may follow a long period of chronic tendinitis, but occasionally it occurs spontaneously after a sprain or jerking injury of the shoulder. There is sudden pain and the patient is unable to abduct the arm. Passive abduction also may, in the early stages, be limited or prevented by pain. If the diagnosis is in doubt, pain can be eliminated by injecting a local anaesthetic into the subacromial space. If active abduction is now possible the tear must be only partial. If active abduction remains impossible, then a complete tear is likely.

CALCIFICATION OF THE ROTATOR CUFF

ACUTE CALCIFIC TENDINITIS

Acute shoulder pain may follow deposition of calcium hydroxyapatite crystals, usually in the 'critical zone' of the supraspinatus tendon slightly medial to its insertion, occasionally elsewhere in the rotator cuff. The condition is not unique to the shoulder, and similar lesions are seen in tendons and ligaments around the ankle, knee, hip and elbow. The cause is unknown but it is thought that local ischaemia leads to fibrocartilaginous metaplasia and deposition of crystals by the chondrocytes. Calcification alone is probably not painful; symptoms, when they occur, are due to the florid vascular reaction which produces swelling and tension in the tendon. Resorption of the calcific material is rapid and it may soften or disappear entirely within a few weeks.

Clinical features

The condition affects 30–50 year-olds. Aching, sometimes following overuse, develops and increases in severity within hours, rising to an agonizing climax. After a few days, pain subsides and the shoulder gradually returns to normal. In some patients the process is less dramatic and recovery slower. During the acute stage the arm is held immobile; the joint is usually too tender to permit palpation or movement.

X-RAYS

Calcification just above the greater tuberosity is always present. An initially well-demarcated depositbecomes more ‘woolly’ and then disappears.

Treatment

NON-OPERATIVE TREATMENT

Conservative treatment is successful in up to 90 per cent of patients. The main methods are non-steroidal anti-inflammatory drugs, subacromial injection of corticosteroids, physiotherapy, extracorporeal shockwave therapy, needle aspiration and irrigation.

Non-steroidal anti-inflammatory drugs are the mainstay of non-operative treatment. Although corticosteroid injections are commonly used in the treatment of calcifying tendinitis, there is no conclusive evidence that they promote resorption of the calcium deposit. The efficacy of physiotherapy in the form of therapeutic ultrasound remains uncertain.

Extracorporeal shockwave therapy employs acoustic waves to induce fragmentation of the mechanically hard crystals. Its use as an alternative treatment for calcifying tendinitis has gained increasing popularity in the last few years and its efficacy has been confirmed in several prospective studies which show that the deposit disappears in up to 86 per cent of cases with a significant reduction in pain. However, most of these studies have only a short-term follow-up.

Needle aspiration and irrigation (barbotage) aims to drain a substantial portion of the calcium deposit, thereby stimulating cell-mediated progressive resorption. Needle aspiration can be readily done underlocal anaesthesia in the outpatient setting with ultrasound guidance. A combination of local anaesthetic and corticosteroid is used. The best results are obtained in patients with an acutely painful shoulder, typically during the resorption stage in which the calcium is of toothpaste-like consistency.

OPERATIVE TREATMENT

While operative treatment is still a controversial issue, there is wide agreement that surgery is indicated for patients with severe disabling symptoms which have persisted for more than 6 months and are resistant to conservative treatment. The procedure involves a gleno-humeral arthroscopy with special attention to the ‘critical zone’ of the rotator cuff. Once the calcium deposit is identified, the capsule is carefully incised from the bursal side with a knife in line with fibre orientation of the tendon; a curette is then used to milk out the toothpaste-like deposit. A subacromial decompression is also usually performed.

CHRONIC CALCIFICATION

Asymptomatic calcification of the rotator cuff is common and often appears as an incidental finding in shoulder x-rays. When it is seen in association with the impingement syndrome, it is tempting to attribute the symptoms to the only obvious abnormality supraspinatus calcification. However, the connection is spurious and treatment should be directed at the impingement lesion rather than the calcification.

LESIONS OF THE BICEPS TENDON

Tendinitis

The long head of biceps is subject to tenosynovitis because of its anatomy; the tendon has a synovial sheath and follows a constrained path in the bicipital groove. Bicipital tendinitis usually occurs together with rotator cuff impingement; rarely, it presents as an isolated problem in young people after unaccustomed shoulder strain. Tenderness is sharply localized to the bicipital groove. Two manoeuvres that often cause pain are: (1) resisted flexion with the elbow straight and the forearm supinated (Speed’s test); and (2) resisted supination of the forearm with the elbow bent (Yergason’s test).

Rest, local heat and deep transverse friction usually bring relief. If recovery is delayed, a corticosteroid injection will help. For refractory cases, a number of surgical solutions have been described including arthroscopic decompression, biceps tenotomy and biceps tenodesis.

Rupture

Rupture of the tendon of the long head of biceps usually accompanies rotator cuff disruption, but sometimes the biceps lesion is paramount. The patient is usually aged over 50. While lifting he or she feels something snap in the shoulder and the upper arm becomes painful and bruised. Ask the patient to flex the elbow: the detached belly of the biceps forms a prominent lump in the lower part of the arm. Isolated tears in elderly patients need no treatment. However, if the rupture is part of a rotator cuff lesion and especially if the patient is young and active - this is an indication for anterior acromioplasty; at the same time the distal tendon stump can be sutured to the bicipital groove (biceps tenodesis). Postoperatively the arm is lightly splinted with the elbow flexed for 4 weeks.

SLAP LESIONS

Compressiv loading of the shoulder in the flexed abducted position (e.g. in a fall on the outstretched hand) can damage the superior labrum anteriorly and posteriorly (SLAP). The injury of the superior labrum begins posteriorly and extends anteriorly, stopping before or at the mid-glenoid notch and including the 'anchor' of the biceps tendon to the labrum.

Four main types are described:

1. non-traumatic superior labral degeneration, usually in older people and often asymptomatic;
2. avulsion of the superior part of the labrum - the commonest type (Fig. 3.17);
3. a 'bucket handle' tear of the superior labrum;
4. as for type 3 with an extension into the tendon of long head of biceps.

Further subtypes that include associated lesions have also been described.

Clinical features

There is usually a history of a fall on the arm. As the initial acute symptoms settle, the patient continues to experience a painful 'click' on lifting the arm above shoulder height, together with loss of power when using the arm in that position. He or she may also complain of an inability to throw.

O'Briens test The patient is instructed to flex the arm to 90 degrees with the elbow fully extended and then to adduct the arm 10-15 degrees medial to the sagittal plane. The arm is then maximally internally rotated and the patient resists the examiner's downward force. The procedure is repeated in supination. Pain elicited by the first manoeuvre which is reduced or eliminated by the second signifies a positive test.

Imaging

MRI is the modality of choice though the diagnosis is best confirmed by arthroscopic examination and at the same time the lesion is treated by debridement or repair.

Treatment

Very few patients with SLAP lesion injuries return to full capability without surgical intervention. Arthroscopic repair of an isolated superior labral lesion is successful in the majority (91 per cent) of patients. However, the results in patients who participate in overhead sports are not as satisfactory as those in patients who are not involved in overhead sports (Seung-Ho Kim et al., 2002)

ADHESIVE CAPSULITIS (FROZEN SHOULDER)

The term 'frozen shoulder' should be reserved for a well-defined disorder characterized by progressive pain and stiffness of the shoulder which usually resolves spontaneously after about 18 months. The cause remains unknown. The histological features are reminiscent of Dupuytren's disease, with active fibroblastic proliferation in the rotator interval, anterior capsule and coraco-humeral ligament (Bunker, 1997. The condition is particularly associated with diabetes, Dupuytren's disease, hyperlipidaemia, hyperthyroidism, cardiac disease and hemiplegia. It occasionally appears after recovery from neurosurgery.

Clinical features

The patient, aged 40-60, may give a history of trauma, often trivial, followed by aching in the arm and shoulder. Pain gradually increases in severity and often prevents sleeping on the affected side. After several months it begins to subside, but as it does so stiffness becomes an increasing problem, continuing for another 6-12 months after pain has disappeared. Gradually movement is regained, but it may not return to normal and some pain may persist. Apart from slight wasting, the shoulder looks quite normal; tenderness is seldom marked. The cardinal feature is a stubborn lack of active and passive movement in all directions.

X-rays are normal unless they show reduced bone density from disuse. Their main value is to exclude other causes of a painful, stiff shoulder.

Diagnosis

Not every stiff or painful shoulder is a frozen shoulder, and indeed there is some controversy over the criteria for diagnosing 'frozen shoulder' (Zuckerman et al., 1994). Stiffness occurs in a variety of conditions -arthritic, rheumatic, post-traumatic and postoperative.

The diagnosis of frozen shoulder is clinical, resting on two characteristic features: (1) painful restriction of movement in the presence of normal xrays; and (2) a natural progression through three successive phases. When the patient is first seen, a number of conditions should be excluded:

Infection In patients with diabetes, it is particularly important to exclude infection. During the first day or two, signs of inflammation may be absent.

Post-traumatic stiffness After any severe shoulder injury, stiffness may persist for some months. It is maximal at the start and gradually lessens, unlike the pattern of a frozen shoulder.

Diffuse stiffness If the arm is nursed over-cautiously (e.g. following a forearm fracture) the shoulder may stiffen. Again, the characteristic pattern of a frozen shoulder is absent.

Reflex sympathetic dystrophy Shoulder pain and stiffness may follow myocardial infarction or a stroke. The features are similar to those of a frozen shoulder and it has been suggested that the latter is a form of reflex sympathetic dystrophy. In severe cases the whole upper limb is involved, with trophic and vasomotor changes in the hand (the shoulder-hand syndrome).

Treatment

CONSERVATIVE TREATMENT

Conservative treatment aims to relieve pain and prevent further stiffening while recovery is awaited. It is important not only to administer analgesics and antiinflammatory drugs but also to reassure the patient that recovery is certain. Exercises are encouraged, the most valuable being 'pendulum' exercises in which the patient leans forward at the hips and moves his arm as if stirring a giant pudding (this is really a form of assisted active movement, the assistance being supplied by gravity). However, the patient is warned that moderation and regularity will achieve more than sporadic masochism. The role of physiotherapy is unproven and the benefits of steroid injection are debatable. Manipulation under general anaesthesia

may improve the range of movement. The shoulder is moved gently but firmly into external rotation, then abduction and flexion. Special care is needed in elderly, osteoporotic patients as there is a risk of fracturing the neck of the humerus. At the end, the joint is injected with methylprednisolone and lignocaine. An alternative method of treatment is to distend the joint by injecting a large volume (50–200 mL) of sterile saline under pressure. Arthroscopy has shown that both manipulation and distension achieve their effect by rupturing the capsule. The results of conservative treatment are subjectively good, most patients eventually regaining painless and satisfactory function; however, examination is likely to show some residual restriction of movement (especially external rotation) in over 50 per cent of cases (Shaffer et al., 1992). Most studies on outcome are small. In the largest of these, Hand et al. (2008) reported on patients who were followed up for a mean of 4.4 years: 59 per cent had normal or near-normal shoulders, and of the remainder 94 per cent had only mild symptoms.

SURGICAL TREATMENT

Surgery does not have a well-defined role. The main indication is prolonged and disabling restriction of movement which fails to respond to conservative treatment. Arthroscopic capsular release is increasingly employed. New techniques enable the surgeon to release intra-articular, subacromial and subdeltoid adhesions without dividing the subscapularis. Active range of motion can be started immediately (Harryman et al., 1997).

INSTABILITY OF THE SHOULDER

The shoulder achieves its uniquely wide range of movement at the cost of stability. The humeral head is held in the shallow glenoid socket by the glenoid labrum, the gleno-humeral ligaments, the coracohumeral ligament, the overhanging canopy of the coracoacromial arch and the surrounding muscles. Failure of any of these mechanisms may result in instability of the joint. One must distinguish between *joint laxity* and *joint instability*. Joint laxity implies a degree of translation in the gleno-humeral joint which falls within a physiological range and which is asymptomatic. Joint is an abnormal symptomatic motion for that shoulder which results in pain, subluxation or dislocation of the joint.

Dislocation is defined as complete separation of the gleno-humeral surfaces, whereas *subluxation* implies a symptomatic separation of the surfaces without dislocation.

Pathogenetic classification

The aetiology and classification of shoulder instability is complex, although the Stanmore Instability Classification system developed at the Royal National Orthopaedic Hospital in London is now increasingly used. It recognizes that there are two broad reasons why shoulders become unstable:

(1) structural changes due to major trauma such as acute dislocation or recurrent micro-trauma; and

(2) unbalanced muscle recruitment (as opposed to muscle weakness) resulting in the humeral head being displaced upon the glenoid.

From a clinical and therapeutic point of view, **three polar types** of disorder can be identified:

Type I Traumatic structural instability.

Type II Atraumatic (or minimally traumatic) structural instability.

Type III Atraumatic non-structural instability (muscular dyskinesia).

The triangular relationship between these conditions allows for the fact there are intermediate types that lie between the ‘poles’; the balance of abnormalities can shift and patients may

'move' from one group to another over time or present with a combination of pathologies: for example, a purely structural disorder which, if allowed to persist, becomes associated with abnormal muscle patterning to the extent that *both conditions* need to be treated and the problems grow in complexity. The system also recognizes that there is a gradation in the opposite direction, from dyskinetic muscle patterning to structural abnormality (Lewis, Kitamura and Bayley, 2004).

RAPIDLY DESTRUCTIVE ARTHROPATHY (MILWAUKEE SHOULDER)

Occasionally, in the presence of longstanding or massive cuff tears, patients develop a rapidly progressive and destructive form of osteoarthritis in which there is severe erosion of the gleno-humeral joint, the acromion process and the acromioclavicular joint - what Neer and his colleagues (1983) called a *cuff tear arthropathy*. The changes are now attributed to hydroxyapatite crystal shedding from the torn rotator cuff and a synovial reaction involving the release of lysosomal enzymes (including collagenases) which lead to cartilage breakdown (McCarty et al., 1981). A similar condition is seen in other joints such as the hip and knee. The shoulder disorder, however, has come to be known as *Milwaukee shoulder*, after the city from whence McCarty hailed.

DISORDERS OF THE SCAPULA AND CLAVICLE

CONGENITAL ELEVATION OF THE SCAPULA

The scapulae normally complete their descent from the neck by the third month of fetal life; occasionally one or both scapulae remain incompletely descended. Associated abnormalities of the cervical spine are common and sometimes there is a family history of scapular deformity.

CLINICAL FEATURES

Two similar, and possibly related, conditions are encountered.

Sprengel's deformity Deformity is the only symptom and it may be noticed at birth. The shoulder on the affected side is elevated; the scapula looks and feels abnormally high, smaller than usual and somewhat prominent; occasionally both scapulae are affected. The neck appears shorter than usual and there may be kyphosis or scoliosis of the upper thoracic spine. Shoulder movements are painless but abduction and elevation may be limited by fixation of the scapula. *X-rays* will show the elevated scapula and any associated vertebral anomalies; sometimes there is also a bony bridge between the scapula and the cervical spine (the omo-vertebral bar).

Klippel–Feil syndrome This is usually a more widespread disorder. There is bilateral failure of scapular descent associated with marked anomalies of the cervical spine and failure of fusion of the occipital bones. Patients look as if they have no neck; there is a low hairline, bilateral neck webbing and gross limitation of neck movement. This condition should not be confused with *bilateral shortness* of the *sternomastoid muscle* in which the head is poked forward and the chin thrust up; the absence of associated congenital lesions is a further distinguishing feature.

CONGENITAL PSEUDARTHROSIS OF THE CLAVICLE

The typical clinical picture is that of a child with a painless lump in the mid-shaft of the clavicle. This always occurs on the right side, except in the presence of dextrocardia. X-ray shows the break in the clavicle, which usually heals only after excision of the ‘nonunion and bone grafting.

Treatment, if required, is by excision of the pseudarthrosis and bone grafting across the gap.

SCAPULAR INSTABILITY

Winging of the scapula is due to weakness of the serratus anterior muscle. It results in asymmetry of the shoulders but the deformity may not be obvious until the patient tries to contract the serratus anterior against resistance. The typical appearance is shown in Figure 13.33.

There are several causes of weakness or paralysis of the serratus anterior muscle:

- neuralgic amyotrophy (see page 259)
- injury to the brachial plexus (a blow to the top of the shoulder, severe traction on the arm or carrying heavy loads on the shoulder
- direct damage to the long thoracic nerve (e.g. during radical mastectomy)
- fascioscapulohumeral muscular dystrophy.

Disability is usually slight and is best accepted.

ARTHROPLASTY OF THE SHOULDER

Shoulder replacement was initially introduced by Neer in the 1950s for the treatment of proximal humeral fractures. Subsequent modifications and the introduction of glenoid resurfacing broadened the indications to include other disease processes, including end-stage gleno-humeral osteoarthritis and rheumatoid arthritis. If non-operative treatment fails, the two surgical options commonly considered are humeral head replacement (HHR) and total shoulder replacement (TSR). The optimal treatment choice, however, remains controversial.

<u>Indications</u>

The indications for arthroplasty are:

1. osteoarthritis causing pain and loss of movement
2. rheumatoid arthritis
3. complex fractures of the proximal humerus
4. avascular necrosis of the humeral head
5. tumours of the proximal humerus
6. severe arthritis with cuff arthropathy.

The elbow and forearm 14

CLINICAL ASSESSMENT

SYMPTOMS

Pain from the elbow is fairly diffuse and may extend into the forearm. Localized pain over the lateral or medial epicondyle of the humerus is usually due to tendinitis. The patient may have noticed that it is triggered, or aggravated, by certain activities. So often is this the case that the symptom has acquired colloquial definitions: 'tennis elbow' for lateral epicondylar pain and 'golfer's elbow' for medial epicondylar pain. Pain over the back of the elbow is often due to an olecranon bursitis. Remember that 'pain in the elbow' is sometimes referred pain from the cervical spine!

Stiffness, if it is mild, may hardly be noticed. If it is severe, it can be very disabling; the patient may be unable to reach up to the mouth (loss of flexion) or the perineum (loss of extension); limited supination makes it difficult to carry large objects. Swelling may be due to injury or inflammation; a soft lump on the back of the elbow suggests an olecranon bursitis. Deformity is uncommon except in rheumatoid arthritis and after trauma. Always ask about previous injuries.

Instability – the feeling that the elbow 'moves out of joint' – is due either to previous trauma or to destructive joint disease. Ulnar nerve symptoms (tingling, numbness and weakness of the hand) may occur in elbow disorders because of the nerve's proximity to the joint. Loss of function is noticed mainly in grooming, carrying and placing activities. However good the hand, if the elbow cannot put it out into the environment and bring it back to the individual, upper limb function is seriously degraded.

SIGNS

Both upper limbs should be completely exposed, and it is essential to look at the back of the elbow as well as the front. Often the neck, shoulders and hands also need to be examined.

Look

Both upper limbs must be completely exposed. The patient holds his or her arms alongside the body, elbows fully extended, with palms forwards. In this position the forearms are normally angled slightly outwards – a valgus or carrying angle of 5–15 degrees. 'Varus' or 'valgus' deformity is determined by angular deviations medialwards or lateralwards beyond those limits or, in unilateral abnormalities, by comparison with the normal side. Varus and valgus deformities (cubitus varus and cubitus valgus) are usually the result of trauma around the elbow. By far the best way to demonstrate a varus deformity is to ask the patient to lift his or her arms sideways to shoulder height; in this position the deformity becomes much more obvious, the arm taking on the appearance of a rifle butt (gunstock deformity, shown in Fig. 14.5).

Feel

Start by identifying the most obvious bony landmarks: the olecranon process posteriorly, the medial and lateral epicondyles and the head of the radius just distal to the lateral epicondyle; pronating and supinating the forearm makes it easier to find the mobile radial head and the lateral joint line. The ulna can be palpated throughout its length, the radius only at its proximal end and in the distal third of the forearm. The back of the elbow is palpated for warmth and swelling (signs of an olecranon bursitis) and subcutaneous nodules (a feature of rheumatoid arthritis). Feel more widely for synovial thickening and fluid (fluctuation on each side of the olecranon). The ulnar nerve is very superficial behind the medial condyle and here it can be rolled under the fingers to feel if it is thickened or hypersensitive. Last of all, feel for tenderness and try to determine which structure is affected.

Move

Active and passive flexion and extension are compared on the two sides. The elbow should be able to extend to the zero position (absolutely straight); people with lax joints can extend even beyond that point. As arough guide, people are normally able to flex the elbow sufficiently to touch the top of the shoulder with their fingers, but bear in mind that those with bulky upper arm muscles may not be able to do so. Pronation and supination of the forearms are tested with the patient holding the arms tucked into the waist and flexed to a right angle; 80–90 degrees each way is normal. *Stability* must also be tested carefully after trauma. The humerus is stabilized, the elbow is flexed to about 25 degrees to unlock any contribution to stability by the olecranon and the elbow is stressed in torsion and collateral stress.

General examination

Clinical examination should include the neck and shoulder (which are sources of referred pain to the elbow) and the hand (for signs of nerve dysfunction).

'PULLED ELBOW'

Downward dislocation of the head of the radius from the annular ligament is a fairly common injury in children under the age of 6 years. There may be a history of the child being jerked by the arm and subsequently complaining of pain and inability to use the arm. The limb is held more or less immobile with the elbow fully extended and the forearm pronated; any attempt to supinate the forearm is resisted. The diagnosis is essentially clinical, though x-rays are usually obtained in order to exclude a fracture. The radial head can be forcibly pulled out of the noose of the annular ligament only when the forearm is pronated; even then the distal attachment of the ligament is sometimes torn. If the history and clinical picture are suggestive, an attempt should be made to reduce the subluxation or dislocation. While the childs attention is diverted, the elbow is quickly supinated and then slightly flexed; the radial head is relocated

with a snap. (This sometimes happens ‘spontaneously’ while the radiographer is positioning the arm!)

RECURRENT ELBOW INSTABILITY

Following a dislocation or severe sprain, the lateral collateral ligament can be stretched or ruptured. The patient may present with painful clunking and locking. On examination, an apprehension response can be elicited by supinating the forearm while applying a valgus force to the elbow during flexion. The lateral collateral ligament can be directly repaired or reconstructed with a tendon autograft (e.g. palmaris longus). Medial instability is less frequent after trauma; a chronic instability can develop in javelin throwers and baseball players. Ligament reconstruction with a tendon graft and careful graduated rehabilitation can give very good results.

EPICONDALGIA

The elbow is prone to painful disorders of the tendon attachment. Sometimes this occurs spontaneously, sometimes after sudden unaccustomed use. These conditions have acquired names derived from the activities in which they were encountered when they were first described.

TENNIS ELBOW (LATERAL EPICONDALGIA)

Pain and tenderness over the lateral epicondyle of the elbow (or, more accurately, the bony insertion of the common extensor tendon) is a common complaint among tennis players - but even more common in non-players who perform similar activities involving forceful repetitive wrist extension. It is the extensor carpi radialis tendon (which automatically extends the wrist when gripping) which is pathological in tennis elbow (Fig. 14.16). Like supraspinatus tendinitis, it may result in small tears, fibrocartilaginous metaplasia, microscopic calcification and a painful vascular reaction in the tendon fibres close to the lateral epicondyle.

Clinical features

The patient is usually an active individual of 30 or 40 years. Pain comes on gradually, often after a period of unaccustomed activity involving forceful gripping and wrist extension. It is usually localized to the lateral epicondyle, but in severe cases it may radiate widely. It is aggravated by movements such as pouring out tea, turning a stiff doorhandle, shaking hands or lifting with the forearm pronated. Among tennis players it is usually blamed on faulty technique. The elbow looks normal, and flexion and extension are full and painless. Characteristically there is localized tenderness at or just below the lateral epicondyle; pain can be reproduced by passively stretching the wrist extensors (by the examiner acutely flexing the patient’s wrist with the forearm pronated) or actively by having the patient extend the wrist with the elbow straight. *X-ray* is usually normal, but occasionally shows calcification at the tendon origin.

Diagnosis

In patients with longstanding symptoms which do not respond to treatment, the possibility of a painful radial nerve entrapment (‘radial tunnel syndrome’) should be considered (see Chapter 11).

Treatment

Many methods of treatment are available but the benefits of most are unclear; it is well to remember that 90 per cent of ‘tennis elbows’ will resolve spontaneously within 6-12 months. The first step is to identify, and then restrict, those activities which cause pain. Modification of sporting style may solve the problem. A tennis elbow clasp is helpful. The role of physiotherapy and manipulation is uncertain. Injection of the tender area with corticosteroid and local anaesthetic relieves pain but is not curative.

OPERATIVE TREATMENT

Some cases are sufficiently persistent or recurrent for operation to be indicated. The origin of the common extensor muscle is detached from the lateral epicondyle. Additional procedures such as division of the orbicular ligament or removal of a ‘synovial fringe’ are sometimes advocated; they probably make very little difference to the outcome. Surgery is successful in about 85 per cent of cases.

GOLFER’S ELBOW (MEDIAL EPICONDYLITIS)

This is similar to tennis elbow but about three times less common. In this case it is the pronator origin that is affected. Often there is an associated ulnar nerve neuropathy. A medial collateral ligament injury should be excluded.

Treatment is the same as for lateral epicondylitis but the outcome of surgery seems less predictable. The abnormal tissue at the flexor‑pronator origin is excised, great care being taken to preserve the medial collateral ligament. The medial antebrachial cutaneous nerve must be respected during the skin incision to avoid a troublesome postoperative neuroma.

BASEBALL PITCHER’S ELBOW

Repetitive, vigorous throwing activities can cause damage to the bones or soft-tissue attachments around the elbow. Professional baseball players may develop hypertrophy of the lower humerus and incongruity of the joint, or loose-body formation and osteoarthritis. The junior equivalent (*‘little leaguer’s elbow’*) is due to partial avulsion of the medial epicondyle. The only remedy ‑ however grudginglycaccepted ‑ is to stay off baseball until the condition clears up completely.

JAVELIN THROWER’S ELBOW

The over-arm action employed by javelin throwerscmay avulse or cause impingement upon the tip of thecolecranon process. However, this sport (like othercthrowing sports) places huge strain on the medial collateralcligament which can become either acutelycinjured or chronically attenuated. There may also be symptoms of ulnar nerve impairment. The pain usually settles down after a period of rest and modification of activities. However, an attenuated medial collateral ligament may need reconstruction with a tendon graft.

AVULSION OF THE DISTAL TENDON OF BICEPS

The typical patient is a man of about 45 years who feels sudden pain and weakness at the front of the elbow after strenuous effort. Feel for the distal biceps tendon while the patient flexes the elbow against resistance (ask him to grip the desk or table as if to lift it; normally the biceps tendon stands out as a taut cord across the elbow crease). Loss of supination power with the elbow flexed (negating supinator muscle) is a good physical sign. The tendon may be partially or completely avulsed from its insertion into the bicipital tuberosity of the radius. The diagnosis is often missed because elbow flexion and supination, although weaker than normal, are preserved by brachialis and supinator action. MRI helps to confirm the diagnosis but must not delay surgical treatment. Clinical diagnosis should usually suffice.

<u>Treatment</u>

Operative repair is not always necessary; some patients are content to manage with slightly reduced elbow flexion: in time, the other elbow flexors will compensate (brachioradialis, brachialis). However, there will be a very obvious cosmetic defect and greatly reduced power of supination. For these reasons, many patients will choose repair. The best results are achieved by operation within 2 weeks, before the tendon retracts and the interosseous tunnel becomes occluded. A two-incision technique is recommended to avoid nerve damage and heterotopic ossification; tissue anchors or sutures-through-drillholes can be used to attach the

tendon to its insertion point. The results of early surgery and careful rehabilitation are usually very good.

BURSITIS

The olecranon bursa sometimes becomes enlarged as a result of *continual pressure* or *friction;* this used to be called 'student's elbow'. If the enlargement is a nuisance the fluid may be aspirated. The commonest non-traumatic cause is *gout*; there may be a sizeable lump with calcification on x-ray. In *rheumatoid arthritis*, also, the bursa may become enlarged, and sometimes nodules can be felt in the lump or just distal to it over the proximal ulna. In both conditions other joints are likely to be affected as well. A chronically enlarged bursa may prove a severe nuisance and need to be excised. However, wound healing can be a problem.

The wrist 15

The wrist and hand function together, for all practical purposes, as a single articulated unit. The hand would be unable to perform its range of complicated movements without the reciprocal movement, positioning and stabilizing action of the wrist. Loss of movement at the wrist limits the manipulative skill of the fingers and thumb; and pain in the wrist makes it impossible to grip or pinch with full strength. Disorders of the wrist and hand are often interrelated and therefore, in the clinical setting, these two units should be examined and analysed together. However, for the sake of emphasis, they are treated here in two separate chapters.

CLINICAL ASSESSMENT

SYMPTOMS

Pain may be localized to the radial side (especially in de Quervain's disease and thumb base arthritis), to the ulnar side (e.g. in distal radio-ulnar joint arthritis and piso-triquetral arthritis) or to the dorsum (in radio-carpal arthritis, Kienbocks disease and occult dorsal wrist ganglion).

Stiffness is often not noticed until it is severe in the flexion-extension plane; loss of rotation is noticed earlier and can be very disrupting.

Swelling may signify involvement of either the joint or the tendon sheaths or a ganglion.

Deformity is a late symptom except after trauma or radial nerve palsy. Ask if it is localized to a particular site (e.g. an overly-prominent head of ulna, suggesting subluxation of the distal radio-ulnar joint) or involving the posture of the wrist as a whole [progressive radial deviation in advanced rheumatoid arthritis (RA)].

Loss of function refers mainly to the hand, though the patient may be aware that the problem lies in the wrist.

Clicks are common and usually of no relevance; *clunks* with pain or weakness may signify instability.

SIGNS

Examination of the wrist is not complete without also examining the elbow, forearm and hand. Both upper limbs should be completely exposed.

Look

The skin is inspected for scars. Both wrists and forearms are compared to see if there is any deformity. If there is swelling, note whether it is diffuse or localized to one of the tendon sheaths. Look also at the hands and fingers to see if there are any related abnormalities. The posture of the wrist at rest and during movement varies with different positions of the hand and fingers. This is discussed in the opening sections of Chapter 16.

Feel

Palpation of the wrist will yield valuable information only if the surface anatomy is thoroughly understood (see Figure 15.1). Tender areas must be accurately localized and the various landmarks compared with those of the normal wrist. The site of tenderness may be diagnostic, for example in de Quervain's disease (tip of radial styloid), scaphoid fracture (anatomical snuffbox), carpo-metacarpal osteoarthritis (base of first metacarpal), Kienbocks disease (over the lunate), triangular fibrocartilage complex (just distal to the head of the ulna) and localized tenosynovitis of any of the wrist tendons. At the same time note if the skin feels unduly warm. If the head of the ulna seems abnormally prominent on the dorsum of the wrist, try to jar the distal radioulnar joint by pressing down sharply on the ulnar prominence; if it moves up and down the joint is unstable (this is aptly named the piano-key sign).

Move

Passive movements To compare passive dorsiflexion of the wrists the patient places his palms together in the position of prayer, then elevates his elbows. Palmar flexion is examined in a similar way. Radial and ulnar deviation are measured in either the palms-up or the palms-down position. With the elbows at right angles and tucked in to the sides, pronation and supination are assessed. While testing passive movements, the presence of abnormal 'clunks' should be noted; they may signify one or other form of carpal instability.

Active movements Ask the patient to pull the hand backwards to its limit (*extension)*, then forwards as far as possible (*flexion*), and then sideways to right and left (*radial and ulnar deviation*). Active *pronation and supination* should be performed with the patient's elbows tucked tightly into the waist. These movements are then repeated but carried out against resistance, to test for muscle power. Finally, grip strength is measured, preferably using a mechanical dynamometer. Loss of power may be due to pain, tendon rupture or muscle weakness.

Provocative tests

Special tests are needed to assess stability of the carpal articulations. The *luno-triquetral joint* is tested by gripping or pinching the lunate with one hand, the triquetral-pisiform with the other, and then applying a sheer stress: pain or clicking suggests an incompetent luno-triquetral ligament. The *piso-triquetral joint* is tested by pushing the pisiform radialwards against the triquetrum. Stability of the *scapho-lunate joint* is tested by pressing hard on the palmar aspect of the scaphoid tubercle while moving the wrist alternately in abduction and adduction: pain or clicking on abduction (radial deviation) is abnormal. The *triangular fibrocartilage* is tested by pushing the wrist medially then flexing and extending it. The *distal radio-ulnar joint* is tested for stability by holding the radius and then ballotting the ulnar head up and down. These tests are mentioned again in the section on carpal instability.

CHRONIC INSTABILITY OF THE WRIST

Movements of the wrist and hand are interdependent, the wrist providing appropriate mobility and stability to position and steady the hand for the remarkable range of actions and tactile sensibility employed in our daily activities. Abnormalities of wrist mechanics are a

common source of functional disability; this is seen most often in rheumatoid arthritis, in association with congenital laxity and after local trauma.

Articulations of the wrist

The wrist comprises three movable joints: the *distal radio-ulnar joint,* the *radio-carpal joint* (between the radius and the proximal row of carpal bones) and the *midcarpal joint* (between the proximal and distal rows of carpal bones).

THE DISTAL RADIO-ULNAR JOINT (DRUJ)

The distal radius and ulna are linked to each other by the interosseous membrane, the capsule of the DRUJ and *the triangular fibrocartilage complex (TFCC).*

The head of the ulna articulates congruently with the sigmoid notch of the distal radius; movement at the joint occurs by the radius both rotating and sliding in an arc around the head of the ulna during pronation and supination of the forearm. Interposed between the head of the ulna and the carpus is a fibrocartilaginous disc, a fan-shaped structure spreading from an apical attachment at the base of the ulnar styloid process to the rim of the radial sigmoid notch. Its dorsal and volar edges are coextensive with the dorsal and palmar radio-ulnar ligaments; further attachments to the joint capsule, the ulno-triquetral and ulno-lunate ligaments, the ulnar collateral ligament and the sheath of the extensor carpi ulnaris tendon complete the fibrocartilage complex. The peripheral attachments of the TFCC have a good vascular supply and can heal after injury; the central area of the triangular plate is avascular and tears do not heal.

KIENBÖCK'S DISEASE

Robert Kienbock, in 1910, described what he called 'traumatic softening' of the lunate bone. This is a form of ischaemic necrosis, probably due to chronic stress or injury, though one cannot be certain about this. It has been suggested that relative shortening of the ulna ('negative ulnar variance') predisposes to stress overload of the lunate between the distal edge of the radius and the carpus, but this has not been proven convincingly.

Pathology

As in other forms of ischaemic necrosis, the pathological changes proceed in four stages: *stage 1*, ischaemia without naked-eye or radiographic abnormality; *stage 2,* trabecular necrosis with reactive new bone formation and increased radiographic density, but little or no distortion of shape; *stage 3,* collapse of the bone; and *stage 4,* disruption of radio-carpal congruence and secondary osteoarthritis.

Clinical features

The patient, usually a young adult, complains of ache and stiffness; only occasionally is there a history of acute trauma. Tenderness is localized over the lunate and grip strength is diminished. In the later stages wrist movements are limited and painful.

Imaging

X-rays at first show no abnormality, but radioscintigraphy may reveal increased activity. Later, x-rays may show either mottled or diffuse density of the bone, and later still the bone looks intensely sclerotic and irregular in shape or squashed. The capitate migrates proximally into the space left by the collapsing lunate and the scaphoid flexes forward. Eventually, there are osteoarthritic changes in the wrist. Ulnar variance should be assessed by standardized x-ray examination with the shoulder abducted to 90 degrees, the forearm in neutral rotation and the wrist in neutral flexion-extension. As the lunate collapses, the relative length of the capitate from third metacarpal bone to distal radius increases.

MRI is the most reliable way of detecting the early changes. A gadolinium-enhanced MRI scan will demonstrate the condition even if plain x-rays are normal.

Treatment

NON-OPERATIVE TREATMENT In early cases, splintage of the wrist for 6–12 weeks relieves pain and possibly reduces mechanical stress. If bone healing catches up with ischaemia, the lunate may remain virtually undistorted; this is more likely in very young patients. However, if pain persists, and even more so if the bone begins to flatten, operative treatment is indicated.

OPERATIVE TREATMENT

In its earliest stages, before collapse, the bone can be revascularized with a pedicled bone graft or vascular bundle implantation.

TENOSYNOVITIS AND TENOVAGINITIS

The extensor retinaculum has six compartments which transmit tendons lined with synovium. Tenosynovitis can be caused by unaccustomed overuse but sometimes it occurs spontaneously. The resulting synovial inflammation causes secondary thickening of the sheath and stenosis of the compartment, which further compromises the tendon. Early treatment, including rest, anti-inflammatory medication and injection of corticosteroids, may break this vicious circle. The first dorsal compartment (abductor pollicis longus and extensor pollicis brevis) and the second dorsal compartment (extensor carpi radialis brevis) are most commonly affected. The flexor tendons are affected far less frequently.

DE QUERVAIN'S DISEASE

Pathology

This condition, first described in 1895, is caused by reactive thickening of the sheath around the extensor pollicis brevis and abductor pollicis longus tendons within the first extensor compartment. It may be initiated by overuse but it also occurs spontaneously, particularly in middle-aged women, and sometimes during pregnancy.

Clinical features The patient is usually a woman aged 40–50, who complains of pain on the radial side of the wrist. There may be a history of unaccustomed activity such as pruning roses or wringing out clothes. Sometimes there is a visible swelling over the radial styloid and the tendon sheath feels thick and hard. Tenderness is most acute at the very tip of the radial styloid. The pathognomonic sign is elicited by *Finkelstein's test*. The examiner places the patient's thumb across the palm in full flexion and then, holding the patients hand firmly, turns the wrist sharply into adduction. In a positive test this is acutely painful; repeating the movement with the thumb left free is relatively painless. Resisted thumb extension (hitch-hikers sign) is also painful.

The *differential diagnosis* includes arthritis at the base of the thumb, scaphoid non-union and the intersection syndrome (see below).

Treatment

The early case can be relieved by a corticosteroid injection into the tendon sheath, sometimes combined with hand therapy (ultrasound, frictions, splintage). Resistant cases need an operation, which consists of slitting the thickened tendon sheath. Sometimes there is duplication of tendons and even of the sheath, in which case both sheaths need to be divided. Care should be taken to prevent injury to the dorsal sensory branches of the radial nerve, which may cause intractable dysaesthesia.

GANGLION CYSTS

Pathology

The ganglion cyst is the most common swelling in the wrist. It arises from leakage of synovial fluid from a ulnaris joint or tendon sheath and contains a glairy, viscous fluid. Although it can

appear anywhere around the carpus, it usually develops on the dorsal surface of the scapho-lunate ligament. Palmar wrist ganglia usually arise from the scapho-lunate or scapho trapeziotrapezoid joint.

Clinical features

The patient, often a young adult, presents with a painless lump, though occasionally there is slight ache and weakness. The lump is well defined, cystic and not tender; it can sometimes be transilluminated. It does not move with the tendons. The back of the wrist is the commonest site; less frequently a ganglion emerges alongside the radial artery on the volar aspect. Occasionally a small, hidden ganglion is found to be the cause of compression of the deep (muscular) branch of the ulnar nerve.

Treatment

Treatment is usually unnecessary. The lump can safely be left alone; it often disappears spontaneously. However, it can be aspirated to reassure the patient. If it becomes troublesome – and certainly if there is any pressure on a nerve – operative removal is justified. Even then it may recur with embarrassing persistence; it is not easy to ensure that every shred of abnormal tissue is removed.

The hand 16

MUSCLE CONTRACTURE

VOLKMANN'S ISCHAEMIC CONTRACTURE

Contracture of the forearm muscles may follow circulatory insufficiency due to injuries at or below the elbow. Shortening of the long flexors causes the fingers to be held in flexion; they can be straightened only when the wrist is flexed so as to relax the long flexors. Sometimes the picture is complicated by associated damage to the ulnar or median nerve (or both). If disability is marked, some improvement may be obtained by lengthening the shortened tendons, or else by excising the fibrosed muscles and restoring finger movement with tendon transfers.

SHORTENING OF THE INTRINSIC MUSCLES

Shortening of the intrinsic muscles in the hand produces a characteristic deformity: flexion at the MCP joints with extension of the IP joints and adduction of the thumb (the so-called *'intrinsic-plus' hand*). Slight degrees of deformity may not be obvious, but can be diagnosed by *Bunnell's 'intrinsic-plus' test*: with the MCP joints pushed passively into hyperextension (thus putting the intrinsics on stretch), it is difficult or impossible to flex the IP joints passively; if the MCP joints are then placed in flexion, the IP joints can be passively flexed. The causes of intrinsic shortening or contracture are: (1) spasticity (e.g. in cerebral palsy); (2) volar subluxation of the MCP joints (e.g. in rheumatoid arthritis); (3) scarring after trauma or infection; and (4) shrinkage due to ischaemia. Moderate contracture can be treated by resecting a triangular segment of the intrinsic 'aponeurosis' at the base of the proximal phalanx (Littler's operation).

TENDON LESIONS

MALLET FINGER

This results from injury to the extensor tendon of the terminal phalanx. It may be due to direct trauma but more often painlessly follows an innocent event when the finger tip is forcibly bent during active extension, perhaps while tucking the blankets under a mattress or trying to catch a ball. The terminal joint is held flexed and the patient cannot straighten it, but passive

movement is normal. With the extensor mechanism unbalanced, the PIP joint may become hyperextended (‘swan-neck’). X-rays are taken to show or exclude a fracture. If there is a fracture but minimal subluxation of the joint, it is treated by splintage with the DIP joint in extension for 6 weeks. Operative treatment is considered only if there is a large fragment (>50 per cent) and subluxation of the DIP joint. Otherwise surgery is ill advised, as the complication rate is high and it is unlikely to improve the outcome. A mallet finger without bone injury is treated with a plastic splint with the DIP joint in extension for 8 weeks, followed by 4 weeks of night splintage. This treatment may still work if presentation is delayed for a few weeks. The great majority do very well. Old lesions need treatment only if the deformity is marked and hand function seriously impaired. The options include fusion for painful arthritic joints or tendon reconstruction.

RUPTURED EXTENSOR POLLICIS LONGUS

The long thumb extensor may rupture after fraying or ischaemia where it crosses the wrist (e.g. after a Colles’ fracture, or in rheumatoid arthritis). The distal phalanx drops into flexion; it can be passively extended, and there may still be weak active extension because of thenar muscle insertion into the extensor expansion; however, the thumb cannot be actively elevated backwards above the plane of the hand (retroposition). Direct repair is unsatisfactory and a tendon transfer, using the extensor indicis, is needed. The results are, in over 90 per cent of cases, satisfactory.

DROPPED FINGER

Sudden loss of finger extension at the MCP joint is usually due to tendon rupture at the wrist (e.g. in rheumatoid arthritis). Because direct repair is not usually possible, the distal portion can be attached to an adjacent finger extensor or a tendon transfer performed. Occasionally the deformity is due to catching of the collateral ligament on a metacarpal osteophyte or rupture of the sagittal band which centralizes the tendon over the back of the knuckle.

BOUTONNIRE DEFORMITY

This lesion presents as a flexion deformity of the PIP joint and extension of the DIP joint. It is due to interruption or stretching of the central slip of the extensor tendon where it inserts into the base of the middle phalanx**.** The lateral slips separate and the head of the proximal phalanx thrusts through the gap like a button through a buttonhole. Ironically while English speakers call it a *‘boutonniere’* deformity, the French refer to it as *‘le buttonhole’*.The usual causes are direct trauma or rheumatoid disease. Initially the deformity is slight and passively correctable; later the soft tissues contract, resulting in fixed flexion of the proximal and hyperextension of the DIP joint. Early diagnosis is therefore important; an impending deformity should be suspected in anyone with tenderness or a cut over the dorsum of the PIP joint, especially if they cannot actively extend the IP joint with the MCP joints and wrist flexed. In the early post-traumatic case, splinting the PIP joint in full extension for 6 weeks usually leads to healing; the DIP joint must be moved passively to prevent the lateral bands from sticking. Open injuries of the central slip should be repaired, with the joint protected by a K-wire for 3 weeks. For later cases where the joint is still passively correctible, several operations have been invented (suggesting that none is too reliable). The easiest and probably most successful procedure is to divide the extensor tendon just proximal to its insertion into the distal phalanx. This allows the extensor mechanism to move proximally, thus enhancing PIP extension and diminishing DIP extension. Longstanding fixed deformities are extremely difficult to correct and may be better left alone.

SWAN-NECK DEFORMITY

This is the reverse of the boutonniere deformity; the PIP joint is hyperextended and the DIP joint flexed. The deformity can be reproduced voluntarily by laxjointed individuals. The clinical disorder has many causes, with two things in common: imbalance of extensor versus flexor action at the PIP joint and laxity of the palmar plate. Thus it may occur: (1) if the PIP extensors overact (e.g. due to intrinsic muscle spasm or contracture, after mallet finger, or following volar subluxation of the MCP joint); (2) if the PIP flexors are inadequate (inhibition or division of the flexor superficialis); or (3) if the palmar plate of the PIP joint fails (in rheumatoid arthritis, lax-jointed individuals or trauma). If the deformity is allowed to persist, secondary contracture of the intrinsic muscles, and eventually of the PIP joint itself, makes correction increasingly difficult and ultimately impossible. Treatment depends on the cause and whether or not the deformity has become fixed. If the deformity corrects passively, then a simple figure-of-eight ring splint to maintain the PIP joint in a few degrees of flexion may be all that is required; if this works but cannot be tolerated, then tenodesis of the PIP joint works well. The options are either to attach one slip of flexor digitorum superficialis to the proximal phalanx, which prevents hyperextension, or to re-route a lateral band anteriorly so it becomes a flexor rather than an extensor of the PIP joint. If the intrinsics are tight they are released. If the deformity is fixed, then it may respond to gentle manipulation supplemented by temporary Kwire fixation in a few degrees of flexion; if not, then lateral band release from the central slip may be needed. The dorsal skin may not close directly after correction. If the swan-neck deformity is secondary to a mallet finger, then the latter should be addressed as described above. If function is severely impaired and does not respond to one of the above measures, the joint is arthrodesed in a more acceptable position.

DUPUYTREN'S CONTRACTURE

This is a nodular hypertrophy and contracture of the superficial palmar fascia (palmar aponeurosis). The condition is inherited as an autosomal dominant trait and is most common in people of European (especially Anglo-Saxon) descent. It is more common in males than females; the prevalence increases with age, but onset at an early stage usually means aggressive disease. There is a high incidence in epileptics receiving phenytoin therapy; associations with diabetes, smoking, alcoholic cirrhosis, AIDS and pulmonary tuberculosis have also been described. There is a contentious and weak association with injury to the hand.

PATHOLOGY

The essential problem in Dupuytren's disease is proliferation of myofibroblasts; where they come from and why they proliferate remains unclear. After an initial proliferative phase, fibrous tissue within the palmar fascia and fascial bands within the fingers contracts, causing flexion deformities of the MCP and PIP joints. Fibrous attachments to the skin lead to puckering. The digital nerve is displaced or enveloped, but not invaded, by fibrous tissue. Occasionally the plantar aponeurosis also is affected.

Clinical features

The patient – usually a middle-aged man – complains of a nodular thickening in the palm. Gradually this extends distally to involve the ring or little finger. Pain may occur early on but is seldom a marked feature. Often both hands are involved, one more than the other. The palm is puckered, nodular and thick. If the subcutaneous cords extend into the fingers they may produce flexion deformities at the MCP and PIP joints. Sometimes the dorsal knuckle pads are thickened (Garrod's pads). About 60 per cent of patients give a family history. Similar nodules may be seen on the soles of the feet (Ledderhose's disease). There is a rare, curious association with fibrosis of the corpus cavernosum (Peyronies disease).

Diagnosis

Dupuytren's contracture must be distinguished from skin contracture (where the previous laceration is usually obvious), tendon contracture (in which the finger deformity changes with wrist position) and PIP joint contracture (in which there may be a history of clinodactyly or joint injury).

Treatment

Operation is indicated if the deformity is a nuisance or rapidly progressing. In particular, PIP joint contractures can become irreversible. The aim is reasonable, not complete, correction. Surgery does not cure the disease, it only partially corrects the deformity, and recurrence or extension is common. Correction of the MCP joint is more predictable than the PIP joint. Only the thickened part of the fascia is excised (complete fasciectomy is usually unnecessary). An isolated cord across the front of the MCP joint can be managed by dividing the contracture under local anaesthetic with a bevelled needle ('needle fasciotomy'). If the disease is more extensive, the affected area is approached through a longitudinal or a Z-shaped incision and, after carefully freeing the nerves and blood vessels, the cords are excised. Skin closure may be facilitated by multiple Z-plasties. This has the dual effect of improving the deformity and, if recurrence occurs, preventing a longitudinal wound contracture. The palmar section of the wound can be left open; it will soon heal with dressings. This makes skin closure easier and allows any haematoma (which may predispose to recurrence) to escape. After operative correction a splint is applied, and removed after a few days for active motion exercises. Night splinting for a few months may reduce recurrence. If there is severe skin involvement (particularly in surgery for recurrent disease), if there is a strong family history, or if the patient is particularly young, then skin grafting should be considered. Amputation or joint fusion is occasionally advisable for severe, recurrent disease in the little finger.

TRIGGER FINGER (DIGITAL TENOVAGINOSIS)

A flexor tendon may become trapped by thickening at the entrance to its sheath; on forced extension it passes the constriction with a snap ('triggering'). A secondary nodule can develop on the tendon. The underlying cause is unknown but the condition is certainly more common in patients with diabetes. People with rheumatoid disease may develop synovial thickening or intratendinous nodules which can also cause triggering. Occupational factors, though sometimes blamed, are unlikely to be causative.

Clinical features

Any digit may be affected, but the thumb, ring and middle fingers most commonly; sometimes several fingers are affected. The patient notices a click as the finger is flexed; when the hand is unclenched, the affected finger initially remains bent at the PIP joint but with further effort it suddenly straightens with a snap. A tender nodule can be felt in front of the MCP joint and the click may be reproduced at this site by alternately flexing and extending the finger.

INFANTILE TRIGGER THUMB

Parents sometimes notice that their baby or infant cannot extend the thumb tip. The diagnosis is often missed, or the condition is wrongly taken for a 'dislocation'. Very occasionally the child grows up with the thumb permanently bent. This condition must be distinguished from the rare *congenitally clasped thumb* in which both the IP joint and the MCP joint are flexed because of congenital insufficiency of the extensor mechanism (see Chapter 15).

Treatment

In adults, early cases may be cured by an injection of corticosteroid carefully placed at the mouth of the tendon sheath. Recurrent triggering up to 6 months later occurs in over 30 per cent of patients – particularly younger patients and those with diabetes, who may then need a second injection. Refractory cases need operation, through an incision over the distal

palmar crease, or in the MCP crease of the thumb – the A1 section of the fibrous sheath is incised until the tendon moves freely. In babies it is worth waiting until the child is about 3 years old, as spontaneous recovery often occurs. If not, then the pulley is released. Care should be taken to avoid injury to the digital neurovascular bundles during surgery. The risk is greatest in the thumb (where the nerves are close to the midline) and the index finger (where the radical digital nerve crosses the tendon). In patients with rheumatoid arthritis the fibrous pulley must be carefully preserved; damage to this structure will predispose to ulnar deviation of the fingers. Flexor synovectomy with excision of one slip of flexor digitorum superficialis is preferred.

ACUTE INFECTIONS OF THE HAND

Infection of the hand is frequently limited to one of several well-defined compartments: under the nail-fold (paronychia); the pulp space (felon) and in the subcutaneous tissues elsewhere; the deep fascial spaces; tendon sheaths; and joints. Usually the cause is a staphylococcus which has been implanted during fairly trivial injury. However, cuts contaminated with unusual organisms account for about 10 per cent of cases.

Pathology

Here, as elsewhere, the response to infection is an acute inflammatory reaction with oedema, suppuration and increased tissue tension. In closed tissue compartments (e.g. the pulp space or tendon sheath) pressures may rise to levels where the local blood supply is threatened, with the risk of tissue necrosis. In neglected cases infection can spread from one compartment to another and the end result may be a permanently stiff and useless hand. There is also a danger of lymphatic and haematogenous spread; even apparently trivial infections may give rise to lymphangitis and septicaemia.

Clinical features

Usually there is a history of trauma (a superficial abrasion, laceration or penetrating wound), but this may have been so trivial as to pass unnoticed. A few hours or days later the finger or hand becomes painful and swollen. There may be throbbing and sometimes the patient feels ill and feverish. Ask if he or she can recall any causative incident: a small cut or superficial abrasion, a prick injury (including plant thorns) or a local injection. Also, do not forget to enquire about predisposing conditions such as diabetes mellitus, intravenous drug abuse and immunosuppression. On examination the finger or hand is red and swollen, and usually exquisitely tender over the site of tension. However, in immune-compromised patients, in the very elderly and in babies, local signs may be mild. With superficial infection the patient can usually be persuaded to flex an affected finger; with deep infections active flexion is not possible. The arm should be examined for lymphangitis and swollen glands, and the patient more generally for signs of septicaemia. X-ray examination may disclose a foreign body but is otherwise unhelpful in the early stages of infection. However, a few weeks later there may be features of osteomyelitis or septic arthritis, and later still of bone necrosis. If pus becomes available, this should be sent for bacteriological examination.

Diagnosis

In making the diagnosis, several conditions must be excluded: an *insect bite or sting* (which can closely mimic a subcutaneous infection), a *thorn prick* (which, itself, can become secondarily infected), acute *tendon rupture* (which may resemble a septic tenosynovitis) and *acute gout* (which is easily mistaken for septic arthritis). *Plant-thorn injuries* are extremely common and the distinction between secondary infection and a nonseptic reaction to a retained fragment can be difficult. Rose thorn and blackthorn are the usual suspects in the UK, but any plant spine (including cactus needles) can be implicated. The local inflammatory response sometimes leads to recurrent arthritis or tenosynovitis, which is arrested only by removing the retained fragment. If the condition is suspected, the fragment may be revealed by ultrasound scanning or MRI. Secondary infection with unusual soil or plant organisms

may occur.

Principles of treatment
Superficial hand infections are common; if their treatment is delayed or inadequate, infection may rapidly extend, with serious consequences. The essentials of treatment are:
· antibiotics
· rest, splintage and elevation
· drainage
· rehabilitation.

NAIL-FOLD INFECTION (PARONYCHIA)
Infection under the nail-fold is the commonest hand infection; it is seen most often in children, or in older people after rough nail-trimming. The edge of the nail-fold becomes red and swollen and increasingly tender. A tiny abscess may form in the nail-fold; if this is left untreated, pus can spread under the nail. At the first sign of infection, treatment with antibiotics alone may be effective. However, if pus is present it must be released by an incision at the corner of the nail-fold in line with the edge of the nail; a pledget of paraffin gauze is used to keep the nail-fold open. If pus has spread under the nail, part or all of the nail may need to be removed.
Chronic paronychia Chronic nail-fold infection may be due to (1) inadequate drainage of an acute infection, or (2) a fungal infection, which requires specific treatment. Topical or oral antifungal agents are used to eradicate fungal infection, but failing this, or for chronic bacterial infection, the nail bed may have to be laid open (‘marsupialized’); care should be taken to avoid damaging the germinal nail matrix.

TENDON SHEATH INFECTION
(SUPPURATIVE TENOSYNOVITIS)
The tendon sheath is a closed compartment extending from the distal palmar crease to the DIP joint. In the thumb and fifth finger, the sheaths are co-extensive with the radial and ulnar bursae, which envelop the flexor tendons in the proximal part of the palm and across the wrist; these bursae also communicate with Parona’s space in the lower forearm. Pyogenic tenosynovitis is uncommon but dangerous. It usually follows a penetrating injury, the commonest organism being *Staphylococcus aureus*; however, streptococcus and Gram-negative organisms are also encountered. The affected digit is painful and swollen; it is usually held in slight flexion, is very tender, and the patient will not move it or permit it to be moved. Early diagnosis is based on clinical findings; x-rays are unhelpful but ultrasound scanning may be useful. Delayed diagnosis results in a progressive rise in pressure within the sheath and a consequent risk of vascular occlusion and tendon necrosis. In neglected cases infection may spread proximally within the radialor ulnar bursa, or from one to the other (*a ‘horse-shoe’ abscess*); it can also spread proximally to the flexor compartment at the wrist and into Parona’s space in the forearm. Occasionally this results in median nerve compression.
Treatment Treatment must be started as soon as the diagnosis is suspected. The hand is elevated and splinted and antibiotics are administered intravenously‐ideally a broad-spectrum penicillin or a systemic cephalosporin. If there is no improvement after 24 hours, surgical drainage is essential. Two incisions are needed, one at the proximal end of the sheath and one at the distal end; using a fine catheter, the sheath is then irrigated (always from

proximal to distal) with Ringer’ s lactate solution. Additional, proximal, incisions may be needed if the synovial bursae are infected. Postoperatively the hand is swathed in absorbent dressings and splinted in the position of safe immobilization. The dressings should not be too bulky, as this will make it difficult to ensure correct positioning of the joints. The flexor sheath catheter is left in place; using a syringe, the sheath is irrigated with 20 mL of saline three or four times a day for the next 2 days. The catheter and dressings are then removed and finger movements are started. Stiffness is a very real risk and so early supervised hand therapy must be arranged.

BITES

ANIMAL BITES

Animal bites are usually inflicted by cats, dogs, farm animals or rodents. Many become infected and, although the common pathogens are staphylococci and streptococci, unusual organisms like *Pasteurella multocida* are often reported.

HUMAN BITES

Human bites are generally thought to be even more prone to infection. A wide variety of organisms (including anaerobes) are encountered, the commonest being *Staphylococcus aureus*, *Streptococcus* Group A and *Eikenella corrodens*. Bites can involve any part of the hand, fingers or thumb; tell-tale signs of a human bite are lacerations on both volar and dorsal surfaces of the finger. Often, though, the ‘bite’ consists only of a dorsal wound over one of the MCP knuckles, sustained during a fistfight.

All such wounds should be assumed to be infected. Moreover, it should be remembered that a laceration of the clenched fist may have penetrated the extensor apparatus and entered the MCP joint; this will not be apparent if the wound is examined with the fingers in extension because the extensor hood and capsule will have retracted proximally.X-rays should be obtained (to exclude a fracture, tooth fragment or foreign body) and swabs taken for bacterial culture and sensitivity.

Treatment

Fresh wounds should be carefully examined in the operating theatre and, if necessary, extended and debrided. Search for a fragment of tooth or - with a knuckle bite - for a *divot* of articular cartilage from the joint. The hand is splinted and elevated and antibiotics are given prophylactically until the laboratory results are obtained. Infected bites will need debridement, wash-outs and intravenous antibiotic treatment. The common infecting organisms are all sensitive to broad-spectrum penicillins (e.g. amoxicillin with clavulanic acid) and cephalosporins. With animal bites one should also consider the possibility of rabies. Postoperative treatment consists, as usual, of copious wound dressings, splintage in the ‘safe ’ position and encouragement of movement once the infection has resolved. Tendon lacerations can be dealt with when the tissues are completely healed.

RAYNAUD’S DISEASE

Raynaud’s syndrome is produced by a vasospastic disorder which affects mainly the hands and fingers. Attacks are usually precipitated by cold; the fingers go pale and icy, then dusky blue (or cyanotic) and finally red. Between attacks the hands look normal. The condition is most commonly seen in young women who have no underlying or predisposing disease. *Raynaud’s phenomenon* is the term applied when these changes are associated with an underlying disease such as scleroderma or arteriosclerosis. Similar, though milder, changes are also seen in thoracic outlet syndrome. The hands must be kept warm. Calcium channel blockade, iloprost infusions or digital sympathectomy (surgical removal of the sympathetic plexus around the digital arteries) may be needed.

HAND–ARM VIBRATION SYNDROME

Excessive and prolonged use of vibrating tools can damage the nerves and vessels in the fingers. The damage is proportional to the duration of exposure and amount of vibration. There are two components: vascular and neurological. The *vascular component* is similar to Raynaud's phenomenon, with the fingertips turning white in cold weather, then changing through blue and red as the circulation is restored. The *neurological component* involves numbness and tingling in the finger-tips. In advanced cases there can be reduced dexterity. Some patients have clear carpal tunnel syndrome as well.

Treatment is generally unsatisfactory, but includes avoidance of cold weather and smoking as well as, of course, vibrating tools. Carpal tunnel syndrome associated with vibration, in the absence of a more diffuse neuropathy, responds fairly well to standard decompression.

ULNAR ARTERY THROMBOSIS

Repeated blows to the hand, especially using the hypothenar eminence as a hammer, can damage the intima of the ulnar artery, leading to either thrombosis or an aneurysm. The patient presents with cold intolerance in the little finger. Microvascular reconstruction of the ulnar artery is needed.

The neck 17

SYMPTOMS

Pain is felt in the neck itself, but it may also be referred to the shoulders or arms. If it starts suddenly after exertion, and is exaggerated by coughing or straining, think of a disc prolapse. Pain spreading down an arm and forearm with paraesthesiae in the hand will strengthen the likelihood of a disc prolapse with cervical root compression. Chronic or recurrent pain in older people is usually due to chronic disc degeneration and spondylosis. Always enquire if any posture or movement makes it worse; or better.

Stiffness may be either intermittent or continuous. Sometimes it is so severe that the patient can scarcely move the head.

Deformity usually appears as a wry neck; occasionally the neck is fixed in flexion.

Numbness, tingling and weakness in the upper limbs may be due to pressure on a nerve root; weakness in the lower limbs may result from cord compression in the neck.

Headache sometimes emanates from the neck, especially occipital headache, but if this is the only symptom other causes should be suspected.

'Tension' is often mentioned as a cause of neck pain and occipital headache. The neck and back are common 'target zones' for psychosomatic illness.

DEFORMITIES OF THE NECK IN CHILDREN

A variety of deformities are encountered, some reflecting postural adjustments to underlying disorders and others due to developmental anomalies.

TORTICOLLIS

This is a description rather than a diagnosis. The chin is twisted upwards and towards one side. There are many causes. The condition may be either *congenital* or *acquired*.

Infantile (congenital) torticollis

This condition is common. The sternomastoid muscle on one side is fibrous and fails to elongate as the child grows; consequently, progressive deformity develops. The cause is unknown; the muscle may have suffered ischaemia from a distorted position in utero (the association with breech presentation and hip dysplasia is supporting evidence), or it may have been injured at birth. A history of difficult labour or breech delivery is common. A lump may be noticed in the first few weeks of life; it is well defined and involves one or both heads of the sternomastoid. At this stage there is neither deformity nor obvious limitation of movement and within a few months the lump has disappeared. Deformity does not become apparent until the child is 1–2 years old. The head is tilted to one side, so that the ear approaches the shoulder; the sternomastoid on that side may feel tight and hard. There may also be asymmetrical development of the face (plagiocephaly). These features become increasingly

obvious as the child grows. Other causes of wry neck (bony anomalies, discitis,lymphadenitis) should be excluded. The history and the typical facial appearance are helpful clues. Radiographs must be taken to exclude a bone abnormality or fracture.

Treatment If the diagnosis is made during infancy, daily muscle stretching by the parents may prevent the incipient deformity. Non-operative treatment is successful in most cases. If the condition persists beyond one year, operative correction is required to avoid progressive facial deformity. The contracted muscle is divided (usually at its lower end but sometimes at the upper end or at both ends) and the head is manipulated into the neutral position. After operation, correction must be maintained, with a temporary rigid orthosis followed by stretching exercises.

Secondary torticollis

Childhood torticollis may be secondary to congenital bone anomalies, atlanto-axial rotatory displacement, infection (lymphadenitis, retropharyngeal abscess, tonsillitis, discitis, tuberculosis), trauma, juvenile rheumatoid arthritis, posterior fossa tumours, intraspinal tumours, dystonia (benign paroxysmal torticollis) or ocular dysfunction.

Atlanto-axial rotatory displacement The aetiology of this condition is unclear, but it is thought to be due to muscle spasm resulting from inflammation of the ligaments, capsule and synovium of the atlanto-axial region. There may be a history of trauma or upper **17** respiratory tract infection. The child presents with a *painful wry neck.* Plain x-rays are difficult to interpret; a CT scan in both neutral and maximum lateral rotation is the most helpful investigation. Most cases are mild and can be managed expectantly with a soft collar and analgesics. If there is no resolution after a week, halter traction, bed rest and analgesics should be prescribed. In more resistant cases, halo traction may be required. Occasionally there is anterior displacement of C1 on C2; the articulation may not stabilize following traction and a C1/2 fusion is then indicated.

ACUTE INTERVERTEBRAL DISC PROLAPSE

Acute disc prolapse is not as common in the neck as in the lower back; both segments of the spine are mobile but the mechanical environment in the cervical region is more favourable than that in the lumbosacral region. The pathological features are similar; these are described in some detail in Chapter 18.

The acute prolapse may be precipitated by local strain or injury, especially sudden unguarded flexion and rotation, and usually occurs immediately above or below the sixth cervical vertebra. In many cases (perhaps in all) there is a predisposing abnormality of the disc with increased nuclear tension. Prolapsed material may press on the posterior longitudinal ligament or dura mater, causing neck pain and stiffness as well as pain referred to the upper limb. Pressure on the nerve roots causes paraesthesia, and sometimes weakness, in one or both arms – usually in the distribution of C6 or C7.

Differential diagnosis

Acute soft-tissue strain Acute strains of the neck are often associated with pain, stiffness and vague 'tingling' in the upper limbs. It is important to bear in mind that pain radiating into the arm is not necessarily due to nerve root pressure.

Neuralgic amyotrophy This condition can closely resemble an acute disc prolapse and should always be thought of if there is no definite history of a strain episode. Pain is sudden and severe, and situated over the shoulder rather than in the neck itself. Careful examination will show that more than one neural level is affected – an extremely rare event in disc prolapse.

Cervical spine infections Pain is unrelenting and local spasm severe. X-rays show erosion of the vertebral end-plates.

Cervical tumours Neurological signs are progressive and x-rays reveal bone destruction.

Rotator cuff lesions Although the distribution of pain may resemble that of a prolapsed cervical disc, tenderness is localized to the rotator cuff and shoulder movements are abnormal.

Treatment

Heat and analgesics are soothing but, as with lumbar disc prolapse, there are only three satisfactory ways of treating the prolapse itself.

Rest A collar will prevent unguarded movement; However, it seldom needs to be worn for more than a week or two.

Reduce Traction may enlarge the disc space, permitting the prolapse to subside. The head of the couch is raised and weights (up to 8 kg) are tied to a harness fitting under the chin and occiput. Traction is applied intermittently for no more than 30 minutes at a time.

Remove If symptoms are refractory and severe enough, if there is a progressive neurological deficit or if there are signs of an acute myelopathy then surgery is indicated. The disc may be removed through an anterior approach; bone grafts are inserted to fuse the affected segment and to restore the normal intervertebral height. If only one level is affected, and there is no bony encroachment on the intervertebral foramen, anterior decompression can be expected to give good long-term relief from radicular symptoms.

CERVICAL SPONDYLOSIS

This vague term is applied to a cluster of abnormalities arising from chronic intervertebral disc degeneration. Changes are most common in the lower two segments of the cervical spine (C5/6 and C6/7), the area which is prone to intervertebral disc prolapse. The discs degenerate, flatten and become less elastic. The facet joints and the uncovertebral joints are slightly displaced and become arthritic, giving rise to pain and stiffness in the neck. Bony spurs, ridges or bars appear at the anterior and posterior margins of the vertebral bodies; those that develop posteriorly may encroach upon the spinal canal or the intervertebral foramina, causing pressure on the dura (which is pain sensitive) and the neural structures.

The back 18

CLINICAL ASSESSMENT

SYMPTOMS

The usual symptoms of back disorders are pain, stiffness and deformity in the back, and pain, paraesthesia or weakness in the lower limbs. The mode of onset is very important: did it start suddenly, perhaps after a lifting strain; or gradually without any antecedent event? Are the symptoms constant, or are there periods of remission? Are they related to any particular posture? Has there been any associated illness or malaise?

Pain, either sharp and localized or chronic and diffuse, is the commonest presenting symptom. Backache is usually felt low down and on either side of the midline, often extending into the upper part of the buttock and even into the lower limbs. Back pain made worse by rest would suggest pain arising from the facet joints. Pain made worse by activity probably comes from any of the soft-tissue supports of the spine (muscles and ligaments) including the annulus of the intervertebral disc.

Sciatica is the term originally used to describe intense pain radiating from the buttock into the thigh and calf – more or less following the distribution of the sciatic nerve and therefore suggestive of nerve root compression or irritation. However Kellgren (1977), in a classic experiment, showed that almost any structure in a spinal segment can, if irritated sufficiently, give rise to *referred pain* radiating into the lower limbs. Unfortunately, with the passage of time, many clinicians have taken to describing all types of pain extending from the lumbar region into the lower limb as 'sciatica'. This is at best confusing and at worst a preparation for misdiagnosis! True *sciatica*, most commonly due to a prolapsed intervertebral disc pressing on a nerve root, is characteristically more intense than referred low back pain, is aggravated by coughing and straining and is often accompanied by symptoms of root pressure such as numbness and paraesthesiae, especially in the foot.

Stiffness may be sudden in onset and almost complete (in a 'locked back' attack, or after a disc prolapse) or continuous and predictably worse in the mornings (suggesting arthritis or ankylosing spondylitis).

Deformity is usually noticed by others, but the patient may become aware of shoulder asymmetry or of clothes not fitting well.

Numbness or *paraesthesia* is felt anywhere in the lower limb, but can usually be mapped fairly accurately over one of the dermatomes. It is important to ask if it is aggravated by standing or walking and relieved by sitting down – the classic symptom of spinal stenosis.
Urinary retention or *incontinence* can be due to pressure on the cauda equina.
Faecal incontinence or urgency, and *impotence*, may also occur.
Other symptoms important in back disorders are: (1) urethral discharge; (2) diarrhoea; (3) sore eyes – classical features of Reiter's disease.

SCOLIOSIS

Scoliosis is an apparent lateral (sideways) curvature of the spine. 'Apparent' because, although lateral curvature does occur, the commonest form of scoliosis is actually a triplanar deformity with lateral, anteroposterior and rotational components (Dickson et al., 1984). Two broad types of deformity are defined: *postural* and *structural*.

Postural Scoliosis

In postural scoliosis the deformity is secondary or compensatory to some condition outside the spine, such as a short leg, or pelvic tilt due to contracture of the hip. When the patient sits (thereby cancelling leg length asymmetry) the curve disappears. Local muscle spasm associated with a prolapsed lumbar disc may cause a skew back; although sometimes called 'sciatic scoliosis' this, too, is a spurious deformity.

Structural scoliosis

In structural scoliosis there is a non-correctable deformity of the affected spinal segment, an essential component of which is vertebral rotation. The spinous processes swing round towards the concavity of the curve and the transverse processes on the convexity rotate posteriorly. In the thoracic region the ribs on the convex side stand out prominently, producing the rib hump, which is a characteristic part of the overall deformity. Dickson and co-workers (1984) have pointed out that this is really a lordoscoliosis associated with rotational buckling of the spine. The initialdeformity is probably correctable, but once it exceeds a certain point of mechanical stability the spine buckles and rotates into a fixed deformity that does not disappear with changes in posture. Secondary (compensatory) curves nearly always develop to counterbalance the primary deformity; they are usually less marked and more easily correctable, but with time they, too, become fixed. Once fully established, the deformity is liable to increase throughout the growth period. Thereafter, further deterioration is slight, though curves greater than 50 degrees may go on increasing by 1 degree per year. With very severe curves, chest deformity is marked and cardiopulmonary function is usually affected.

Most cases have no obvious cause (*idiopathic scoliosis)*; other varieties are *congenital* or *osteopathic* (due to bony anomalies), *neuropathic, myopathic* (associated with some muscle dystrophies) and a miscellaneous group of connective-tissue disorders.

IDIOPATHIC SCOLIOSIS

This group constitutes about 80 per cent of all cases of scoliosis. The deformity is often familial and the population incidence of serious curves (over 30 degrees and therefore needing treatment) is three per 1000; trivial curves are very much more common. The age at onset has been used to define three groups: *adolescent*, *juvenile* and *infantile*. A simpler division now in general use is *early-onset* (before puberty) and *late-onset scoliosis* (after puberty).

Treatment of scoliosis

The aims of treatment are: (1) to prevent a mild deformity from becoming severe; (2) to correct an existing deformity that is unacceptable to the patient. A period of preliminary observation may be needed before deciding between conservative and operative treatment. At

4–9-monthly intervals the patient is examined, photographed and x-rayed so that curves can be measured and checked for progression.

NON-OPERATIVE TREATMENT

If the patient is approaching skeletal maturity and the deformity is acceptable (which usually means it is less than 30 degrees and well balanced), treatment is probably unnecessary unless sequential x-rays show definite progression.

Exercises are often prescribed; they have no effect on the curve but they do maintain muscle tone and may inspire confidence in a favourable outcome.

Bracing has been used for many years in the treatment of progressive scoliotic curves between 20 and 30 degrees.

The *Milwaukee brace* is principally a thoracic support consisting of a pelvic corset connected by adjustable steel supports to a cervical ring carrying occipital and chin pads; its purpose is to reduce the lumbar lordosis and encourage active stretching and straightening of the thoracic spine.

The *Boston brace* is a snug-fitting underarm brace that provides lumbar or low thoracolumbar support. Corrective pads may be added to these devices to apply pressure at a particular site. A well-made brace can be worn 23 hours out of 24 and does not preclude full daily activities, including sport and exercises. It has long been recognized that bracing will not improve the curve – at best it will merely stop it from getting worse. Many orthopaedic surgeons no longer employ this method of treatment, arguing that there is insufficient evidence of its benefits. Their preference now is to wait for the curve to progress to the stage when corrective surgery would be justified.

OPERATIVE TREATMENT

Surgery is indicated: (1) for curves of more than 30 degrees that are cosmetically unacceptable, especially in pre-pubertal children who are liable to develop marked progression during the growth spurt; (2) for milder deformity that is deteriorating rapidly. Balanced, double primary curves require operation only if they are greater than 40 degrees and progressing.

The objectives are: (1) to halt progression of the deformity; (2) to straighten the curve (including the rotational component) by some form of instrumentation; (3) to arthrodese the entire primary curve by bone grafting.

KYPHOSIS

Rather confusingly, the term ‘kyphosis’ is used to describe both the normal (gentle rounding of the thoracic spine) and the abnormal (excessive thoracic curvature or straightening out of the cervical or lumbar lordotic curves). Excessive thoracic curvature might be better described as ‘hyperkyphosis’. *Kyphos*, or gibbus, is a sharp posterior angulation due to localized collapse or wedging of one or more vertebrae. This may be the result of a congenital defect, a fracture (sometimes pathological) or spinal tuberculosis (see Fig. 18.24).

SPINAL INFECTION

The axial skeleton accounts for 2–7 per cent of all cases of osteomyelitis. Predisposing factors include diabetes mellitus, malnutrition, substance abuse, human immunodeficiency virus (HIV) infection, malignancy, long-term use of steroids, renal failure and septicaemia.

DISCITIS

Infection limited to the intervertebral disc is rare and when it does occur it is usually due to direct inoculation following discography, chemonucleolysis or discectomy. The vertebral end-plates are rapidly attacked and the infection then spreads into the vertebral body.

Clinical features and investigations

With direct infection there is always a history of some invasive procedure. Acute back pain and muscle spasm following an injection into the disc should never be attributed merely to the irritant effect of the injection. Systemic features are usually mild, but the ESR is elevated. *In children* the infection is assumed to be bloodborne. There may be a history of a flu-like illness followed by back pain, muscle spasm and severe limitation of movement. X-rays, radioscintigraphy and MRI show the same features as in pyogenic spondylitis.

Treatment

Prevention is always better than cure. Following an injection into the disc, a broad-spectrum antibiotic should be administered intravenously. Non-iatrogenic discitis is usually self-limiting. During the acute stage bed rest and analgesics are essential. If symptoms do not resolve rapidly, a needle biopsy is advisable. Only if there are signs of abscess formation or cord or nerve root pressure is surgical evacuation or decompression indicated. This is rarely necessary.

ACUTE INTERVERTEBRAL DISC PROLAPSE

Acute disc herniation (prolapse, rupture) is much less common but considerably more dramatic than chronic degeneration. Physical stress (a combination of flexion and compression) is the proximate cause but, even at L4/5 or L5/S1 (where stress is most severe) it seems unlikely that a disc would rupture unless there was also some disturbance of the hydrophilic properties of the nucleus. A *'protrusion'* is a posteriorly bulging disc with some outer annulus intact. When *rupture* does occur, fibrocartilaginous disc material is extruded posteriorly (*'extrusion'*) and usually bulges to one or other side of the posterior longitudinal ligament. With a complete rupture, part of the nucleus may sequestrate and lie free in the spinal canal or work its way into the intervertebral foramen (*sequestration*). A large central rupture may cause compression of the cauda equina. A posterolateral rupture presses on the nerve root proximal to its point of exit throughthe intervertebral foramen; thus a herniation at L4/5 will compress the fifth lumbar nerve root, and a herniation at L5/S1, the first sacral root. Sometimes a local inflammatory response with oedema aggravates the symptoms. *Acute back* pain at the onset of disc herniation probably arises from disruption of the outermost layers of the annulus fibrosus and stretching or tearing of the posterior longitudinal ligament. If the disc protrudes to one side, it may irritate the dural covering of the adjacent nerve root causing pain in the buttock, posterior thigh and calf (*sciatica*). Pressure on the nerve root itself causes *paraesthesia* and/or *numbness* in the corresponding dermatome, as well as *weakness* and *depressed reflexes* in the muscles supplied by that nerve root.

Clinical features

Acute disc prolapse may occur at any age, but is uncommon in the very young and the very old. The patient is usually a fit adult aged 20–45 years. Typically, while lifting or stooping he has severe back pain and is unable to straighten up. Either then or a day or two later pain is felt in the buttock and lower limb (sciatica). Both backache and sciatica are made worse by coughing or straining. Later there may be paraesthesia or numbness in the leg or foot, and occasionally muscle weakness. Cauda equina compression is rare but may cause urinary retention and perineal numbness. The patient usually stands with a slight list to one side (‘sciatic scoliosis’). Sometimes the knee on the painful side is held slightly flexed to relax tension on the sciatic nerve; straightening the knee makes the skew back more obvious. All back movements are restricted, and during forward flexion the list may increase. There is often tenderness in the midline of the low back, and paravertebral muscle spasm. Straight leg

raising is restricted and painful on the affected side; dorsiflexion of the foot and bowstringing of the lateral popliteal nerve may accentuate the pain. Sometimes raising the unaffected leg causes acute sciatic tension on the painful side (‘crossed sciatic tension’). With a high or mid-lumbar prolapse the femoral stretch test may be positive. Neurological examination may show muscle weakness (and, later, wasting), diminished reflexes and sensory loss corresponding to the affected level. L5 impairment causes weakness of knee flexion and big toe extension as well as sensory loss on the outer side of the leg and the dorsum of the foot. Normal reflexes at the knee and ankle are characteristic of L5 root compression. Paradoxically, the knee reflex may appear to be *increased*, because of weakness of the antagonists (which are supplied by L5). S1 impairment causes weak plantar-flexion and eversion of the foot, a depressed ankle jerk and sensory loss along the lateral border of the foot. Occasionally an L4/5 disc prolapse compresses both L5 and S1. Cauda equina compression causes urinary retention and sensory loss over the sacrum.

Imaging

X-rays are helpful, not to show an abnormal disc space but to exclude bone disease. After several attacks the disc space may be narrowed and small osteophytes appear.

Myelography (radiculography) using iopamidol (Niopam) is a fairly reliable method of confirming the nerve root distortion resulting from a disc protrusion, localizing it and excluding intrathecal tumours; however, it carries a significant risk of unpleasant sideeffects, such as headache (in over 30 per cent), nausea and dizziness. Myelography is unsuitable for diagnosing a far lateral disc protrusion (lateral to the intervertebral foramen); if this is suspected CT or MRI is essential.

CT and *MRI* are more reliable than myelography and have none of its disadvantages. These are now the preferred methods of spinal imaging.

Differential diagnosis

The full-blown syndrome is unlikely to be misdiagnosed, but with repeated attacks and with lumbar spondylosis gradually supervening (see later), the features often become atypical. There are two diagnostic aphorisms:

· *Lower limb pain* is not always the sciatica of root compression; frequently it is referred pain from backache and can occur in other lumbar spine disorders.

· *Disc rupture* affects at most two neurological levels; if multiple levels are involved, suspect a cauda equina compression (see box) or a neurological disorder.

Inflammatory disorders such as infection or ankylosing spondylitis, cause severe stiffness, a raised ESR and erosive changes on x-ray.

Vertebral tumours cause severe pain, marked muscle spasm and pain through the night. With metastases the patient is ill, the ESR is raised and the x-rays show bone destruction or sclerosis.

Nerve tumours such as a neurofibroma of the cauda equina, may cause ‘sciatica’ but pain is continuous. Advanced imaging will confirm the diagnosis.

FEATURES OF CAUDA EQUINA SYNDROME
Bladder and bowel incontinence
Perineal numbness
Bilateral sciatica
Lower limb weakness
Crossed straight-leg raising sign
Note: Scan urgently and operate urgently if a large central disc is revealed.

Treatment

Heat and analgesics soothe, and exercises strengthen muscles, but there are only three ways of treating the prolapse itself – *rest, reduction* or *removal,* followed by *rehabilitation*:

Rest With an acute attack the patient should be kept in bed, with hips and knees slightly flexed. A nonsteroidal anti-inflammatory drug is useful.

Reduction Continuous bed rest and traction for 2 weeks may reduce the herniation. If the symptoms and signs do not improve during that period, an epidural injection of corticosteroid and local anaesthetic may help.

Chemonucleolysis dissolution of the nucleus pulposus by percutaneous injection of a proteolytic enzyme (chymopapain) – is in theory an excellent way of reducing a disc prolapse. However, controlled studies have shown that it is less effective (and potentially more dangerous) than surgical removal of the disc material (Ejeskar et al., 1982).

Removal The indications for operative removal of a prolapse are: (1) a cauda equina compression syndrome – this is an emergency; (2) neurological deterioration while under conservative treatment; (3) persistent pain and signs of sciatic tension (especially crossed sciatic tension) after 2–3 weeks of conservative treatment. The presence of a prolapsed disc, and the level, must be confirmed by CT, MRI or myelography before operating. Surgery in the absence of a clear preoperative diagnosis is usually unrewarding. The two operations most widely performed are *laminotomy* and *microdiscectomy.*

Laminotomy is nowadays preferred to the older, more destructive type of laminectomy. Ligamentum flavum on the relevant side and at the relevant level is removed, if necessary with some margin of the bordering laminae and medial third of the facet joint. The dura and nerve root are then gently retracted towards the midline and the pea-like disc bulge or extrusion/ sequestration is displayed. If the outer layer of the annulus is seen still to be intact, it is incised and the mushy disc material plucked out piecemeal with pituitary forceps. The nerve is traced to its point of exit in order to exclude other pathology. A far lateral disc protrusion is very difficult to expose by the standard interlaminar approach without damaging the facet joint. An intertransverse approach may be more suitable for these cases. The main intraoperative complication is bleeding from epidural veins. This is less likely to occur if the patient is placed on his side or in the kneeling position, thus minimizing pressure on the abdomen and a rise in venous pressure. The major postoperative complication is disc space infection, but fortunately this is rare. Recurrent prolapse with sciatica is more common and may require revision decompression surgery.

Microdiscectomy is essentially similar to the standard posterior operation, except that the exposure is very limited and the procedure is carried out with the aid of an operating microscope. Morbidity and length of hospitalization are certainly less than with conventional surgery, but there are drawbacks: careful x-ray control is needed to ensure that the correct level is entered; intraoperative bleeding may be difficult to control; there is a considerable ‘learning curve’ and the inexperienced operator risks injuring the dura or a stretched nerve root, or missing essential pathology; there is a slightly increased risk of disc space infection, and prophylactic antibiotics are advisable.

Rehabilitation After recovery from an acute disc rupture, or disc removal, the patient is taught isometric exercises and how to lie, sit, bend and lift with the least strain. Ideally this should be done as part of an education programme in a ‘back school’ (Zachrisson, 1981).

SPONDYLOLISTHESIS

‘Spondylolisthesis’ means forward translation of one segment of the spine upon another. The shift is nearly always between L4 and L5, or between L5 and the sacrum. Normal discs, laminae and facets constitute a locking mechanism that prevents each vertebra from moving

forwards on the one below. Forward shift (or slip) occurs only when this mechanism has failed.

APPROACH TO DIAGNOSIS IN PATIENTS WITH LOW BACK PAIN

Chronic backache is such a frequent cause of disability in the community that it has become almost a disease in itself. The following is a suggested approach to more specific diagnosis. Careful history taking and examination will uncover one of five pain patterns:

1. Transient backache following muscular activity This suggests a simple back strain that will respond to a short period of rest followed by gradually increasing exercise. People with thoracic kyphosis (of whatever origin), or fixed flexion of the hip, are particularly prone to back strain because they tend to compensate for the deformity by holding the lumbosacral spine in hyperlordosis.

2. Sudden, acute pain and sciatica In young people (those under the age of 20) it is important to exclude *infection* and *spondylolisthesis*; both produce recognizable x-ray changes. Patients aged 20–40 years are more likely to have an acute disc prolapse: diagnostic features are: (1) a history of a lifting strain, (2) unequivocal sciatic tension; (3) neurological symptoms and signs. Elderly patients may have osteoporotic *compression fractures*, but *metastatic disease* and *myeloma* must be excluded.

3. Intermittent low back pain after exertion Patients of almost any age may complain of recurrent backache following exertion or lifting activities and this is relieved by rest. Features of disc prolapse are absent but there may be a history of acute sciatica in the past. In early cases x-rays usually show no abnormality; later there may be signs of *lumbar spondylosis* in those over 50 years and *osteoarthritis* of the facet joints is common. These patients need painstaking examination to: (1) uncover any features of radiological segmental instability or facet joint osteoarthritis; (2) determine whether those features are incidental or are likely to account for the patient's symptoms. In the process, disorders such as ankylosing *spondylitis*, *chronic infection*, *myelomatosis* and other *bone diseases* must be excluded by appropriate imaging and blood investigations.

4. Back pain plus pseudoclaudication These patients are usually aged over 50 and may give a history of previous, longstanding back trouble. The diagnosis of *spinal stenosis* should be confirmed by CT and/or MRI.

5. Severe and constant pain localized to a particular site This suggests local bone pathology, such as a compression fracture, Paget's disease, a tumour or infection. Spinal osteoporosis in middle-aged men is pathological and calls for a full battery of tests to exclude primary disorders such as *myelomatosis*, *carcinomatosis*, *hyperthyroidism*, *gonadal insufficiency*, *alcoholism* or *corticosteroid usage*.

The hip 19

CLINICAL ASSESSMENT

SYMPTOMS

Pain arising in the hip joint is felt in the groin, down the front of the thigh and, sometimes, in the knee; occasionally knee pain is the only symptom! Pain at the back of the hip is seldom from the joint; it usually derives from the lumbar spine. *Limp* is the next most common symptom. It may simply be a way of coping with pain, or it may be due to a change in limb length, weakness of the hip abductors or joint instability. *Snapping* or *clicking* in the hip suggests a number of causes: slipping of the gluteus maximus tendon over the greater trochanter, detachment of the acetabular labrum or psoas bursitis. *Stiffness* and *deformity* are late symptoms, and tend to be well compensated for by pelvic mobility. *Walking distance* may be curtailed; or, reluctantly, the patient starts using a walking stick.

SIGNS WITH THE PATIENT UPRIGHT

Start by standing face to face with the patient and note his or her general build and the symmetry of the lower limbs. First impressions are important and can be put to the test as the examination proceeds. The patient in Figure 19.1, for example, seems to have unusually short lower limbs in comparison to his trunk length. Is it a mild type of bone dysplasia, or are the hips dislocated?

Trendelenburg's sign

This is a test for postural stability when the patient stands on one leg. In normal two-legged stance the body's centre of gravity is placed midway between the two feet. Normally, in one-legged stance, the pelvis is pulled up on the unsupported side and the centre of gravity is placed directly over the standing foot. If the weightbearing hip is unstable, the pelvis *drops* on the unsupported side; to avoid falling, the person has to throw his body towards the loaded side so that the centre of gravity is again over that foot. If the difference between the two hips is marked you can detect it by simply looking at the patient's stance. However, small differences are not so obvious. In the classical Trendelenburg test the examiner stands behind the patient and looks at the buttockfolds. Normally in one-legged stance the buttock on the opposite side rises as the person lifts that leg; in a positive (abnormal) test the opposite buttock-fold drops (Fig. 19.2).

The causes of a positive Trendelenburg sign are: (1) pain on weightbearing; (2) weakness of the hip abductors; (3) shortening of the femoral neck; and (4) dislocation or subluxation of the hip.

Gait

Now ask the patient to walk, and observe each phase of the gait. The commonest abnormalities are a *shortleg limp* (a regular, even dip on the short side), an *antalgic gait* (an irregular limp, with the patient moving more quickly off the painful side) and a *Trendelenburg lurch* (a variant of Trendelenburg's sign). While the patient is upright, take the opportunity to examine the spine for deformity or limitation of movement.

SIGNS WITH THE PATIENT SITTING

This is the best way to test for iliopsoas function. The patient should be sitting on the edge of the examination couch. Place a hand firmly on his thigh and ask him to lift the thigh (flex the hip) against resistance. This is a predominantly psoas action; pain or weakness suggests a local disorder such as tendinitis or psoas bursitis.

SIGNS WITH THE PATIENT LYING

Look

Scars or sinuses may be seen (or they may be at the back of the hip). Compare the two sides for signs of muscle wasting or swelling. Check that the pelvis is horizontal (both anterior superior iliac spines at the same level) and the legs placed symmetrically. Limb length can be gauged by looking at the ankles and heels, but measurement is more accurate. With the two legs in identical positions, measure the distance from the anterior superior iliac spine to the medial malleolus on each side. The limb may lie in an abnormal position; excessive rotation is easy to detect but other deformities are often masked by tilting of the pelvis. Sometimes the *real length*, as determined by measuring between two bony points, is quite different from the *apparent length* with the patient lying in repose. This happens when the pelvis is tilted and one limb is hitched upwards. Almost invariably this is due to an uncorrectable deformity at the hip: with fixed adduction on one side, the limbs would tend to be crossed; when the legs are placed side by side the pelvis has to tilt upwards on the affected side, giving the impression of a shortened limb. The exact opposite occurs when there is fixed abduction, and the limb seems to be longer on the affected side. If real shortening is present it is usually possible to establish where the fault lies. With the knees flexed and the heels together, it can be seen whether the discrepancy is below or above the knee. If it is above, the next question is whether the abnormality lies above the greater trochanter. The thumbs are pressed firmly against the anterior superior iliac spines and the middle fingers grope for the tops of the greater trochanters; any elevation of the trochanter on one side is readily appreciated.

Feel

Skin temperature and *soft-tissue* contours can be felt, but are unhelpful unless the patient is very thin. *Bone* contours are felt when levelling the pelvis and judging the height of the greater trochanters. *Tenderness* may be elicited in and around the joint.

Move

The assessment of hip movements is difficult because any limitation can easily be obscured by movement of the pelvis. Thus, even a gross limitation of extension, causing a *fixed flexion deformity,* can be completely masked simply by arching the back into excessive lordosis. Fortunately it can be just as easily unmasked by performing *Thomas' test* (Fig. 19.5): both hips are flexed simultaneously to their limit, thus completely obliterating the lumbar lordosis; holding the 'sound' hip firmly in position (and thus keeping the pelvis still), the other limb is lowered gently; with any flexion deformity the knee will not rest on the couch. Meanwhile the full range of *flexion* will also have been noted; the normal range is about 130 degrees. Similarly, when testing *abduction* the pelvis must be prevented from tilting sideways. This is achieved by placing the 'sound' hip (the hip opposite to the one being examined) in full abduction and keeping it there. A hand is placed on one iliac crest to detect the slightest movement of the pelvis. Then, after checking that the anterior superior iliac spines are level, the affected joint is moved gently into abduction. The normal range is about 40 degrees.

Adduction is tested by crossing one limb over the other; the pelvis must be watched and felt to determine the point at which it starts to tilt. The normal range of adduction is about 30 degrees. To test *rotation* both legs, lifted by the ankles, are rotated first internally (medially) and then externally (laterally); the patellae are watched to estimate the amount of rotation. Rotation in flexion is tested with the hip and knee each flexed 90 degrees. If internal rotation is full with the hip extended, but restricted in flexion, this suggests pathology in the anterosuperior portion of the femoral head, probably avascular necrosis (the so-called ‘sectoral sign’). However, in a young person, pain on internal rotation with the hip flexed may indicate a torn acetabular labrum.
Abnormal movement is rarely elicited. Telescoping (excessive movement when the limb is alternately pulled and pushed in its long axis) is a sign of gross instability.
Do not forget the back of the hip. Ask the patient to roll over into the prone position. Check for scars and sinuses. Feel for tenderness and test the range of hip extension. Rotation can also be assessed by flexing both knees and moving the legs, first away from each other (producing internal rotation at the hips) and then towards or crossing each other (external rotation).

DEVELOPMENTAL DYSPLASIA OF THE HIP

The terminology used to describe abnormalities of the paediatric hip is imprecise and confusing. The term ‘congenital dislocation of the hip’ (CDH) has been largely superseded by *developmental dysplasia of the hip (DDH)* in an attempt to describe the range and evolution of abnormalities that occur in this condition. This comprises a spectrum of disorders including acetabular dysplasia without displacement, subluxation and dislocation. Teratological forms of malarticulation leading to dislocation are also included. Normal hip development depends on proportionate growth of the acetabular triradiate cartilages and the presence of a concentrically located femoral head. Whether the instability comes first and then affects acetabular development because of imperfect seating of the femoral head, or is a result of a primary acetabular dysplasia, is still uncertain. Both mechanisms might be important. The reported incidence of neonatal hip instability in northern Europe is approximately 1 per 1000 live births, but this is dependent on the definition of ‘instability’. Barlow (1962) described an incidence of 1:60; however 60 per cent stabilized by one week and 88 per cent by 8 weeks. The incidence is considerably higher in some ethnic groups - 25-50 cases per 1000 live births in Lapps and Native Americans!
Girls are much more commonly affected than boys, the ratio being about 7:1. The left hip is more often affected than the right; in 1 in 5 cases the condition is bilateral.

Clinical features

The ideal, still unrealized, is to diagnose every case at birth. For this reason, every newborn child should be examined for signs of hip instability. Where there is a family history of congenital instability, and with breech presentations or signs of other congenital abnormalities, extra care is taken and the infant may have to be examined more than once. Even then some cases are missed.
In the neonate There are several ways of testing for instability. In ***Ortolani's test***, the baby’s thighs are held with the thumbs medially and the fingers resting on the greater trochanters; the hips are flexed to 90 degrees and gently abducted. Normally there is smooth abduction to almost 90 degrees. In congenital dislocation the movement is usually impeded, but if pressure is applied to the greater trochanter there isva soft ‘clunk’ as the dislocation reduces, and then thevhip abducts fully (the ‘jerk of entry’). If abductionvstops halfway and there is no jerk of entry, there may be an irreducible dislocation.

Barlow's test is performed in a similar manner, but here the examiner's thumb is placed in the groin and, by grasping the upper thigh, an attempt is made to lever the femoral head in and out of the acetabulum during abduction and adduction. If the femoral head is normally in the reduced position, but can be made to slip out of the socket and back in again, the hip is classed as 'dislocatable' (i.e. unstable).

Every hip with signs of instability - however slight - should be examined by *ultrasonography.* This shows the shape of the cartilaginous socket and the position of the femoral head. If there is any abnormality, the infant is placed in a splint with the hips flexed and abducted (see under Management) and is recalled for re-examination - in the splint - at 2 weeks and at 6 weeks. By then it should be possible to assess whether the hip is reduced and stable, reduced but unstable (dislocatable by Barlow's test), subluxated or dislocated.

Late features An observant mother may spot asymmetry, a clicking hip, or difficulty in applying the napkin (diaper) because of limited abduction. With unilateral dislocation the skin creases look asymmetrical and the leg is slightly short (Galeazzi's sign) and externally rotated; a thumb in the groin may feel that the femoral head is missing. With bilateral dislocation there is an abnormally wide perineal gap. Abduction is decreased. Contrary to popular belief, late walking is not a marked feature; nevertheless, in children who do not walk by 18 months dislocation must be excluded. Likewise, a limp or Trendelenburg gait, or a waddling gait could be a sign of missed dislocation.

Imaging

Ultrasonography Ultrasound scanning has replaced radiography for imaging hips in the newborn. The radiographically 'invisible' acetabulum and femoral head can, with practice, be displayed with static and dynamic ultrasound. Sequential assessment is straightforward and allows monitoring of the hip during a period of splintage.

Plain x-rays X-rays of infants are difficult to interpret and in the newborn they can be frankly misleading. This is because the acetabulum and femoral head are largely (or entirely) cartilaginous and therefore not visible on x-ray. X-ray examination is more useful after the first 6 months, and assessment is helped by drawing lines on the x-ray plate to define three geometric indices (Fig. 19.10).

Management

THE FIRST 3–6 MONTHS

Where facilities for ultrasound scanning are available, all newborn infants with a high-risk background or a suggestion of hip instability are examined by ultrasonography. If this shows that the hip is reduced and has a normal cartilaginous outline, no treatment is required but the child is kept under observation for 3-6 months. In the presence of acetabular dysplasia or hip instability, the hip is splinted in a position of flexion and abduction (see below) and ultrasound scanning is repeated at intervals until stability and normal anatomy are restored or a decision is made to abandon splintage in favour of more aggressive treatment.

If ultrasound is not available, the simplest policy is to regard all infants with a high-risk background or a positive Ortolani or Barlow test, as 'suspect' and to nurse them in double napkins or an abduction pillow for the first 6 weeks. At that stage they are re-examined: those with stable hips are left free but kept under observation for at least 6 months; those with persistent instability are treated by more formal abduction splintage (see below) until the hip is stable and x-ray shows that the acetabular roof is developing satisfactorily (usually 3-6 months) There are two drawbacks to this approach: (1) the sensitivity of the clinical tests is not high enough to ensure that all cases will be spotted (Jones, 1994); and (2) of those hips that are unstable at birth, 80-90 per cent will stabilize spontaneously in 2-3 weeks. It therefore seems more sensible not to start splintage immediately unless the hip is already

dislocated. This reduces the small (but significant) risk of epiphyseal necrosis that attends any form of restrictive splintage in the neonate. Thus: if a hip is dislocatable but not habitually dislocated, the baby is left untreated but reexamined weekly; if at 3 weeks the hip is still unstable, abduction splintage is applied (see below). If the hip is already dislocated at the first examination, it is gently placed in the reduced position and abduction splintage is applied from the outset. Reduction is maintained until the hip is stable; this may take only a few weeks, but the safest policy is to retain some sort of splintage until x-ray shows a good acetabular roof.

Splintage The object of splintage is to hold the hips somewhat flexed and abducted; extreme positions are avoided and the joints should be allowed some movement in the splint. Von Rosen's splint is an H-shaped malleable splint that has the merit of being easy to apply (and the demerit of being equally easy to take off!). The Pavlik harness is more difficult to apply but gives the child more freedom while still maintaining position. The three golden rules of splintage are:

(1)the hip must be properly reduced before it is splinted;

(2) extreme positions must be avoided;

(3) the hips should be able to move.

If the hip is splinted in a subluxed/ dislocated position, the posterior wall of the acetabulum is at risk of growth disturbance, leading to considerable difficulties with later reconstruction. This situation must be avoided; if the hip fails to locate, splintage should be abandoned in favour of closed or operative reduction at a later date.

Follow-up Whatever policy is adopted, follow-up is continued until the child is walking. Sometimes, even with the most careful treatment, the hip may later show some degree of acetabular dysplasia.

PERSISTENT DISLOCATION: 6–18 MONTHS

If, after early treatment, the hip is still incompletely reduced, or if the child presents late with a 'missed' dislocation, the hip must be reduced - preferably by closed methods but if necessary by operation - and held reduced until acetabular development is satisfactory.

Closed reduction Closed reduction is suitable after the age of 3 months and is performed under general anaesthesia with an arthrogram to confirm a concentric reduction. To minimize the risk of avascular necrosis, reduction must be gentle and may be preceded by gradual traction to both legs. Failure to achieve concentric reduction should lead to abandoning this method in favour of an operative approach at approximately 1 year of age. The hips should be stable in a safe zone of abduction, which may be increased with a closed adductor tenotomy.

Splintage The concentrically reduced hip is held in a plaster spica at 60 degrees of flexion, 40 degrees of abduction and 20 degrees of internal rotation. After 6 weeks the spica is changed and the stability of the hips assessed under anaesthesia. Provided the position and stability are satisfactory the spica is retained for a further 6 weeks. Following plaster removal the hip is either left unsplinted or managed in a removable abduction splint which is retained for up to 6 months depending on radiological evidence of satisfactory acetabular development.

Operation If, at any stage, concentric reduction has not been achieved, open operation is needed. The psoas tendon is divided; obstructing tissues (redundant capsule and thickened ligamentum teres) are removed and the hip is reduced. It is usually stable in 60 degrees of flexion, 40 degrees of abduction and 20 degrees of internal rotation. A spica is applied and the hip is splinted as described above. If stability can be achieved only by markedly internally rotating the hip, a corrective subtrochanteric osteotomy of the femur is carried out, either at the time of open reduction or 6 weeks later. In young children this usually gives a good result (Fig. 19.12a,b).

ACETABULAR DYSPLASIA AND SUBLUXATION OF THE HIP

Acetabular dysplasia may be genetically determined or may follow incomplete reduction of a congenital dislocation, damage to the lateral acetabular epiphysis or maldevelopment of the femoral head (either congenital or, for example, after Perthes' disease). The socket is unusually shallow, the roof is sloping and there is deficient coverage of the femoral head superolaterally and anteriorly; in some cases the hip subluxates. Faulty load transmission in the lateral part of the joint may lead to secondary osteoarthritis (OA).

Clinical features

During infancy, dysplasia may be clinically silent and only apparent on ultrasound examination. If there is associated instability, Barlow's test may be positive, but other clinical indicators including loss of abduction may be absent.

In children the condition is usually asymptomatic and discovered only when the pelvis is x-rayed for some other reason. Sometimes, however, the hip is painful – especially after strenuous activity – and the child may develop a limp. If there is subluxation the Trendelenburg sign is positive, leg length may be asymmetrical and the femoral head may be felt as a lump in the groin; movement – particularly abduction in flexion – is restricted.

Older adolescents and *young adults* may complain of pain over the lateral side of the hip, probably due to muscle fatigue and/or segmental overload towards the edge of the acetabulum. Some experience episodes of sharp pain in the groin, possibly the result of a labral tear or detachment.

Older adults (predominantly in their thirties and forties) usually present with features of secondary OA. Indeed, in southern Europe dysplasia of the hip is the commonest cause of symptomatic OA.

NOTE: It is worth emphasizing that most people with mild acetabular dysplasia go through life without knowing that they are in any way abnormal and the condition exists only as a 'x-ray diagnosis'.

IMAGING

X-rays should be taken lying and standing (the latter may show minor degrees of incongruity). In the supine anteroposterior radiograph, the acetabulum looks shallow, the roof is sloping and the femoral head is uncovered. Subtle abnormalities are revealed by measuring the depth of the socket and the relationship between the centre of the femoral head and the edge of the acetabulum – Wiberg's centre-edge (CE) angle. With subluxation, Shenton's line is broken.

The *faux profil* (oblique view) of the hip in the standing position will demonstrate acetabular dysplasia and incipient OA in the young adult. Congruity and stability of the hip may be best assessed by examination and dynamic arthrography under anaesthesia (Catterall, 1992).

CT and *MRI* are helpful in those who are considered for operative treatment. Three dimensional CT reconstruction is particularly useful in providing an accurate picture of the anatomy.

Diagnosis

It is often difficult to be sure that the patient's symptoms are due to the dysplastic acetabulum; other conditions causing pain and limp must be excluded (see Box on page 514). Bilateral dysplasia is a feature of developmental disorders, such as multiple epiphyseal dysplasia.

Treatment

Infants with subluxation are treated as for dislocation: the hip is splinted in abduction until the acetabular roof looks normal.

Young children (4–10) are treated with a Salter innominate osteotomy, provided the dysplastic

acetabulum remains congruent. It is often difficult to recommend surgery for an asymptomatic condition, but significant persistent dysplasia, without improvement of the acetabular index, in a child over 5 years old merits serious discussion.
Older children and *young adolescents*, provided the hip is reducible and congruent, often manage with no more than muscle-strengthening exercises. If symptoms persist, they may need an operation to augment the acetabular roof, either a lateral shelf procedure or a limited pelvic osteotomy such as the Chiari operation, either of which may be combined with a varus osteotomy of the proximal femur.
Older adolescents and *young adults* with pain, weakness, instability and subluxation of the hip are candidates for peri-acetabular osteotomy and threedimensional re-orientation of the entire hip (Ganz et al., 1998).
Patients with *secondary OA* may need inter trochanteric osteotomy or total hip replacement.

THE IRRITABLE HIP (TRANSIENT SYNOVITIS)

This condition is defined as a non-specific, short-lived synovitis, resulting in an effusion of the hip joint. It is the most common cause of an acute limp or hip pain in children, with a reported frequency of 14 per 1000. The most commonly affected age group is 3–8-yearolds with boys affected twice as often as girls. It affects both hips in 5 per cent of cases, although this is rarely simultaneous.

Aetiology

While viral infections, trauma and allergy have been suggested, the exact aetiology remains unclear. The pathological process involves a synovial effusion resulting in an increased intra-articular pressure.

Clinical features

The typical patient presents with pain and a limp, often intermittent and following activity. Pain is felt in the groin or front of the thigh, sometimes reaching as far as the knee. Slight wasting may be detectable but the cardinal sign is restriction of all movements with pain at the extremes of the range in all directions. The diagnosis is based primarily on the clinical features. Standard laboratory investigations including white cell count, erythrocyte sedimentation rate (ESR) and C-reactive protein concentration are usually within normal limits. X-rays do not demonstrate any bony defects, but occasionally there may be a subtle widening of the medial joint space (1–2 mm) when compared with the unaffected side. This is caused by the effusion which allows the femoral head to sublux slightly; it may be confirmed by ultrasonography. Characteristically, symptoms last for 1–2 weeks and then subside spontaneously; hence the synonym ‘transient synovitis’. The child may experience more than one episode, with an interval of months between attacks of pain.

Differential diagnosis

The condition is important largely because it resembles a number of serious disorders which have to be excluded.
Perthes' disease is the main worry. Acute symptoms usually last longer than 2 weeks and x-rays may show an increased ‘joint space’. Later, of course, the x-ray features are unmistakable.
Slipped epiphysis may present as an ‘irritable hip’. Initially the x-ray looks normal and this may lead to complacency. If the age and general build are suggestive, or if the symptoms persist, the x-ray should be repeated.
Tuberculous synovitis produces a raised ESR and the Heaf test is positive.

Juvenile chronic arthritis and *ankylosing spondylitis* may start with synovitis of one hip and it may take months before other joints are affected. Look for systemic features and a raised ESR. In doubtful cases, synovial biopsy may be helpful.
Septic arthritis should always be borne in mind. The early symptoms and signs are sometimes misleading, especially if someone has already prescribed antibiotics 'just in case!'
Treatment
Treatment involves bed rest, reduced activity and observation, which may be supervised at home or in hospital. Most children recover within a few days and any deterioration in signs or symptoms requires urgent reassessment. Traction, although popular in the past, is not currently recommended as it may increase the intra-articular pressure. Joint aspiration is ineffective; any relief in symptoms tends to be shortlived as the effusion rapidly recurs. Ultrasonography is repeated at intervals and weightbearing is allowed only when the symptoms disappear and the effusion resolves. Although this condition carries a good prognosis, recurrence rates of up to 10 per cent have been reported. A causal relationship with Perthes' disease has been suspected but remains unproven.

PERTHES' DISEASE

Perthes' disease – or rather Legg-Calve-Perthes disease, for in 1910 the condition was described independently by three different people – is a painful disorder of childhood characterized by avascular necrosis of the femoral head. It is uncommon in any community – the quoted incidence is about 1 in 10 000 – with a higher incidence in Japanese, Inuits and central Europeans and a lower incidence in native Australians, native Americans, Polynesians and blacks. Patients are usually 4-10 years old and boys are affected four times as often as girls. The condition may be part of a general disorder of growth. Epidemiological studies in the UK have shown that there is a higher than usual incidence in underprivileged communities. Affected children and their siblings have slightly retarded growth of the trunk and limbs. As in other forms of non-traumatic osteonecrosis, inherited thrombophilia has been postulated as a contributory cause and antithrombotic factor deficiencies and hypofibrinolysis have been reported in children with Perthes' disease (Glueck et al., 1996). This hypothesis has been questioned by others (Editorial by R. J. Liesner, 1999).
Pathogenesis
The precipitating cause of Perthes' disease is unknown but the cardinal step in the pathogenesis is ischaemia of the femoral head. Up to the age of 4 months, the femoral head is supplied by (1) metaphyseal vessels which penetrate the growth disc, (2) lateral epiphyseal vessels running in the retinacula and (3) scanty vessels in the ligamentum teres. The metaphyseal supply gradually declines until, by the age of 4 years, it has virtually disappeared; by the age of 7, however, the vessels in the ligamentum teres have developed. Between 4 and 7 years of age the femoral head may depend for its blood supply and venous drainage almost entirely on the lateral epiphyseal vessels whose situation in the retinacula makes them susceptible to stretching and to pressure from an effusion. Although such pressure may be insufficient to block off the arterial flow, it could easily cause venous stasis resulting in a rise in intraosseous pressure and consequent ischaemia (Lin and Ho, 1991). This may be enough to tip the balance towards infarction and necrosis in children who are constitutionally predisposed. The immediate cause of capsular tamponade may be an effusion following trauma (of which there is a history in over half the cases) or a non-specific synovitis. Two or more such incidents may be needed to produce the typical bone changes.
Pathology
The pathological process goes through several stages which in total may last up to 3 or 4 years.

Stage 1: Ischaemia and bone death All or part of the bony nucleus of the femoral head is dead; it still looks normal on plain x-ray but stops enlarging. The cartilaginous part of the femoral head, being nourished by synovial fluid, remains viable and becomes thicker than normal. There may also be thickening and oedema of the synovium and capsule.
Stage 2: Revascularization and repair Within weeks (possibly even days) of infarction, a number of changes begin to appear. Dead marrow is replaced by granulation tissue, which sometimes calcifies. The bone is revascularized and new lamellae are laid downon the dead trabeculae, producing the appearance of increased density on x-ray. Some of the dead trabecular fragments are resorbed and replaced by fibrous tissue; when this happens, the alternating areas of sclerosis and fibrosis appear on the x-ray as *'fragmentation'* of the epiphysis. The metaphysis may become hyperaemic and on x-ray looks rarefied or cystic. In older children, and more severe cases, morphological changes may also appear in the acetabulum.
Stage 3: Distortion and remodelling If the repair process is rapid and complete, the bony architecture may be restored before the femoral head loses its shape. If it is tardy, the bony epiphysis may collapse and subsequent growth of the femoral head and neck will be distorted: the head becomes oval or flattened – like the head of a mushroom – and enlarged laterally, while the neck is often short and broad. Slowly the femoral head is displaced laterally in relation to the acetabulum. Any residual deformity is likely to be permanent.
Clinical features
The patient – typically a boy of 4–8 years – complains of pain and starts limping. Symptoms continue for weeks on end or may recur intermittently. The child appears to be well, though often somewhat undersized. In 4 per cent there is an associated urogenital anomaly. The hip looks deceptively normal, though there may be a little wasting. Early on, the joint is irritable so that all movements are diminished and their extremes painful. Often the child is not seen till later, when most movements are full; but abduction (especially in flexion) is nearly always limited and usually
internal rotation also.
X-rays
Although the condition may be suspected from the clinical appearances, diagnosis hinges on the x-ray changes. At first the x-rays may seem normal, though subtle changes such as widening of the 'joint space' and slight asymmetry of the ossific centres are usually present. Radionuclide scanning may show a 'void' in the anterolateral part of the femoral head. The classic feature of increased density of the ossific nucleus occurs somewhat later. This is often referred to as the '*necrotic phase*', though the radiographically dense areas must surely be due to the new bone formation that always follows bone necrosis. This progresses to the phase of radiographic '*fragmentation*' – alternating patches of density and lucency, or sometimes a crescentic subarticular fracture often best seen in the lateral view. Epiphyseal density increases (the phase of *re-ossification*) and scintigraphy shows increased activity. With *healing* the femoral head may regain its normal (or near-normal) shape; however, in less fortunate cases the femoral head becomes mushroom- shaped, larger than normal and laterally displaced in a dysplastic acetabular socket.

The Catterall classification The radiographic picture varies with the age of the child, the stage of the disease and the amount of head that is necrotic.
Catterall (1982) described four groups, based on the appearances in both anteroposterior and lateral x-rays.
In group 1 the epiphysis has retained its height and
less than half the nucleus is sclerotic.

In group 2 up tohalf the nucleus is sclerotic and there may be some collapse of the central portion.

In group 3 most of the nucleus is involved, with sclerosis, fragmentation and collapse of the head. Metaphyseal resorption may be present.

Group 4 is the worst: the whole head is involved, the ossific nucleus is flat and dense and metaphyseal resorption is marked.

The Herring classification This classification embodies a greater degree of predictive value for the outcome of the Perthes changes and is therefore preferred by many orthopaedic surgeons. The features are described below and illustrated in Figure 19.25.

Prognostic features

The outlook for children with Perthes' disease, as a group, is well summarized by Herring (1994): 'A small percentage of patients have a very difficult course, with recurrent loss of motion, pain, and an eventual poor outcome. However, most children have moderate problems in the active phase of the disease and then improve steadily, eventually having a satisfactory outcome.' This does not, of course, absolve one from undertaking careful analysis and planning in dealing with the individual case. *Age* is the most important prognostic factor: in children under 6 years the outlook is almost always excellent; thereafter, the older the child the less good is the prognosis. There is a poorer prognosis, too, for *girls* than for boys. A widely used radiographic guide is the *Catterall classification* (see above). The greater the degree of femoral head involvement, the worse the outcome. This is recognized in the simpler *classification of Salter and Thompson*, into those with more and those with less than half the head involved (Simmons et al., 1990). There is also the concept of the *head at risk* - radiographic signs which presage increasing deformity and displacement of the femoral head: (1) progressive uncovering of the epiphysis; (2) calcification in the cartilage lateral to the ossific nucleus; (3) a radiolucent area at the lateral edge of the bony epiphysis (Gage's sign); and (4) severe metaphyseal resorption. Common to all these predictive systems is the importance of the structural integrity of the superolateral (principal load-bearing) part of the femoral head. This is reflected in Herring's *lateral pillar classification*. In the anteroposterior x-ray, the femoral head is divided into three 'pillars' by lines at the medial and lateral edges of the central 'sequestrum'. Group A are those with normal height of the lateral pillar. Group B are patients with partial collapse (but still more than 50 per cent height) of the lateral pillar; those under 9 years of age usually have a good outcome but older children are likely to develop flattening of the femoral head. Group C cases show more severe collapse of the lateral pillar (less than 50 per cent of normal height); these take longer to heal and usually end up with significant distortion of the femoral head.

Differential diagnosis

The irritable hip of early Perthes' disease must be differentiated from other causes of irritability; the child's fitness, the increased joint space and the patchy bone density are characteristic. In transient synovitis the x-ray is normal. Morquio's disease, cretinism, multiple epiphyseal dysplasia, sickle-cell disease and Gaucher's disease may resemble Perthes' disease radiologically, especially if they are bilateral; however, in bilateral Perthes' disease the two sides are likely to be at different stages. Moreover, in the other conditions general diagnostic features are usually apparent.

'Old Perthes deformities' in adults, in the 10 per cent of cases with bilateral involvement, may resemble those of certain bone dysplasias, especially multiple epiphyseal dysplasia. Look for changes in other epiphyses.

Management

The initial management of the child with Perthes' disease is determined by the severity of symptoms. Analgesia and modification of activities are often sufficient, but hospitalization for bed rest and short periods of traction are sometimes necessary. Wheelchair use and crutch walking should be discouraged in order to avoid unnecessary joint stiffness and contracture. Once joint irritability has subsided, which usually takes about 3 weeks, movement is encouraged, particularly cycling and swimming. Preservation of abduction is also important, with formal stretching used in some children. The clinical and radiographic features are then reassessed and the bone age is determined from x-rays of the wrist. The choice of further management is between (a) symptomatic treatment and (b) containment.

Symptomatic treatment means pain control (if necessary by further spells of traction), gentle exercise to maintain movement and regular reassessment. During asymptomatic periods the child is allowed out and about but sport and strenuous activities are avoided.

Containment means taking active steps to seat the femoral head congruently and as fully as possible in the acetabular socket, so that it may retain its sphericity and not become displaced during the period of healing and remodelling. This is achieved (a) by holding the hips widely abducted, in plaster or in a removable brace (ambulation, though awkward, is just possible, but the position must be maintained for at least a year); or (b) by operation, either a varus osteotomy of the femur or an innominate osteotomy of the pelvis, or both. In earlier years there was a good deal of support for non-operative containment, and this is still applicable where specialized surgical facilities are unavailable. However, this has been questioned by more recent outcome studies and the preferred approach is to achieve containment by operative methods (Martinez, 1992; Meehan 1992).

Operative reconstruction provides the advantages of improved containment and early mobilization. Shortterm studies also suggest an improvement in the anatomy of the hip, but there is no convincing evidence of any alteration in the natural history of the disorder or (in particular) the likelihood of needing an arthroplasty in later life.

GUIDELINES TO TREATMENT

There is no general agreement on the 'correct' course of treatment for all cases. Decisions are based on an assessment of the stage of the disease, the prognostic x-ray classifications, the age of the patient and the clinical features, particularly range of abduction and extension. The following guidelines are derived from the review by Herring (1994).

Children under 6 years No specific form of treatment has much influence on the outcome. Symptomatic treatment, including activity modification, is appropriate.

Children aged 6–8 years In this group the bone age is more important than the chronological age.

Bone age at or below 6 years

Lateral pillar group A and B (or Catterall stage I and II) - symptomatic treatment.

Lateral pillar group C (or Catterall stage III and IV)- abduction brace.

Bone age over 6 years

Lateral pillar group A and B (Catterall stage I and II)

- abduction brace or osteotomy.

Lateral pillar group C (Catterall stage III and IV) - outcome probably unaffected by treatment, but some would operate.

Children 9 years and older Except in very mild cases (which is rare), operative containment is the treatment of choice.

PYOGENIC ARTHRITIS

(see also Chapter 2)

Pyogenic arthritis of the hip is usually seen in children under 2 years of age. The organism (usually a staphylococcus) reaches the joint either directly from a distant focus or by local spread from osteomyelitis of the femur. Unless the infection is rapidly aborted, the femoral head, which is largely cartilaginous at this age, is liable to be destroyed by the proteolytic enzymes of bacteria and pus. Adults, also, may develop pyogenic hip infection, either as a primary event in states of debilitation or (more often) secondary to invasive procedures around the hip.

Clinical features

The child is ill and in pain, but it is often difficult to tell exactly where the pain is! The affected limb may be held absolutely still and all attempts at moving the hip are resisted. With care and patience it may be possible to localize a point of maximum tenderness over the hip; the diagnosis is confirmed by aspirating pus or fluid from the joint and submitting it for laboratory examination and bacteriological culture. In the acute stage x-rays are of little value but sometimes they show soft-tissue swelling, displacement of the femoral head and a vacuum sign in the joint. Ultrasonography will reveal the joint effusion. Diagnosis can be difficult, especially in neonates who may be almost asymptomatic. If the baby looks ill and no cause is apparent, think of *deep sepsis* and look for a possible source (e.g. an intravascular line). A high index of suspicion is the best aid.

Treatment

Intravenous antibiotics should be given as soon as the diagnosis is reasonably certain. The joint is aspirated under general anaesthesia and, if pus is withdrawn, anterior arthrotomy is performed; antibiotics are instilled locally and the wound is closed without drainage. Systemic antibiotics are essential, and the hip is kept on traction or splinted in abduction until all evidence of disease activity has disappeared.

Complications

If the infection is unchecked the head and neck of the femur may be destroyed and a pathological dislocation result. The pus may escape and, when the child recovers, the sinus heals. The hip signs then resemble those of a congenital dislocation, but the telltale scar remains and on x-ray the femoral head is completely absent.

OSTEOARTHRITIS

(see also Chapter 5)

The hip joint is one of the commonest sites of OA, though in some populations (e.g. African Negroes and southern Chinese) this joint seems peculiarly immune to the disease. This may simply be because certain predisposing conditions (acetabular dysplasia, Perthes' disease, slipped epiphysis) show a similar differential incidence in these populations. Where there is an obvious underlying cause the term '*secondary osteoarthritis'* is applied (Table 19.2); these patients are often in their third or fourth decade and the appearance of the joint reflects the preceding abnormality. Thus in regions where congenital dislocation and acetabular dysplasia are common (e.g. in southern Europe), women are more often affected than men, the hips may be the only joints affected and lateral subluxation is common. When no underlying cause is apparent, the term '*primary osteoarthritis*' is used. It is now believed that even in these cases there is some preceding disorder that leads to articular cartilage damage and subtle abnormalities are being sought in patients who would otherwise fall into the 'primary' category. In the case of the hip particular attention has been given to anatomical and mechanical factors that affect joint congruency and predispose to femoro-acetabular impingement and erosion of the articular surface. This comparatively new field of enquiry is explored on page 524.

Pathology

The articular cartilage becomes soft and fibrillated while the underlying bone shows cyst formation and sclerosis. These changes are most marked in the area of maximal loading (chiefly the top of the joint); at the margins of the joint there are the characteristic osteophytes. Synovial hypertrophy is common and capsular fibrosis may account for joint stiffness. The pathology of OA is discussed in greater detail in Chapter 5. Sometimes articular destruction progresses very rapidly, with erosion of the femoral head or acetabulum (or both), occasionally going on to perforation of the pelvis. This could be due to basic calcium crystal deposition in the joint (see Chapter 4).

Clinical features

Pain is felt in the groin but may radiate to the knee. Typically it occurs after periods of activity but later it aged to use a walking stick and to try to preserve movement and stability by non-weightbearing exercises. In early cases physiotherapy (including manipulation) may relieve pain for long periods. Activities are adjusted so as to reduce stress on the hip.

Operative treatment The indications for operation are

(1) progressive increase in pain,
(2) severe restriction of activities,
(3) marked deformity and
(4) progressive loss of movement (especially abduction), together with
(5) x-ray signs of joint destruction.

In the usual case – a patient aged over 60 years with a long history of pain and increasing disability – the preferred operation is *total joint replacement* (see below). In those between 40 and 60 years this may still be the best operation if joint destruction is severe.

In younger patients, particularly those with some preservation of articular cartilage, an *intertrochanteric realignment osteotomy* may be considered. If performed early, it can arrest or delay further cartilage destruction, and if the operation is well planned it does not preclude later replacement arthroplasty.

In recent years *osteochondroplasty* has gained attention following the realization that primary ' or 'idiopathic' OA of the hip is often associated with malposition or malcongruency of this ball-and-socket joint. This is discussed in the next section.

Arthrodesis of the hip is a practical solution for young adults with marked destruction of a singlejoint, and particularly when the conditions foradvanced reconstructive surgery are less than ideal. If well executed, the operation guarantees freedom from pain and permanent stability, though it has the dis - advantages of restricted mobility and a significant incidence of later backache, as well as deformity and discomfort in other nearby joints (Solomon, 1998).

The knee 20

CLINICAL ASSESSMENT
SYMPTOMS
Pain, either insidious in onset or more acute, is the most common knee symptom. With inflammatory or degenerative disorders it is usually diffuse, but with mechanical disorders (and especially after injury) it is often localized ‑ the patient can, and should, point to the painful spot. If the patient can describe the mechanism of the injury, this is extremely useful: a direct blow to the front of the knee may damage the patello-femoral joint; a blow to the side may rupture the collateral ligament; twisting injuries are more likely to cause a torn meniscus or a cruciate ligament rupture.
Swelling may be diffuse or localized. If there was aninjury, it is important to ask whether the swelling appeared immediately (suggesting a haemarthrosis) or only after some hours (typical of a torn meniscus). A complaint of recurrent swelling, with more or less normal periods in between, suggests a longstandinginternal derangement ‑ possibly an old meniscal tear, degeneration of the meniscus, a small osteoarticular fracture or loose bodies in the joint. Chronic swelling is typical of synovitis or arthritis. A small, localized swelling on the anteromedial or anterolateral side of the joint makes one think of a cyst of the meniscus (always on the medial side) or a floating loose body. Swelling over the front of the knee could be due to a prepatellar bursitis; a localized bulge in the popliteal fossa can also be caused by a bursal swelling, but is more often due to ballooning of the synovial membrane and capsule at the back of thejoint.

'Stiffness' is a common complaint, but it must be distinguished from inhibition of movement due to pain, or simple weakness of the extensor apparatus. Particularly characteristic is stiffness that appears regularly after periods of rest - so-called 'post-inactivity stiffness' - which suggests some type of chronic arthritis.
Locking is different from stiffness. The knee, quite suddenly, cannot be straightened fully, although flexion is still possible. This happens when a torn meniscus or loose body is caught between the articular surfaces. By wiggling the knee around, the patient may be able to 'unlock' it; sudden *unlocking* is the most reliable evidence that something mobile had previously obstructed full extension. Do not be misled by 'pseudo-locking', when movement is suddenly stopped by pain or the fear of impending pain.
Deformity is seldom a leading symptom; patients are not keen to admit to having 'knock knees' or 'bandy legs'. However, a unilateral deformity, especially if it is progressive, will be more worrying.
Giving way, a feeling of instability, or a lack of trust in the knee suggests a mechanical disorder caused by ligamentous, meniscal or capsular injury, or simple muscle weakness. Giving way, particularly if it occurs when climbing up or down stairs, may also be due to patello-femoral pain or instability. Excessive use of an unstable knee produces post-exercise swelling (effusionm or haemarthrosis) and diffuse pain within the joint.
Limp may be due to either pain or instability.
Loss of function manifests as a progressively diminishing walking distance, inability to run and difficulty going up and down steps. Squatting or kneeling may be painful, either because of pressure on the patellofemoral joint or because the knee cannot flex fully.

Move
Passive extension can be tested by the examiner simply holding both legs by the ankles and lifting them off the couch; the knees should straighten fully (or even into a few degrees of hyperextension) and symmetrically.
Active extension can be roughly tested by the examiner slipping a hand under each knee and then asking the patient to force the knees into the surface of the couch; it is usually easy to feel whether the hands are trapped equally strongly on the two sides. Another way is to have the patient sitting on the edge of the couch with his or her legs hanging over the side and then asking them to extend each knee as far as possible; the test can be repeated with the patient extending the knees against resistance.
Passive and *active flexion* are tested with the patient lying supine. Normally the heel can be pulled up close to the buttock, with the knee moving through a range of 0-150 degrees. The 'heel-to-buttock' distance is compared on the two sides.
Internal and *external rotation*, though normally no more than about 10 degrees, should also be assessed. The patient's hip and knee are flexed to 90 degrees; one hand steadies and feels the knee, the other rotates the foot.
Crepitus during movement may be felt with a hand placed on the front of the knee. It usually signifies patello-femoral roughness.
Movement with compartmental loading is a useful test for localizing the site of joint pain; the medial or lateral compartment of the knee can be loaded separately by applying varus or valgus stress during flexion and noting which manoeuvre is more painful.

<u>Tests for intra-articular fluid</u>
Cross fluctuation This test is applicable only if there is a large effusion. The left hand compresses and empties the suprapatellar pouch while the right hand straddles the front of the

joint below the patella; by squeezing with each hand alternately, a fluid impulse is transmitted across the joint.
The patellar tap Again the suprapatellar pouch is compressed with the left hand to squeeze any fluid from the pouch into the joint. With the other hand the patella is then tapped sharply backwards onto the femoral condyles. In a positive test the patella can be felt striking the femur and bouncing off again (a type of ballottement).
The bulge test This is a useful method of testing when there is very little fluid in the joint, though it takes some practice to get it right! After squeezing any fluid out of the suprapatellar pouch, the medial compartment is emptied by pressing on the inner aspect of the joint; that hand is then lifted away and the lateral side is sharply compressed – a distinct ripple is seen on the flattened medial surface as fluid is shunted across.
The juxta-patellar hollow test Normally, when the knee is flexed, a hollow appears lateral to the patellar ligament and disappears with further flexion; if there is excess fluid, the hollow fills and disappears at a lesser angle of flexion (Mann et al., 1991). Compare this in the two knees.

The patello-femoral joint
The *size, shape and position* of the patella are noted. The bone is felt, first on its anterior surface and then along its edges and at the attachments of the quadriceps tendon and the patellar ligament. Much of the posterior surface, too, is accessible to palpation if the patella is pushed first to one side and then to the other; tenderness suggests synovial irritation or articular cartilage softening. Moving the patella up and down while pressing it lightly against the femur (*the 'friction test'*) causes painful grating if the central portion of the articular cartilage is damaged. Pressing the patella laterally with the thumb while flexing the knee slowly may induce anxiety and sharp resistance to further movement; this, the *'apprehension test'*, is diagnostic of recurrent patellar subluxation or dislocation.

Tests for stability
Collateral ligaments The medial and lateral ligaments are tested by stressing the knee into valgus and varus: this is best done by tucking the patient's foot under your arm and holding the extended knee firmly with one hand on each side of the joint; the leg is then angulated alternately towards abduction and adduction. The test is performed at full extension and again at 30 degrees of flexion. There is normally some medio-lateral movement at 30 degrees, but if this is excessive (compared to the normal side) it suggests a torn or stretched collateral ligament. Sideways movement in full extension is always abnormal; it may be due to either torn or stretched ligaments and capsule or loss of articular cartilage or bone, which allows the affected compartment to collapse.
Cruciate ligaments Routine tests for cruciate ligament stability are based on examining for abnormal gliding movements in the sagittal plane. With both knees flexed 90 degrees and the feet resting on the couch, the upper tibia is inspected from the side; if its upper end has dropped back, or can be gently pushed back, this indicates a tear of the posterior cruciate ligament (the 'sag sign'). With the knee in the same position, the foot is anchored by the examiner sitting on it (provided this is not painful); then, using both hands, the upper end of the tibia is grasped firmly and rocked backwards and forwards to see if there is any anteroposterior glide (the '*drawer test'*). Excessive anterior movement (a positive anterior drawer sign) denotes anterior cruciate laxity; excessive posterior movement (a positive posterior drawer sign) signifies posterior cruciate laxity.

More sensitive is the ***Lachman test,*** but this is difficult if the patient has big thighs (or the examiner has small hands). The patient’ s knee is flexed 20 degrees; with one hand grasping the lower thigh and the other the upper part of the leg, the joint surfaces are shifted backwards and forwards upon each other. If the knee is stable, there should be no gliding. In both the drawer test and Lachman test, note whether the endpoint of abnormal movement is ‘soft’ or ‘hard’.

Complex ligament injuries When only a collateral or cruciate ligament is damaged the diagnosis is relatively easy: the direction of unstable movement is either sideways or front-to-back. With combined injuries the direction of instability may be oblique or rotational. Special clinical tests have been developed to detect these abnormalities (see Chapter 30); the best known is the *pivot shift test.* The patient lies supine with the lower limb completely relaxed. The examiner lifts the leg with the knee held in full extension and the tibia internally rotated (the position of slight rotational subluxation). A valgus force is then applied to the lateral side of the joint as the knee is flexed; a sudden posterior movement of the tibia is seen and felt as the joint is fully re-located. The test is sometimes quite painful.

Tests for meniscal injuries

McMurray’s test This classic test for a torn meniscus is seldom used now that the diagnosis can easily be made by MRI. However, advanced imaging is not always available and the clinical test has not been altogether discarded. The test is based on the fact that the loose meniscal tag can sometimes be trapped between the articular surfaces and then induced to snap free with a palpable and audible click. The knee is flexed as far as possible; one hand steadies the joint and the other rotates the leg medially and laterally while the knee is slowly extended. The test is repeated several times, with the knee stressed in valgus or varus, feeling and listening for the click. A positive test is helpful but not pathognomonic; a negative test does not exclude a tear.

Thessaly test This test is based on a dynamic reproduction of load transmission in the knee joint under normal or trauma conditions. With the affected knee flexed to 20 degrees and the foot placed flat on the ground, the patient takes his or her full weight on that leg while being supported (for balance) by the examiner (Fig. 20.9). The patient is then instructed to twist his or her body to one side and then to the other three times (thus, with each turn, exerting a rotational force in the knee) while keeping the knee flexed at 20 degrees. Patients with meniscal tears experience medial or lateral joint line pain and may have a sense of locking. The test has shown a high diagnostic accuracy rate at the level of 95 per cent in detecting meniscal tears, with a low number of false positive and negative recordings (Karachalios et al., 2005).

THE DIAGNOSTIC CALENDAR

While most disorders of the knee can occur at any age, certain conditions are more commonly encountered during specific periods of life.

Congenital knee disorders may be present at birth or may become apparent only during the first or second decade of life.

Adolescents with anterior knee pain are usually found to have chondromalacia patellae, patellar instability,osteochondritis or a plica syndrome. But remember‐knee pain may be referred from the hip!

Young adults engaged in sports are the most frequent victims of meniscal tears and ligament injuries. Examination should include a variety of tests for ligamentous instability that would be quite inappropriate in elderly patients.

Patients above middle age with chronic pain and stiffness probably have osteoarthritis. With primary osteoarthritis of the knees, other joints also are often affected; polyarthritis does not necessarily (nor even most commonly) mean rheumatoid arthritis.

DEFORMITIES OF THE KNEE

By the end of growth the knees are normally in 5–7 degrees of valgus. Any deviation from this may be regarded as 'deformity', though often it bothers no one – least of all the possessor of the knees. The three common deformities are bow leg (genu varum), knock knee (genu valgum) and hyperextension (genu recurvatum).

BOW LEGS AND KNOCK KNEES IN CHILDREN

Deformity is usually gauged from simple observation. Bilateral bow leg can be recorded by measuring thedistance between the knees with the child standing and the heels touching; it should be less than 6 cm. Similarly, knock knee can be estimated by measuring the distance between the medial malleoli when the knees are touching with the patellae facing forwards; it is usually less than 8 cm.

DEFORMITIES OF THE KNEE IN ADULTS

GENU VARUM AND GENU VALGUM

Angular deformities are common in adults (usually bow legs in men and knock knees in women). They may be the *sequel to childhood deformity* and if so usually cause no problems. However, if the deformity is associated with joint instability, this can lead to osteoarthritis – of the medial compartment in varus knees and the lateral compartment in valgus knees. Genu valgum may also cause abnormal tracking of the patella and predispose to patello-femoral osteoarthritis. Even in the absence of overt osteoarthritis, if the patient complains of severe pain, or if there are clinical or radiological signs of joint damage, a 'prophylactic osteotomy can be performed – above the knee for valgus deformity and below the knee for varus. Preoperative planning should include radiographic measurements to determine the mechanical and anatomical axes of both bones and the lower limb, as well as estimation of the centre of rotation of angulation. Deformity may be *secondary to arthritis* – usually varus in osteoarthritis and valgus in rheumatoid arthritis. In these cases the joint is often unstable and corrective osteotomy less predictable in its effect. Stress x-rays are essential in the assessment of these cases. Other causes of varus or valgus deformity are *ligament injuries, malunited fractures* and *Paget's disease*. Where possible, the underlying disorder should be dealt with; provided the joint is stable, corrective osteotomy may be all that is necessary.

GENU RECURVATUM (HYPEREXTENSION OF THE KNEE)

Congenital recurvatum This may be due to abnormal intra-uterine posture; it usually recovers spontaneously. Rarely, gross hyperextension is the precursor of true congenital dislocation of the knee.

LESIONS OF THE MENISCI

The menisci have an important role in (1) improving articular congruency and increasing the stability of the knee, (2) controlling the complex rolling and gliding actions of the joint and (3) distributing load during movement. During weightbearing, at least 50 per cent of the contact stresses are taken by the menisci when the knee is loaded in extension, rising to

almost 90 per cent with the knee in flexion. If the menisci are removed, articular stresses are markedly increased; even a partial meniscectomy of one-third of the width of the meniscus will produce a threefold increase in contact stress in that area. The medial meniscus is much less mobile than the lateral, and it cannot as easily accommodate to abnormal stresses. This may be why meniscal lesions are more common on the medial side than on the lateral. Even in the absence of injury, there is gradual stiffening and degeneration of the menisci with age, so splits and tears are more likely in later life – particularly if there is any associated arthritis or chondrocalcinosis. In young people, meniscal tears are usually the result of trauma.

TEARS OF THE MENISCUS

The meniscus consists mainly of circumferential fibres held by a few radial strands. It is, therefore, more likely to tear along its length than across its width. The split is usually initiated by a rotational grinding force, which occurs (for example) when the knee is flexed and twisted while taking weight; hence the frequency in footballers. In middle life, when fibrosis has restricted mobility of the meniscus, tears occur with relatively little force.

Clinical features

The patient is usually a young person who sustains a twisting injury to the knee on the sports field. Pain (usually on the medial side) is often severe and further activity is avoided; occasionally the knee is 'locked' in partial flexion. Almost invariably, swelling appears some hours later, or perhaps the following day. With rest the initial symptoms subside, only to recur periodically after trivial twists or strains. Sometimes the knee gives way spontaneously and this is again followed by pain and swelling. It is important to remember that in patients aged over 40 the initial injury may be unremarkable and the main complaint is of recurrent 'giving way' or 'locking'. 'Locking' – that is, the sudden inability to extend the knee fully – suggests a bucket-handle tear. The patient sometimes learns to 'unlock' the knee by bending it fully or by twisting it from side to side. On examination the joint may be held slightly flexed and there is often an effusion. In longstanding cases the quadriceps will be wasted. Tenderness is localized to the joint line, in the vast majority of cases on the medial side. Flexion is usually full but extension is often slightly limited. Between attacks of pain and effusion there is a disconcerting paucity of signs. The history is helpful, and McMurray's test, Apley's grinding test or the Thessaly test may be positive.

Investigations

Plain x-rays are usually normal, but *MRI* is a reliable method of confirming the clinical diagnosis, and may even reveal tears that are missed by arthroscopy.

Arthroscopy has the advantage that, if a lesion is identified, it can be treated at the same time.

Differential diagnosis

Loose bodies in the joint may cause true locking. The history is much more insidious than with meniscal tears and the attacks are variable in character and intensity. A loose body may be palpable and is often visible on x-ray.

Recurrent dislocation of the patella causes the knee to give way; typically the patient is caught unawares and collapses to the ground. Tenderness is localized to the medial edge of the patella and the apprehension test is positive.

Fracture of the tibial spine follows an acute injury and may cause a block to full extension. However, swelling is immediate and the fluid is blood-stained. X-ray may show the fracture.

A partial tear of the medial collateral ligament may heal with adhesions where it is attached to the medial meniscus, so that the meniscus loses mobility. The patient complains of recurrent attacks of pain and giving way, followed by tenderness on the medial side. Sleep may be disturbed if the medial side rests upon the other knee or the bed. As with a meniscus injury, rotation is painful; but unlike a meniscus lesion, the grinding test gives less pain and the distraction test more pain.

A torn anterior cruciate ligament can cause chronic instability, with a sense of the knee ‘giving way’ or buckling when the patient turns sharply towards the side of the affected knee. Careful examination should reveal signs of rotational instability, a positive Lachman test or a positive anterior drawer sign. MRI or arthroscopy will settle any doubts.

Treatment

Dealing with the locked knee Usually the knee ‘unlocks’ spontaneously; if not, gentle passive flexion and rotation may do the trick. Forceful manipulation is unwise (it may do more damage) and is usually unnecessary; after a few days’ rest the knee may well unlock itself. However, if the knee does not unlock, or if attempts to unlock it cause severe pain, arthroscopy is indicated. If symptoms are not marked, it may be better to wait a week or two and let the synovitis settle down, thus making the operation easier; if the tear is confirmed, the offending fragment is removed.

Conservative treatment If the joint is not locked, it is reasonable to hope that the tear is peripheral and can therefore heal spontaneously. After an acute episode, the joint is held straight in a plaster backslab for 3–4 weeks; the patient uses crutches and quadriceps exercises are encouraged. Operation can be put off as long as attacks are infrequent and not disabling and the patient is willing to abandon those activities that provoke them. MRI will show if the meniscus has healed.

Operative treatment Surgery is indicated (1) if the joint cannot be unlocked and (2) if symptoms are recurrent. For practical purposes, the lesion is often dealt with as part of the ‘diagnostic’ arthroscopy. Tears close to the periphery, which have the capacity to heal, can be sutured; at least one edge of the tear should be red (i.e. vascularized). In appropriate cases the success rate for both open and arthroscopic repair is almost 90 per cent. Tears other than those in the peripheral third are dealt with by excising the torn portion (or the bucket handle). Total meniscectomy is thought to cause more instability and so predispose to late secondary osteoarthritis; certainly in the short term it causes greater morbidity than partial meniscectomy and has no obvious advantages. Arthroscopic meniscectomy has distinct advantages over open meniscectomy: shorter hospital stay, lower costs and more rapid return to function. However, it is by no means free of complications (Sherman et al., 1986). Postoperative pain and stiffness are reduced by prophylactic non-steroidal anti-inflammatory drugs. Quadriceps-strengthening exercises are important.

Outcome

Neither a meniscal tear by itself nor removal of the meniscus necessarily leads to secondary osteoarthritis. However, the likelihood is increased if the patient has (a) a pre-existing varus deformity of the knee, (b) signs of cruciate ligament insufficiency or (c) features elsewhere of a generalized osteoarthritis.

MENISCAL DEGENERATION

Patients over 45 years old may present with symptoms and signs of a meniscal tear. Often, though, they can recall no preceding injury. At arthroscopy there may be a horizontal cleavage in the medial meniscus – the characteristic ‘degenerative’ lesion – or detachment of the anterior or posterior horn without an obvious tear. Associated osteoarthritis or chrondrocalcinosis is common. A detached anterior or posterior horn can be sutured firmly in place. Meniscectomy is indicated only if symptoms are marked or if, at arthroscopy, there is a major tear causing mechanical block.

DISCOID LATERAL MENISCUS

In the fetus the meniscus is not semilunar but disclike; if this shape persists, symptoms are likely. A young patient complains that, without any history of injury, the knee gives way and ‘thuds’ loudly. A characteristic clunk may be felt at 110 degrees as the knee is bent and at

10 degrees as it is being straightened. The diagnosis is easily confirmed by MRI. If there is only a clunk, treatment is not essential. If pain is disturbing, the meniscus may be excised, though a more attractive procedure is arthroscopic partial excision leaving a normally shaped meniscus (Dimakopoulos and Patel, 1990).

MENISCAL CYSTS

Cysts of the menisci are probably traumatic in origin, arising from either a small horizontal cleavage tear or repeated squashing of the peripheral part of the meniscus. It is also suggested that synovial cells infiltrate into the vascular area between meniscus and capsule and there multiply. The multilocular cyst contains gelatinous fluid and is surrounded by thick fibrous tissue.

Clinical features

The lateral meniscus is affected much more frequently than the medial. The patient complains of an ache or a small lump at the side of the joint. Symptoms may be intermittent, or worse after activity. On examination the lump is situated at or slightly below the joint line, usually anterior to the collateral ligament. It is seen most easily with the knee slightly flexed; in some positions it may disappear altogether. Lateral cysts are often so firm that they are mistaken for a solid swelling. Medial cysts are usually larger and softer.

Differential diagnosis

Apart from cysts, various conditions may present with a small lump along the joint line.

A *ganglion* is quite superficial, usually not as 'hard' as a cyst, and unconnected with the joint.

Calcific deposits in the collateral ligament usually appear on the medial side, are intensely painful and tender, and often show on the x-ray.

A prolapsed, torn meniscus occasionally presents as a rubbery, irregular lump at the joint line. In some cases the distinction from a 'cyst' is largely academic.

Various tumours, both of soft tissue (lipoma, fibroma) and of bone (osteochondroma), may produce a medial or lateral joint lump. Careful examination will show that the lump does not arise from the joint itself.

Treatment

If the symptoms warrant operation, the cyst may be removed. In the past this was usually combined with total meniscectomy, in order to prevent an inevitable recurrence of the cyst. However, it is quite feasible to examine the meniscus by arthroscopy, remove only the torn or damaged portion and then decompress the cyst from within the joint. The recurrence rate following such arthroscopic surgery is negligible (Parisien, 1990).

CHRONIC LIGAMENTOUS INSTABILITY

The knee is a complex hinge which depends heavily on its ligaments for medio-lateral, anteroposterior and rotational stability. Ligament injuries, from minor strains through partial ruptures to complete tears, are common in sportsmen, athletes and dancers. Whatever the nature of the acute injury, the victim may be left with chronic instability of the knee – a sense of the joint wanting to give way, or actually giving way, during unguarded activity. Sometimes this is accompanied by pain and recurrent episodes of swelling. There may be a meniscal tear, but meniscectomy is likely to make matters worse; sometimes patients present with meniscectomy scars on both sides of the knee!

Examination should include special tests for ligamentous instability as well as radiological investigation and arthroscopy. It is important not only to establish the nature of the lesion but also to measure the level of functional impairment against the needs and demands of the individual patient before advocating treatment.

RECURRENT DISLOCATION OF THE PATELLA

In 15–20 per cent of cases (mostly children) the first episode is followed by recurrent dislocation or subluxation after minimal stress. This is due, in some measure, to disruption or stretching of the ligamentous structures which normally stabilize the extensor mechanism. However, in a significant proportion of cases there is no history of an acute strain and the initial episode is thought to have occurred 'spontaneously'. It is now recognized that in all cases of recurrent dislocation, but particularly in the latter group, one or more *predisposing factors* are often present: (1) generalized ligamentous laxity; (2) under - development of the lateral femoral condyle and flattening of the intercondylar groove; (3) maldevelopment of the patella, which may be too high or too small; (4) valgus deformity of the knee; (5) external tibial torsion; or (6) a primary muscle defect. Repeated dislocation damages the contiguous articular surfaces of the patella and femoral condyle; this may result in further flattening of the condyle, so facilitating further dislocations. Dislocation is almost always towards the lateral side; medial dislocation is seen only in rare iatrogenic cases following overzealous lateral release or medial transposition of the patellar tendon.

PATELLO-FEMORAL PAIN SYNDROME (CHONDROMALACIA OF THE PATELLA; PATELLOFEMORAL OVERLOAD SYNDROME)

There is no clear consensus concerning the terminology, aetiology and treatment of pain and tenderness in the anterior part of the knee. This syndrome is common among active adolescents and young adults. It is often (but not invariably) associated with softening and fibrillation of the articular surface of the patella – *chondromalacia patellae.* Having no other pathological label, orthopaedic surgeons have tended to regard chondromalacia as the cause (rather than one of the effects) of the disorder. Against this are the facts that (1) chondromalacia is commonly found at arthroscopy in young adults who have no anterior knee pain, and (2) some patients with the typical clinical syndrome have no cartilage softening.

OSTEOCHONDRITIS DISSECANS

The prevalence of osteochondritis dissecans is between 15 and 30 per 100 000 with males being affected more often than females (ratio 5:3). An increase in its incidence has been observed in recentyears, probably due to the growing participation of young children of both genders in competitive sports.

A small, well-demarcated, avascular fragment ofbone and overlying cartilage sometimes separates from one of the femoral condyles and appears as a loose body in the joint. The most likely cause is trauma, either a single impact with the edge of the patella or repeated microtrauma from contact with an adjacent tibial ridge. The fact that over 80 per cent of lesions occur on the lateral part of the medial femoral condyle, exactly where the patella makes contact in full flexion, supports the first of these. There may also be some general predisposing factor, because several joints can be affected, or several members of one family. Lesions are bilateral in 25 per cent of cases.

Pathology

The lower, lateral surface of the medial femoral condyle is usually affected, rarely the lateral condyle,

and still more rarely the patella. An area of subchondral bone becomes avascular and within this area an ovoid osteocartilaginous segment is demarcated from the surrounding bone. At first the overlying cartilage is intact and the fragment is stable; over a period of months the fragment separates but remains in position; finally the fragment breaks free to become a loose body in the joint. The small crater is slowly filled with fibrocartilage, leaving a depression on the articular surface.

Classification

Osteochondritis dissecans of the knee is classified according to anatomical location, arthroscopic appearance, scintigraphic or MRI findings and chronological age. For prognostic and treatment purposes it is divided into juvenile and adult forms, either stable or unstable (Kocher et al., 2006).

Clinical features

The patient, usually a male aged 15–20 years, presents with intermittent ache or swelling. Later, there are attacks of giving way such that the knee feels unreliable;"locking' sometimes occurs. The quadriceps muscle is wasted and there may be a small effusion. Soon after an attack there are two signs that are almost diagnostic: (1) tenderness localized to one femoral condyle; and (2) Wilson's sign: if the knee is flexed to 90 degrees, rotated medially and then gradually straightened, pain is felt; repeating the test with the knee rotated laterally is painless.

Imaging

Plain x-rays may show a line of demarcation around a lesion in situ, usually in the lateral part of the medial femoral condyle. This site is best displayed in special intercondylar (tunnel) views, but even then a small lesion or one situated far back may be missed. Once the fragment has become detached, the empty hollow may be seen – and possibly a loose body elsewhere in the joint.

Radionuclide scans show increased activity around the lesion, and *MRI* consistently shows an area of low signal intensity in the T1 weighted images; the adjacent bone may also appear abnormal, probably due to oedema. These investigations usually indicate whether the fragment is 'stable' or 'loose'. MRI may also allow early prediction of whether the lesion will heal or not.

Arthroscopy

With early lesions the articular surface looks intact, but probing may reveal that the cartilage is soft. Loose segments are easily visualized.

Differential diagnosis

Avascular necrosis of the femoral condyle – usually associated with corticosteroid therapy or alcohol abuse – may result in separation of a localized osteocartilaginous fragment. However, it is seen in an older age group and on x-ray the lesion is always on the dome of the femoral condyle, and this distinguishes it from osteochondritis dissecans.

Treatment

For the purposes of management, it is useful to 'stage' the lesion; hence the importance of radionuclide scanning, MRI and arthroscopy. Lesions in adults have a greater propensity to instability whereas juvenile osteochondritis is typically stable. Those lesions with an intact articular surface have the greatest potential to heal with non-operative treatment if repetitive impact loading is avoided.

In the earliest stage, when the cartilage is intact and the lesion is 'stable', no treatment is needed but activities are curtailed for 6–12 months. Small lesions often heal spontaneously.

If the fragment is 'unstable', i.e. surrounded by a clear boundary with radiographic 'sclerosis' of the underlying bone, or showing MRI features of separation, treatment will depend on the size of the lesion. A small fragment should be removed by arthroscopy and the base drilled; the bed will eventually be covered by fibrocartilage, leaving only a small defect. A large fragment (say more than 1 cm in diameter) should be fixed in situ with pins or Herbert screws. In addition, it may help to drill the underlying sclerotic bone to promote union of the necrotic fragment. For drilling, the area is approached from a point some distance away, beyond the articular cartilage.

If the fragment is completely detached but in one piece and shown to fit nicely in its bed, the crater is cleaned and the floor drilled before replacing the loose fragment and fixing it with

Herbert screws. If the fragment is in pieces or ill-shaped, it is best discarded; the crater is drilled and allowed to fill with fibrocartilage. In recent years attempts have been made to fill the residual defects by articular cartilage transplantation: either the insertion of osteochondral plugs harvested from another part of the knee or the application of sheets of cultured chondrocytes. This approach should still be regarded as in the ‘experimental’ stage. After any of the above operations the knee is held in a cast for 6 weeks; thereafter movement is encouraged but weightbearing is deferred until x-rays show signs of healing.

LOOSE BODIES

The knee – relatively capacious, with large synovial folds – is a common haven for loose bodies. These may be produced by: (1) injury (a chip of bone or cartilage); (2) osteochondritis dissecans (which may produce one or two fragments); (3) osteoarthritis (pieces of cartilage or osteophyte); (4) Charcot’s disease (large osteocartilaginous bodies); and (5) synovial chondromatosis (cartilage metaplasia in the synovium, sometimes producing hundreds of loose bodies).

OSTEOARTHRITIS

The knee is the commonest of the large joints to be affected by osteoarthritis (see Chapter 5). Often there is a predisposing factor: injury to the articular surface, a torn meniscus, ligamentous instability or preexisting deformity of the hip or knee, to mention a few. However, in many cases no obvious cause can be found. Underlying all of these, there may also be a genetic component. Curiously, while the male:female distribution is more or less equal in white (Caucasian) peoples, black African women are affected far more frequently than their male counterparts. Osteoarthritis is often bilateral and in these cases there is a strong association with Heberden’s nodes and generalized osteoarthritis.

Clinical features

Patients are usually over 50 years old; they tend to be overweight and may have longstanding bow-leg deformity. Pain is the leading symptom, worse after use, or (if the patello-femoral joint is affected) on stairs. After rest, the joint feels stiff and it hurts to ‘get going’ after sitting for any length of time. Swelling is common, and giving way or locking may occur. On examination there may be an obvious deformity (usually varus) or the scar of a previous operation. The quadriceps muscle is usually wasted. Except during an exacerbation, there is little fluid and no warmth; nor is the synovial membrane thickened. Movement is somewhat limited and is often accompanied by patello-femoral crepitus. It is useful to test movement applying first a varus and then a valgus force to the knee; pain indicates which tibio-femoral compartment is involved. Pressure on the patella may elicit pain. The natural history of osteoarthritis is one of alternating ‘bad spells’ and ‘good spells’. Patients may experience long periods of lesser discomfort and only moderate loss of function, followed by exacerbations of pain and stiffness (perhaps after unaccustomed activity).

X-ray

The anteroposterior x-ray *must* be obtained with the patient standing and bearing weight; only in this way can small degrees of articular cartilage thinning be revealed. The tibio-femoral joint space is diminished (often only in one compartment) and there is subchondral sclerosis. Osteophytes and subchondral cysts are usually present and sometimes there is soft-tissue calcification in the suprapatellar region or in the joint itself (chondrocalcinosis). If only the patello-femoral joint is affected, suspect a pyrophosphate arthropathy.

Treatment

If symptoms are not severe, treatment is conservative. Joint loading is lessened by using a walking stick. Quadriceps exercises are important. Analgesics are prescribed for pain, and warmth (e.g. radiant heat or shortwave diathermy) is soothing. A simple elastic support may do wonders, probably by improving proprioception in an unstable knee. Intra-articular corticosteroid injections will often relieve pain, but this is a stopgap, and not a very good one, because repeated injections may permit (or even predispose to) progressive cartilage and bone destruction. New forms of medication have been introduced in recent years, particularly the oral administration of glucosamine and intra-articular injection of hyalourans. There is, as yet, no agreement about the long-term efficacy of these products.

OPERATIVE TREATMENT

Persistent pain unresponsive to conservative treatment, progressive deformity and instability are the usual indications for operative treatment.

Arthroscopic washouts, with trimming of degenerate meniscal tissue and osteophytes, may give temporary relief; this is a useful measure when there are contraindications to reconstructive surgery.

Patellectomy is indicated only in those rare cases where osteoarthritis is strictly confined to the patellofemoral joint. However, bear in mind that extensor power will be reduced and if a total joint replacement is later needed pain relief will be less predictable than usual (Paletta and Laskin, 1995).

Realignment osteotomy is often successful in relieving symptoms and staving off the need for ‘end-stage’ surgery. The ideal indication is a ‘young’ patient (under 50 years) with a varus knee and osteoarthritis confined to the medial compartment: a high tibial valgus osteotomy will redistribute weight to the lateral side of the joint. The degree and accuracy of angular correction are the most important determinants of mid- and long-term clinical outcome.

Replacement arthroplasty is indicated in older patients with progressive joint destruction. This is usually a ‘resurfacing’ procedure, with a metal femoral condylar component and a metal-backed polyethylene table on the tibial side. If the disease is largely confined to one compartment, a unicompartmental replacement can be done as an alternative to osteotomy. With modern techniques, and meticulous attention to anatomical alignment of the knee, the results of replacement arthroplasty are excellent.

Arthrodesis is indicated only if there is a strong contraindication to arthroplasty (e.g. previous infection) or to salvage a failed arthroplasty.

OSTEONECROSIS

Osteonecrosis of the knee, though not as common as femoral head necrosis, has the same aetiological and pathogenetic background (see Chapter 6). The usual site is the dome of one of the femoral condyles, but occasionally the medial tibial condyle is affected. Two main categories are identified:

(1) *osteonecrosis associated with a definite background disorder* [e.g. corticosteroid therapy, alcohol abuse, sickle-cell disease, hyperbaric decompression sickness, systemic lupus erythematosus (SLE) or Gaucher's disease], and

(2) '*spontaneous' osteonecrosis* of the knee, popularly known by the acronym SONK, which is due to a small insufficiency fracture of a prominent part of the osteoarticular surface in osteoporotic bone; the vascular supply to the free fragment is compromised (Yamamoto and Bullough, 2000).

A third type, *postmeniscectomy osteonecrosis,* has been reported; its prevalence and pathophysiology are still unclear (Patel et al., 1998).

Treatment

Treatment is conservative in the first instance and consists of measures to reduce loading of the joint and analgesics for pain. If symptoms or signs increase, operative treatment may be considered. Surgical options include arthroscopic debridement, drilling with or without bone grafting, core decompression of the femoral condyle at a distance from the lesion, and (for patients with persistent symptoms and well-marked articular surface damage) a valgus osteotomy or unicompartmental arthroplasty. Resurfacing with osteochondral allografts has also been employed, with variable results.

RUPTURES OF THE EXTENSOR APPARATUS

Resisted extension of the knee may tear the extensor mechanism. The patient stumbles on a stair, catches his or her foot while walking or running, or may only be kicking a muddy football. In all these incidents, active knee extension is prevented by an obstacle. The precise location of the lesion varies with the patient's age. In the elderly the injury is usually above the patella; in middle life the patella fractures; in young adults the patellar ligament can rupture. In adolescents the upper tibial apophysis is occasionally avulsed; much more often it is merely 'strained'. Tendon rupture sometimes occurs with minimal strain; this is seen in patients with connective tissue disorders (e.g. SLE) and advanced rheumatoid disease, especially if they are also being treated with corticosteroids.

RUPTURE ABOVE THE PATELLA

Rupture may occur in the belly of the rectus femoris. The patient is usually elderly, or on long-term corticosteroid treatment. The torn muscle retracts and forms a characteristic lump in the thigh. Function is usually good, so no treatment is required. Avulsion of the quadriceps tendon from the upper pole of the patella is seen in the same group of people. Sometimes it is bilateral. Operative repair is essential.

RUPTURE BELOW THE PATELLA

This occurs mainly in young people. The ligament may rupture or may be avulsed from the lower pole of the patella. Operative repair is necessary. Pain and tenderness in the middle portion of the patellar ligament may occur in athletes; CT or ultrasonography will reveal an abnormal area. If rest fails to provide relief the paratenon should be stripped (King et al., 1990). Partial rupture or avulsion sometimes leads to a traction tendinitis and calcification in the patellar ligament‐ the *Sinding‐Larsen Johansson syndrome* (see below).

OSGOOD–SCHLATTER DISEASE

(‘APOPHYSITIS’ OF THE TIBIAL TUBERCLE)

In this common disorder of adolescence the tibial tubercle becomes painful and ‘swollen’. Although often called osteochondritis or apophysitis, it is nothing more than a traction injury of the apophysis into which part of the patellar tendon is inserted (the remainder is inserted on each side of the apophysis and prevents complete separation). There is no history of injury and sometimes the condition is bilateral. A young adolescent complains of pain after activity, and of a lump. The lump is tender and its situation over the tibial tuberosity is diagnostic. Sometimes active extension of the knee against resistance is painful and x-rays may reveal fragmentation of the apophysis. Spontaneous recovery is usual but takes time, and it is wise to restrict such activities as cycling, jumping and soccer. Occasionally, symptoms persist and, if patience or wearing a back-splint during the day are unavailing, a separate ossicle in the tendon is usually responsible; its removal is then worthwhile.

TENDINITIS AND CALCIFICATION AROUND THE KNEE CALCIFICATION IN THE MEDIAL LIGAMENT

Acute pain in the medial collateral ligament may be due to a soft calcific deposit among the fibres of the ligament. There may be a small, exquisitely tender lump in the line of the ligament. Pain is dramatically relieved by operative evacuation of the deposit.

PELLEGRINI–STIEDA DISEASE

X-rays sometimes show a plaque of bone lying next to the femoral condyle under the medial collateral ligament. Occasionally this is a source of pain. It is generally ascribed to ossification of a haematoma following a tear of the medial ligament, though a history of injury is not always forthcoming. Treatment is rarely needed.

PATELLAR ‘TENDINOPATHY’ (SINDING–LARSEN JOHANSSON SYNDROME).

This condition was described independently by Sinding- Larsen in 1921 and Johansson in 1922. Following a strain or partial rupture of the patellar ligament the patient (usually a young athletic individual) develops a traction ‘tendinitis’ characterised by pain and point tenderness at the lower pole of the patella. Sometimes, if the condition does not settle, calcification appears in the ligament (Medlar and Lyne, 1978). CT or ultrasonography may reveal the abnormal area in the ligament. A similar disorder has been described at the proximal pole of the patella. The condition is comparable to Osgood-Schlatter’s disease and usually recovers spontaneously. If rest fails to provide relief, the abnormal area is removed and the paratenon stripped (King et al., 1990; Khan et al., 1998).

ACUTE SWELLING OF THE ENTIRE JOINT POST-TRAUMATIC HAEMARTHROSIS

Swelling immediately after injury means blood in the joint. The knee is very painful and it feels warm, tense and tender. Later there may be a ‘doughy’ feel. Movements are restricted. X-rays are essential to see if there is a fracture; if there is not, then suspect a tear of the anterior cruciate ligament. The joint should be aspirated under aseptic conditions. If a ligament injury is suspected, examination under anaesthesia is helpful and may indicate the need for operation; otherwise a crepe bandage is applied and the leg cradled in a back-splint. Quadriceps exercises are practised from the start. The patient may get up when comfortable, retaining the back-splint until muscle control returns.

BLEEDING DISORDERS

In patients with clotting disorders, the knee is the most common site for acute bleeds. If the appropriate clotting factor is available, the joint should be aspirated and treated as for a traumatic haemarthrosis. If the factor is not available, aspiration is best avoided; the knee is splinted in slight flexion until the swelling subsides.

ACUTE SEPTIC ARTHRITIS

Acute pyogenic infection of the knee is not uncommon. The organism is usually *Staphylococcus aureus*, but in adults gonococcal infection is almost as common. The joint is swollen, painful and inflamed; the white cell count and ESR are elevated. Aspiration reveals pus in the joint; fluid should be sent for bacteriological investigation, including anaerobic culture. Treatment consists of systemic antibiotics and drainage of the joint – ideally by arthroscopy, with irrigation and complete synovectomy; if fluid reaccumulates, it can be aspirated through a wide-bore needle. As the inflammation subsides, movement is begun, but weightbearing is deferred for 4–6 weeks.

TRAUMATIC SYNOVITIS

Injury stimulates a reactive synovitis; typically the swelling appears only after some hours, and subsides spontaneously over a period of days. There is inhibition of quadriceps action and the thigh wastes. The knee may need to be splinted for several days but movement should be encouraged and quadricepsexercise is essential. If the amount of fluid is considerable, its aspiration hastens muscle recovery. In addition, any internal injury will need treatment.

ASEPTIC NON-TRAUMATIC SYNOVITIS

Acute swelling, without a history of trauma or signs of infection, suggests *gout* or *pseudogout*. Aspiration will provide fluid which may look turbid, resembling pus, but it is sterile and microscopy (using polarized light) reveals the crystals. Treatment with anti-inflammatory drugs is usually effective.

CHRONIC SWELLING OF THE JOINT

The diagnosis can usually be made on clinical andx-ray examination. The more elusive disorders should be fully investigated by joint aspiration, synovial fluid examination, arthroscopy and synovial biopsy.

ARTHRITIS

The commonest causes of chronic swelling are *osteoarthritis* and *rheumatoid arthritis*. Other signs, such as deformity, loss of movement or instability, may be present and x-ray examination will usually show characteristic features.

SYNOVIAL DISORDERS

Chronic swelling and synovial effusion without articular destruction should suggest conditions such as *synovial chondromatosis* and *pigmented villonodular synovitis*. The diagnosis will usually be obvious on arthroscopy and can be confirmed by synovial biopsy. The most important condition to exclude is *tuberculosis*. There has been a resurgence of cases during the last ten years and the condition should be seriouslyconsidered whenever there is no obvious alternative diagnosis. Investigations should include Mantoux testing and synovial biopsy. The ideal is to start antituberculous chemotherapy before joint destruction occurs.

SWELLINGS IN FRONT OF THE JOINT

PREPATELLAR BURSITIS (‘HOUSEMAID’S KNEE’)

The fluctuant swelling is confined to the front of the patella and the joint itself is normal. This is an uninfected bursitis due not to pressure but to constant friction between skin and bone. It is seen mainly in carpet layers, paving workers, floor cleaners and miners who do not use protective knee pads. Treatment consists of firm bandaging, and kneeling is avoided; occasionally aspiration is needed. In chronic cases the lump is best excised. Infection

(possibly due to foreign body implantation) results in a warm, tender swelling. Treatment is by rest, antibiotics and, if necessary, aspiration or excision.

INFRAPATELLAR BURSITIS (‘CLERGYMAN’S KNEE’)

The swelling is below the patella and superficial to the patellar ligament, being more distally placed than prepatellar bursitis; it used to be said that one who prays kneels more uprightly than one who scrubs! Treatment is similar to that for prepatellar bursitis. Occasionally the bursa is affected in gout.

OTHER BURSAE

Occasionally a bursa deep to the patellar tendon or the pes anserinus becomes inflamed and painful. Treatment is non-operative.

SWELLINGS AT THE BACK OF THE KNEE

SEMIMEMBRANOSUS BURSA

The bursa between the semimembranosus and the medial head of gastrocnemius may become enlarged in children or adults. It presents usually as a painless lump behind the knee, slightly to the medial side of the midline and most conspicuous with the knee straight. The lump is fluctuant but the fluid cannot be pushed into the joint, presumably because the muscles compress and obstruct the normal communication. The knee joint is normal. Occasionally the lump aches, and if so it may be excised through a transverse incision. However, recurrence is common and, as the bursa normally disappears in time, a waiting policy is perhaps wiser.

POPLITEAL ‘CYST’

Bulging of the posterior capsule and synovial herniation may produce a swelling in the popliteal fossa. The lump, which is usually seen in older people, is in the midline of the limb and at or below the level of the joint. It fluctuates but is not tender. Injection of radio-opaque medium into the joint, and x-ray, will show that the ‘cyst’ communicates with the joint. The condition was originally described by Baker, whose patients were probably suffering from tuberculous synovitis. Nowadays it is more likely to be caused by rheumatoid or osteoarthritis, but it is still often called a ‘Baker’s cyst’. Occasionally the ‘cyst’ ruptures and the synovial contents spill into the muscle planes causing pain and swelling in the calf‑a combination which can easily be mistaken for deep vein thrombosis. The swelling may diminish following aspiration and injection of hydrocortisone; excision is not advised, because recurrence is common unless the underlying condition is treated.

POPLITEAL ANEURYSM

This is the commonest limb aneurysm and is sometimes bilateral. Pain and stiffness of the knee may precede the symptoms of peripheral arterial disease, so it is essential to examine any lump behind the knee for pulsation. A thrombosed popliteal aneurysm does not pulsate, but it feels almost solid.

The ankle and foot 21

CLINICAL ASSESSMENT

SYMPTOMS

Adults with foot and ankle problems often present complaining of pain, swelling, deformity and impaired function including difficulties with work, social and domestic activities. Questions should include those that flag up the possibility of neoplastic or generalized inflammatory disease and diabetes.

Pain over a bony prominence or a joint is probably due to some local disorder; ask the patient to point to the painful spot. Symptoms tend to be well localized to the structures involved, but vague pain across the forefoot (*metatarsalgia*) is less specific and is often associated with uneven loading and muscle fatigue. Often the main complaint is of shoe pressure on a tender corn over a toe joint or a callosity on the sole. Osteoarthritic pain at the first metatarsophalangeal (MTP) joint is often better in firm-soled shoes; hallux valgus/bunions will be exacerbated by close-fitting shoes; a functionally or mechanically unstable ankle often feels better in boots; metatarsalgia is worse in shoes with a higher heel. Morton's neuroma or a prominent metatarsal head feels like a marble or pebble in the shoe.

Deformity is sometimes the main complaint; the patient may abhor a ‘crooked toe’ or a ‘twisted foot’, even if it is not painful, and parents often worry about their children who are ‘flat-footed’ or ‘pigeon-toed’. Elderly patients may complain chiefly of having difficulty fitting shoes.

Swelling is common, even in normal people, but it gains more significance if it is unilateral or strictly localized.

Instability of the ankle or subtalar joint produces repeated episodes of the joint ‘giving way’. Ask about any previous injury (a ‘twisted ankle’).

Numbness and *paraesthesia* may be felt in all the toes or in a circumscribed field served by a single nerve or one of the nerve roots from the spine.

General questions that help in reaching a diagnosis, assessing the impact of the condition on function and deciding on treatment in foot and ankle problems are:

Have you any pain or stiffness in your muscles, joints or back? Can you dress yourself completely without any difficulty? Can you walk up and down stairs without any difficulty?

SIGNS WITH PATIENT UPRIGHT

It is important to see the patient stand, as deformities will often be much better shown once the patient is weightbearing. The patient, whose lower limbs should be exposed from the knees down, stands first facing the surgeon, then with his or her back to the surgeon. Ask the patient to rise up on tiptoes and then settle back on the heels. Note the posture of the feet throughout this movement. Normally the heels are in slight valgus while standing and inverted on tiptoes; the degree of inversion should be equal on the two sides, showing that the subtalar joint is mobile and the tibialis posterior functioning. Viewed from behind, if there is excessive eversion of one foot, the lateral toes are more easily visible on that side (the ‘*too-many-toes*’ sign).

Gait Observing the gait also helps to identify dynamic problems and pathology on other lower limb joints. The patient is asked to walk normally. Note whether the gait is smooth or halting and whether the feet are well balanced. Gait is easier to analyze if concentrating on the sequence of movements that make up the walking cycle. It begins with heel-strike, then moves into stance, then push-off and finally swing-through before making the next heel-strike. The stance phase itself can be further divided into three intervals: (1) from heel-strike to flat foot; (2) progressive ankle dorsiflexion as the body passes over the foot; (3) ankle plantarflexion leading to toe-off. Gait may be disturbed by pain, muscle weakness, deformity or stiffness. The position and mobility of each ankle is of prime importance. A fixed equinus deformity results in the heel failing to strike the ground at the beginning of the walking cycle; sometimes the patient forces heel contact by hyperextending the knee. If the ankle dorsiflexors are weak, the forefoot may hit the ground prematurely, causing a ‘slap’; this is

referred to as foot-drop (or drop-foot). During swingthrough the leg is lifted higher than usual so that the foot can clear the ground (a high-stepping gait). Hindfoot and midfoot deformities may interfere with level ground-contact in the second interval of stance; the patient walks on the inner or outer border of the foot. Toe contact, especially of the great toe, is also important; pain or stiffness in the first MTP joint may prevent normal push-off.

SIGNS WITH PATIENT SITTING OR LYING

A systematic approach to examination, following the 'look, feel, move' steps, will lead to a diagnosis in the majority of cases. Next the patient is examined lying on a couch, or it may be more convenient if he or she sits opposite the examiner and places each foot in turn on the examiner's lap.

Look

The heel is held square so that any foot deformity can be assessed. The toes and sole should be inspected for *skin changes*. The foot shows areas of overload by producing callosities, and there are often corresponding areas of wear and signs of overload on the footwear. Thickening and keratosis may be seen over the proximal toe joints or on the soles. Atrophic changes in theskin and toe-nails are suggestive of a neurological or vascular disorder.

Deformity may be in the ankle, the foot or toes. A foot that is set flat on the ground at a right angle to the tibia is described as *plantigrade*; if it is set in fixed plantarflexion (pointing downwards) it is said to be in *equinus*; a dorsiflexed position is called *calcaneus*. Common defects are a 'flat-footed' stance (*pes valgus*); an abnormally high instep (*pes cavus*); a downward- arched forefoot (*pes plantaris*); lateral deviation of the great toe (*hallux valgus*); fixed flexion of a single interphalangeal (IP) joint (*hammer toe*) or of all the toes (*claw toes*).

Swelling may be diffuse and bilateral, or localized; unilateral swelling nearly always has a surgical cause and bilateral swelling is more often 'medical' in origin. Swelling over the medial side of the first metatarsal head (a *bunion*) is common in older women.

Corns are usually obvious; *callosities* must be looked for on the soles of the feet.

Feel

Pain and tenderness in the foot and ankle localize very well to the affected structures - the patient really does show us where the problem is. The skin temperature is assessed and the pulses are felt. Remember that one in every six normal people does not have a dorsalis pedis artery. If all the foot pulses are absent, feel for the popliteal and femoral pulses; the patient may need further evaluation by Doppler ultrasound. If there is tenderness in the foot it must be precisely localized, for its site is often diagnostic. Any swelling, oedema or lumps must be examined. Sensation may be abnormal; the precise distribution of any change is important. If a neuropathy is suspected (e.g. in a diabetic patient) test also for vibration sense, protective sensation and sense of position in the toes.

Move

The foot comprises a series of joints that should be examined methodically:

· *Ankle joint* – With the heel grasped in the left hand and the midfoot in the right, the ranges of plantarflexion (flexion) and dorsiflexion (extension) are estimated. Beware not to let the foot go into valgus during passive dorsiflexion as this will give an erroneous idea of the range of movement.

· *Subtalar joint* – It is important to 'lock' the ankle joint when assessing subtalar inversion and eversion. This is done simply by ensuring that the ankle is plantigrade when the heel is moved. It is often easier to record the amount of subtalar movement if the patient is examined prone. Inversion is normally greater than eversion.

· *Midtarsal joint* – One hand grips the heel firmly to stabilize the hindfoot while the other hand movesthe forefoot up and down and from side to side.

· *Toes* – The MTP and IP joints are tested separately. Extension (dorsiflexion) of the great toe at the MTP joint should normally exceed 70 degrees and flexion 10 degrees.

Stability

Stability is assessed by moving the joints across the normal physiological planes and noting any abnormal 'clunks'. Ankle stability should be tested in both coronal and sagittal planes, always comparing the two joints. Patients with recent ligament injury may have to be examined under anaesthesia. *Medial* and *lateral stability* are checked by stressing the ankle first in valgus and then in varus.

Anteroposterior stability is assessed by performing an anterior 'drawer test': the patient lies on the examination couch with hips and knees flexed and the feet resting on the couch surface; the examiner grasps the distal tibia with both hands and pushes firmly backwards, feeling for abnormal translation of the tibia upon the talus. Another way of doing this is to stabilize the distal tibia with one hand while the other grasps the heel and tries to shift the hindfoot forwards and backwards. The same tests can be performed under x-ray and the positions of the two ankles measured and compared.

Muscle power

Power is tested by resisting active movement in each direction. The patient will be more cooperative if the movement required is demonstrated precisely. While the movement is held, feel the muscle belly and tendon to establish whether they are intact and functioning.

Shoes

Footwear often adds additional clues when examining the foot and ankle, providing valuable information about faulty stance or gait.

General examination

If there are any symptoms or signs of vascular or neurological impairment, or if multiple joints are affected, a more general examination is essential.

IMAGING

There are practical problems with imaging in children, and babies in particular because: (1) babies tend not to keep still during examination; (2) their bones are not completely ossified and their shape and position may be hard to define.

X-rays In the adult, the standard views of the ankle are anteroposterior (AP), mortise (an AP view with the ankle internally rotated 15‑20 degrees) and lateral. Although the subtalar joint can be seen in a lateral view of the foot, medial and lateral oblique projections allow better assessment of the joint. These views are often used to check articular congruity after treatment of calcaneal fractures. The calcaneum itself is usually x-rayed in axial and lateral views, but a weightbearing view is helpful in defining its relationship to the talus and tibia. The foot, toes and intertarsal joints are well displayed in standing anteroposterior and medial oblique views, but occasionally a true lateral view is needed.

Stress x-rays These complement the clinical tests for ankle stability. The patient should be completely relaxed; if the ankle is too painful, stress x-rays can be performed under regional or general anaesthesia. Both ankles should be examined, for comparison.

CT scan CT provides excellent coronal views and is important in assessing fractures and congenital bony coalitions.

Radioscintigraphy Radioisotope scanning, though nonspecific, is excellent for localizing areas of abnormal blood flow or bone remodelling activity; it is useful in the diagnosis of covert infection.

MRI and ultrasound These methods are used to demonstrate soft tissue problems, such as tendon and ligament injuries.

PEDOBAROGRAPHY

A record of pressures beneath the foot can be obtained by having the patient stand or walk over a force plate; sensors in the plate produce a dynamic map of the peak pressures and the time over which these are recorded can be obtained. Although this is sometimes helpful in clinical decision making, or for comparing pre- and postoperative function, the investigation is used mainly as a research tool.

WHERE DOES IT HURT; WHERE IS IT TENDER?

Anterior ankle joint line – impingement from osteophytes in OA

Anterolateral angle of ankle joint – lateral gutter impingement in post-traumatic ankle with soft tissue

problems

Bony tip/lateral malleolus – ankle fracture (Ottawa guidelines)

Posterior/inferior to lateral malleolus – peroneal tenosynovitis or tear

Posterior to medial malleolus/line of tibialis posterior – tibialis posterior tendinitis or tear, and in plano-valgus

collapse of hindfoot

Base of fifth metatarsal – fracture/insertional problem with peroneus brevis

Achilles tendon – Achilles tendinitis/paratendinitis

Achilles insertion – insertional tendinitis

Retrocalcaneal bursa – bursitis

Plantar fascia – plantar fasciitis

Medial to first MTP joint – bunion

Dorsal to first MTP joint – OA, hallux limitus/rigidus

Beneath first MTP joint – sesamoiditis

Beneath metatarsal heads – 'metatarsalgia'

In third interspace – Morton's neuroma

CONGENITAL DEFORMITIES

Congenital deformities of the foot are common. Many appear as part of a more widespread genetic disorder; only those in which the foot is the main (or only) problem are considered in this section. Isolated abnormalities of the toes are also dealt with elsewhere.

TALIPES EQUINOVARUS (IDIOPATHIC CLUB-FOOT)

The term '*talipes*' is derived from *talus* (Latin = ankle bone) and *pes* (Latin = foot). Equinovarus is one of several different talipes deformities; others are talipes calcaneus and talipes valgus. In the full-blown equinovarus deformity the heel is in equinus, the entire hindfoot in varus and the midand forefoot adducted and supinated. The abnormality is relatively common, the incidence ranging from 1‑2 per thousand births; boys are affected twice as often as girls and the condition is bilateral in one-third of cases. The exact cause is not known, although the resemblance to other disorders suggests several possible mechanisms. It could be a germ defect, or a form ofarrested development. Its occurrence in neurological disorders and neural tube defects (e.g. myelo -meningocele and spinal dysraphism) points to a neuromuscular disorder. Severe examples of club-foot are seen in

association with arthrogryposis, tibial deficiency and constriction rings. In some cases it is no more than a postural deformity caused by tight packing in an overcrowded uterus.

Clinical features

The deformity is usually obvious at birth; the foot is both turned and twisted inwards so that the sole faces posteromedially. More precisely, the ankle is in equinus, the heel is inverted and the forefoot is adducted and supinated; sometimes the foot also has a high medial arch (cavus), and the talus may protrude on the dorsolateral surface of the foot. The heel is usually small and high, and deep creases appear posteriorly and medially; some of these creases are incomplete constriction bands. In some cases the calf is abnormally thin. In a normal baby the foot can be dorsiflexed and everted until the toes touch the front of the leg. In club-foot this manoeuvre meets with varying degrees of resistance and in severe cases the deformity is fixed. The infant must always be examined for associated disorders such as congenital hip dislocation and spina bifida. The absence of creases suggests arthrogryposis; look to see if other joints are affected.

Treatment

The aim of treatment is to produce and maintain a plantigrade, supple foot that will function well. There are several methods of treatment but relapse is common, especially in babies with associated neuromuscular disorders.

PES PLANUS AND PES VALGUS(‘FLAT-FOOT’)

“Our feet are no more alike than our faces.” This truism from a *British Medical Journal* editorial sums up the problem of ‘normally abnormal’ feet. The medial arch may be normally high or normally low. The term ‘flatfoot’ applies when the apex of the arch has collapsed and the medial border of the foot is in contact (or nearly in contact) with the ground; the heel becomes valgus and the foot pronates at the subtalar-midtarsal complex. The problems associated with flat-foot differ in babies, children and adults and these three categories will therefore be considered separately.

FLAT-FOOT IN CHILDREN AND ADOLESCENTS

Flat-foot is a common complaint among children. Or rather their parents, grandparents, and assistants in the shoe-shop‐the children themselves usually dont seem to notice it!

Flexible flat-foot Flexible pes valgus appears in toddlers as a normal stage in development, and it usually disappears after a few years, when medial arch development is complete; occasionally, though, it persists into adult life. The arch can often be restored by simply dorsiflexing the great toe (*Jack’s test*), and during this manoeuvre the tibia rotates externally (Rose et al., 1985). Many of these children have ligamentous laxity and there may be a family history of both flat feet and joint hypermobility.

Stiff (or ‘rigid’) flat-foot A deformity that cannot be corrected passively should alert the examiner to an underlying abnormality. *Congenital vertical talus* is dealt with earlier. In older

children, conditions to be considered are: (1) *tarsal coalition*; (2) *an inflammatory joint disorder*; (3) a *neurological disorder*.

. *Compensatory flat-foot* This is a spurious deformity that occurs in order to accommodate some other postural defect. For example, a tight tendo Achillis (or a mild fixed equinus) may be accommodated by everting the foot; or if the lower limbs are externally rotated the body weight falls anteromedial to the ankle and the feet go into valgus – the Charlie Chaplin look.

Treatment

Physiological flat-foot Young children with flexible flat feet require no treatment. Parents need to be reassured and told that the 'deformity' will probably correct itself in time; even if it does not fully correct, function is unlikely to be impaired. Some parents will cite examples of other children who were helped by insoles or moulded heel-cups. These appliances serve mainly to alter the pattern of weightbearing and hence that of shoe wear; simply put, they are more effective in treating the shoes than the feet.

Tight tendo Achillis Flat-foot associated with a tight tendo Achillis and restricted dorsiflexion at the ankle may benefit from tendon-stretching exercises.

Accessory navicular Sometimes the main complaint (with a flexible flat-foot) is tenderness over an unusually prominent navicular on the medial border of the midfoot. X-rays may show an extra ossicle at this site – the accessory navicular. Symptoms are due to pressure (and possibly a 'bursitis') over the bony prominence, or repetitive strain at the synchondrosis between the accessory ossicle and the navicular proper. If symptoms warrant it, the accessory bone can be shelled out from within the tibialis posterior tendon. If the medial arch has 'dropped' significantly, the tibialis posterior tendon can be used as a 'hitch' by reinserting it through a hole drilled in the navicular and suturing the loop with the foot held in maximum inversion (Kidner's operation).

Rigid flat-foot (tarsal coalition) One of the problems with treatment of this condition is that the presence of a tarsal coalition is not necessarily the cause of the patient's symptoms; the anomaly is sometimes discovered as an incidental finding in asymptomatic feet. For this reason the initial treatment should always be conservative. A walking plaster is applied with the foot plantigrade and is retained for 6 weeks; splintage with an outside iron and inside T-strap may have to be continued for another 3–6 months. Obviously if an inflammatory joint disorder is discovered, this will have to be treated. If symptoms do not settle, operative treatment is needed. A calcaneonavicular bar can be resected without much difficulty through a lateral approach, and the operation may be performed before puberty; a portion of the bar is removed and the gap filled with fat or a piece of muscle (e.g. extensor digitorum brevis) to prevent recurrence. Talocalcaneal coalitions are more difficult to deal with and it may be wiser to wait until after the patient reaches puberty and then perform a triple arthrodesis.

FLAT-FOOT IN ADULTS

As in children, the usual picture is of a flexible flatfoot with no obvious cause. However, underlying disorders are common enough to always warrant a careful search for abnormal ligamentous laxity, tarsal coalitions, disorders of the tibialis posterior tendon, post-traumatic deformity, degenerative arthritis, neuropathy and conditions resulting in muscular imbalance. Painful acquired flat-foot often results from tibialis posterior dysfunction. Tibialis posterior tendon dysfunction affects predominantly women in later midlife. It is usually of insidious onset, affecting one foot much more than the other, and with identifiable systemic factors such as obesity, diabetes, steroids or surgery. There may be recollection of a minor episode of trauma, such as a twisting injury to the foot. The patient experiences aching discomfort in the line of the tibialis posterior tendon, often radiating up the inner aspect of the lower leg. The foot often feels ‘tired’. As the tendon stretches out the foot drifts into plano-valgus, producing the typical acquired flat-foot deformity. As the tendon ruptures the ache or pain will often improve, temporarily, but as the foot deformity then worsens the plantar fascia becomes painful and there may be lateral hindfoot pain as the fibula starts to impinge against the calcaneum Examination There is usually swelling and tenderness in the line of tibialis posterior, at and distal to the medial malleolus. The hindfoot collapse is best appreciated by viewing the patient from behind, when the valgus deformity of the heel is appreciated, and the forefoot abduction leads to ‘too many toes’ being seen from this position, compared to the contralateral foot. It is difficult for the patient to do a single leg heel raise, as the tibialis posterior cannot stabilize and invert the heel, impairing the heel-raise action of the Achilles tendon.

Imaging

Weightbearing x-rays show the altered foot axes. The tendon can be assessed with ultrasound or magnetic resonance imaging (MRI) scan.

Treatment

The key point is to recognize the condition. If it is in the early stages then relative rest (sticks or crutches), support with a temporary insole, elasticated foot/ ankle support and oral non-steroidal anti-inflammatory drugs (NSAIDs) may be effective. Whether or not to inject the tendon *sheath* with corticosteroid is contentious; but to inject the tendon itself is just plain wrong! These temporary measures may offer the opportunity to institute more permanent solutions, such as modification of weight and activity, and assessment for definitive orthotics.

ORTHOSES

Functional foot orthoses (FFOs) have a role to play in the adult flexible but symptomatic flat-foot. These appliances (usually called *orthotics*) are used to correct abnormal foot function or biomechanics and, in so doing, they also correct for abnormal lower extremity function; they are very much more than an ‘arch support’. Orthotics are useful in the treatment of a range of painful conditions of the foot and lower extremities, in particular first MTP joint arthritis, metatarsalgia, arch and instep pain, ankle pain and heel pain. Since abnormal foot function may cause abnormal leg, knee and hip function, orthotics can be used to treat painful tendinitis and bursitis conditions in the ankle, knee and hip, as well as exercise-induced leg

pain (‘shin splints’). Some types of FFOs are also designed to accommodate painful areas on the soles of the feet (like accommodative foot orthoses). Orthoses may be made of flexible, semi-rigid or rigid plastic or graphite materials. They are relatively thin and fit easily into several types of shoe. They are fabricated from a three-dimensional model of the foot or scanning the foot with a mechanical or optical scanner. Assessment for orthotics can be performed by a podiatrist, who can also advise on whether the usual/intended footwear will accommodate such a device and offer the support needed for it to be effective. ‘Off-the-shelf’ insoles are cheaper, but there are several advantages to prescription foot orthoses. They are custom-made to precisely fit each foot, and are made in relatively rigid, durable materials with a minimal chance of discomfort or irritation to the foot and a greater potential to relieve pain.

PHYSIOTHERAPY

Local treatment of the associated inflammation with physiotherapy might be of benefit. Assessment of the hindfoot biomechanics by a podiatrist might help to prevent progression, and could offer protection to the contralateral side, which is often much less severely

affected.

SURGERY

If the condition does not improve with a few weeks of conservative treatment, or the patient presents several months after onset of the symptoms, then surgical intervention should be considered. Options include surgical decompression and tenosynovestomy, or reconstruction of the tendon. The latter is often combined with a calcaneal osteotomy to help to protect the tendon and improve the axis. If there is already degeneration in the hindfoot joints then triple arthrodesis might be indicated (fusing the subtalar, calcaneo-cuboid and talonavicular joints - the ankle joint is not arthrodesed in this procedure, so foot plantarflexion and dorsiflexion are maintained).

PES CAVUS (HIGH-ARCHED FEET)

In pes cavus the arch is higher than normal, and often there is also clawing of the toes. The close resemblance to deformities seen in neurological disorders where the intrinsic muscles are weak or paralyzed suggests that all forms of pes cavus are due to some type of muscle imbalance. There are rare congenital causes, such as arthrogryposis, but in the majority of cases pes cavus results from an acquired neuromuscular disorder see Box opposite. A specific abnormality can often be identified; hereditary motor and sensory neuropathies and spinal cord abnormalities (tethered cord syndrome, diastematomyelia) are the commonest in Western countries but poliomyelitis is the most common cause worldwide. Occasionally the deformity follows trauma - burns or a compartment syndrome resulting in Volkmann’s contracture of the sole.

NEUROMUSCULAR CAUSES OF PES CAVUS

Muscular dystrophies Duchenne, Becker

Neuropathies HMSN I and II

Cord lesions Poliomyelitis, syringomyelia, diastomatomyelia, tethered cord

Cerebral disorders Cerebral palsy, Friedreich's ataxia

HALLUX VALGUS

Hallux valgus is the commonest of the foot deformities (and probably of all musculoskeletal deformities). In people who have never worn shoes the big toe is in line with the first metatarsal, retaining the slightly fanshaped appearance of the forefoot. In people who habitually wear shoes the hallux assumes a valgus position; but only if the angulation is excessive is it referred to as 'hallux valgus'. Splaying of the forefoot, with varus angulation of the first metatarsal, predisposes to lateral angulation of the big toe in people wearing shoes - and most of all in those who wear high-heeled shoes. *Metatarsus primus varus* may be congenital, or it may result from loss of muscle tone in the forefoot in elderly people. Hallux valgus is also common in rheumatoid arthritis, probably due to weakness of the joint capsule and ligaments. Heredity plays an important part; a positive family history is obtained in over 60 per cent of cases.

Clinical features

The commonest complaints are pain over the bunion, worries about cosmesis and difficulty fitting shoes. Often there is also deformity of the lesser toes and pain in the forefoot. With the patient standing, planovalgus hindfoot collapse may become apparent. The great toe is in valgus and the bunion varies in appearance from a slight prominence over the medial side of the first metatarsal head to a red and angrylooking bulge that is tender. The MTP joint often retains a good range of movement, but in longstanding cases it may be osteoarthritic. Always check the circulation and sensation.

X-rays

Standing views will show the degree of metatarsal and hallux angulation. Lines are drawn along the middle of the first and second metatarsals and the proximal phalanx of the great toe; normally the intermetatarsal angle is less than 9 degrees and the valgus angle at the MTP joint less than 15 degrees. Any greater degree of angulation should be regarded as 'hallux valgus Not all types of valgus deformity are equally progressive and troublesome. Based on the x-ray appearances, patients with hallux valgus can be divided into three types (Piggott, 1960):

(1) those in whom the MTP joint is normally centred but the articular surfaces, though congruent, are tilted towards valgus;

(2) those in whom the articular surfaces are not congruent, the phalangeal surface being tilted towards valgus;

(3) those in whom the joint is both incongruent and slightly subluxated (Fig. 21.23).

Type 1 is a stable joint and any deformity is likely to progress very slowly or not at all. Type 2 is somewhat unstable and likely to progress. Type 3 is even more unstable and almost certain to progress.

Treatment

ADOLESCENTS

Many young patients are asymptomatic, but worry over the shape of the toe and an anxious mother keen not to let the condition become as severe as her own will bring the patient to the clinic. It is wise to try conservative measures first, mainly because surgical correction in this age group carries a 20–40 per cent recurrence rate. This consists essentially of encouraging the patient to wear shoes with wide and deep toeboxes, soft uppers and low heels – 'trainers' are a good choice. If x-rays show a type 1 (congruous) deformity, the patient can be reassured that it will progress very slowly, if at all. If there is an incongruous deformity, surgical correction will sooner or later be required.

There are a number of non-operative strategies that may be adopted to deal with the deformity and the resulting limitations, but none that will get rid of the bunion itself. Accommodating, comfortable shoes can help, but are not acceptable for some patients (or professions). Lace-up or Velcro-fastening shoes are better than slip-ons, and flat shoes are probably better than those with a raised heel. Bunion pads (like a Polo/doughnut shape) can help to offload the tender bunion, but strapping and overnight splints are probably a waste of money with no quality research to support their use. Chiropody can help by taking care of the callosities and skin compromise. Podiatrists may help to correct the foot biomechanics, but there is no good evidence that antipronatory orthoses are effective in the longer term management of the bunion. Diabetic services often provide specialized foot-care.

Operative treatment In the adolescent with mild deformities, where the hallux valgus angle is less than 25 degrees, correction can be obtained by either a softtissue rebalancing operation (see later) or by a metatarsal osteotomy. If the x-ray shows a congruent articulation, the deformity is largely bony and therefore amenable to correction by a distal osteotomy.

ADULTS-In the adult, when self-care is insufficient and the bunion is causing pain and difficulty with footwear, surgical options are appropriate. Recurrent infection or ulceration are also indications for operative treatment. The type of surgery proposed will depend on the level and extent of the deformity. This will usually comprise: (1) an osteotomy to re-align the first metatarsal; (2) soft tissue procedures to rebalance the joint.

HALLUX RIGIDUS

'Rigidity' (or stiffness) of the first MTP joint occurs at almost any age from adolescence onwards. In young people it may be due to local trauma or osteochrondritis dissecans of the

first metatarsal head. In older people it is usually caused by longstanding joint disorders such as gout, pseudogout or osteoarthritis (OA), and is very often bilateral. In contrast to hallux valgus, men and women are affected with equal frequency. A family history is common.

DISORDERS OF THE TENDO ACHILLIS

ACHILLES TENDINITIS

Athletes, joggers and hikers often develop pain and swelling around the tendo Achillis, due to local irritation of the tendon sheath or the paratenon.

Pathology

The condition usually affects the 'watershed' area about 4 cm above the insertion of the tendon, an area where the blood supply to the tendon is poorer than elsewhere. The tendon sheath or the flimsy tissue around it may become inflamed. In a minority of cases the changes appear at the tendon insertion, or there may be inflammation of the retrocalcaneal bursa just above the calcaneum and deep to the tendon; anatomical deformity of the posterior part of the calcaneum may contribute to the pathogenesis.

Clinical features

The condition may come on gradually, or rapidly following a change in sporting activity (or a change of sports footwear). Less commonly there is a history of direct trauma to the Achilles tendon. The area above the heel may look inflamed and function is inhibited because of pain in the heel-cord, especially at pushoff. The tendon feels thickened in the watershed area about 4 cm above its insertion. In chronic cases an ultrasound scan may be helpful in confirming the diagnosis. If the onset is very sudden, suspect tendon rupture (see later).

Treatment

If the condition starts acutely, it will often settle within about 6 weeks if treated appropriately. Referral for early physiotherapy is important. In the interim, advice on rest, ice, compression and elevation (RICE) and the use of an NSAID (oral or topical) are helpful. When the symptoms improve, stretching exercises, followed by a muscle strengthening programme, should be advised. The use of a removeable in-shoe heel-raise might be helpful. If there is a plano-valgus hindfoot, correction with orthotics will often bring about improvement and reduce the risk of recurrence. When the onset is insidious and treatment is started late, symptoms will be prolonged and may last for 9 months or longer.

Operative treatment is seldom necessary but if symptoms fail to settle with physiotherapy then surgery may be appropriate – even more so if there is suspicion of an acute (or missed) tendon rupture. This will involve some type of 'decompressive' operation. Treatments such as radiofrequency coblation or extracorporeal shockwave lithotripsy are now showing some promise.

Potential pitfalls

Injection with corticosteroids should be avoided. Tendon rupture is a real risk and could well give rise to litigation. Do not diagnose ‘partial rupture’ of the Achilles tendon; this should only be entertained if there is clearly some discontinuity of the tendon on ultrasound scan.

ACHILLES TENDON RUPTURE

A ripping or popping sensation is felt, and often heard, at the back of the heel. This most commonly occurs in sports requiring an explosive push-off: squash, badminton, football, tennis, netball. The patient will often report having looked round to see who had hit them over the back of the heel, the pain and collapse are so sudden. The typical site for rupture is at the vascular watershed about 4 cm above the tendon insertion onto the calcaneum. The condition is often associated with poor muscle strength and flexibility, failure to warm up and stretch before sport, previous injury or tendinitis and corticosteroid injection.

Examination

Plantarflexion of the foot is usually inhibited and weak (although it may be possible, as the long flexors of the toes are also ankle flexors). There is often a palpable gap at the site of rupture; bruising comes out a day or two later. The calf squeeze test (Thompson’s or Simmond’s test) is diagnostic of Achilles tendon rupture: normally, with the patient prone, if the calf is squeezed the foot will plantarflex involuntarily; if the tendon is ruptured the foot remains still. Clinical assessment is often sufficient. *Ultrasound scans* must be used to confirm or refute the diagnosis.

Differential diagnosis

Incomplete tear A complete rupture is often mistaken for a partial tear (which is rare). The mistake arises because, if a complete rupture is not seen within 24 hours, the gap is difficult to feel; moreover, the patient may by then be able to stand on tiptoe (just), by using his or her long toe flexors.

Tear of soleus muscle A tear at the musculotendinous junction causes pain and tenderness halfway up the calf. This recovers with the aid of physiotherapy and raising the heel of the shoe.

Treatment

If the patient is seen early, the ends of the tendon may approximate when the foot is passively plantarflexed. If so, a plaster cast or special boot is applied with the foot in equinus; rehabilitation and physiotherapy regimes vary, but it is probably safe, and may be better for eventual tendon strength, to commence physiotherapy within 4–6 weeks. A shoe with a raised heel should be worn for a further 6–8 weeks. The ‘re-rupture rate’ is about 10 per cent. Operative repair is associated with an earlier return to function, better tendon and calf muscle

strength and a lower re-rupture rate. Supported rehabilitation and physiotherapy are commenced early (within a week or two of repair) There are, however, risks associated with operative tendon repair, including wound healing problems and sural nerve neuroma. For ruptures that present late, reconstruction using local tendon substitutes (e.g. flexor hallucis longus tendon) or strips of fascia lata is still possible.

PAINFUL FEET

"My feet are killing me!" This complaint is common but the cause is often elusive. Pain may be due to: (1) mechanical pressure (which is more likely if the foot is deformed or the patient obese); (2) joint inflammation or stiffness; (3) a localized bone lesion; (4) peripheral ischaemia; (5) muscular strain – usually secondary to some other abnormality. Remember, too, that local disorders may be part of a generalized disease (e.g. diabetes or rheumatoid arthritis), so examination of the entire patient may be indicated.

POSTERIOR HEEL PAIN

Two common causes of heel pain are traction 'apophysitis' and calcaneal bursitis:

Traction 'apophysitis' (Sever's disease) This condition usually occurs in boys aged about 10 years. It is not a 'disease' but a mild traction injury. Pain and tenderness are localized to the tendo Achillis insertion. The x-ray report usually refers to increased density and fragmentation of the apophysis, but often the painless heel looks similar. The heel of the shoe should be raised a little and strenuous activities restricted for a few weeks.

Calcaneal bursitis Older girls and young women often complain of painful bumps on the backs of their heels. The posterolateral portion of the calcaneum is prominent and shoe friction causes retrocalcaneal bursitis. Symptoms are worse in cold weather and when wearing high-heeled shoes (hence the use of colloquial labels such as 'winter heels' and 'pumpbumps'). Treatment should be conservative – attention to footwear (open-back shoes are best) and padding of the heel. Operative treatment – removal of the bump or dorsal wedge osteotomy of the calcaneum – is feasible but the results are unpredictable; despite the reduction in the size of the bumps, patients often continue to experience discomfort, potentially added to by an operation scar.

INFERIOR HEEL PAIN

Calcaneal bone lesions Any bone disorder in the calcaneum can present as heel pain: a stress fracture, osteomyelitis, osteoid osteoma, cyst-like lesions and Paget's disease are the most likely. X-rays usually provide the diagnosis.

PLANTAR FASCIITIS

This is an annoying and painful condition that limits function. There is pain and tenderness in the sole of the foot, mostly under the heel, with standing or walking. The condition usually comes on gradually, without any clear incident or injury but sometimes there is a history of sudden increase in sporting activity, or a change of footwear, sports shoes or running surface. There may be an associated tightness of the Achilles tendon. The pain is often worse when first getting up in the morning, with typical hobbling downstairs, or when first getting up from a period of sitting - the typical start-up pain and stiffness. The pain can at times be very sharp, or it may change to a persistent background ache as the patient walks about. The condition can take 18-36 months or longer to resolve, but is generally self-limiting, given time.

PAIN OVER THE MIDFOOT

In children, pain in the midtarsal region is rare: one cause is *Kohler's disease* (osteochondritis of the navicular). The bony nucleus of the navicular becomes dense and fragmented. The child, under the age of 5, has a painful limp and a tender warm thickening over the navicular. Usually no treatment is needed as the condition resolves spontaneously. If symptoms are severe, a short period in a below-knee plaster helps.

A comparable condition occasionally affects middle- aged women (*Brailsford's disease*); the navicular becomes dense, then altered in shape, and later the midtarsal joint may degenerate.

In adults, especially if the arch is high, a ridge of bone sometimes develops on the adjacent dorsal surfaces of the medial cuneiform and the first metatarsal (the '*overbone*'). A lump can be seen, which feels bony and may become bigger and tender if the shoe presses on it. If shoe adjustment fails to provide relief the lump may be bevelled off.

GENERALIZED PAIN IN THE FOREFOOT

Metatarsalgia Generalized ache in the forefoot is a common expression of foot strain, which may be due to a variety of conditions that give rise to faulty weight distribution (e.g. flattening of the metatarsal arch, or undue shortening of the first metatarsal), or merely the result of prolonged or unaccustomed walking, marching, climbing or standing. These conditions have this in common: they give rise to a mismatch between the loads applied to the foot, the structure on which those loads are acting, and the muscular effort required loads. Aching is felt across the forefoot and the anterior metatarsal arch may have flattened out. There may even be callosities under the metatarsal heads.

Treatment involves: (1) dealing with the mechanical disorder (correcting a deformity if it is correctable, supplying an orthosis that will redistribute the load, fitting a shoe that will accommodate the foot); and (2) performing regular muscle strengthening exercises, especially for the intrinsic muscles that maintain the anterior (metatarsal) arch of the foot. A good 'do-ityourself' exercise is for the patient to stand barefoot on the floor, feet together, and then drag their body forwards by repeatedly crimping the toes to produce traction upon the floor. Ten minutes a day should suffice.

Pain in metatarsophalangeal joints Inflammatory arth - ritis (e.g. rheumatoid disease) may start in the foot with synovitis of the MTP joints. Pain in these cases is associated with swelling and tenderness of the forefoot joints and the features are almost always bilateral and symmetrical.

LOCALIZED PAIN IN THE FOREFOOT

Whereas metatarsalgia involves the entire forefoot, localized pain and tenderness is related to a specific anatomical site in the forefoot and could be due to a variety of bone or soft tissue disorders: ‘sesamoiditis’, osteochondritis of a metatarsal head (Freiberg’s disease), a metatarsal stress fracture or digital nerve entrapment (Morton’s disease).

Sesamoiditis

Pain and tenderness directly under the first metatarsal head, typically aggravated by walking or passive dorsiflexion of the great toe, may be due to sesamoiditis. This term is a misnomer: symptoms usually arise from irritation or inflammation of the peritendinous tissues around the sesamoids‑more often the medial (tibial) sesamoid, which is subjected to most stress during weightbearing on the ball of the foot.

Acute sesamoiditis may be initiated by direct trauma (e.g. jumping from a height) or unaccustomed stress (e.g. in new athletes and dancers).

Chronic sesamoid pain and *tenderness* should signal the possibility of sesamoid displacement, local infection (particularly in a diabetic patient) or avascular necrosis.

Sesamoid chondromalacia is a term coined by Apley (1966) to explain changes such as fragmentation and cartilage fibrillation of the medial sesamoid. X-rays in these cases may show a bipartite or multipartite medial sesamoid, which is often mistaken for a fracture.

Treatment, in the usual case, consists of reduced weightbearing and a pressure pad in the shoe. In resistant cases, a local injection of methylprednisolone and local anaesthetic often helps; otherwise the sesamoid should be shaved down or removed, taking great care not to completely interrupt the flexor hallucis brevis tendon.

Freiberg’s disease (osteochondritis; osteochondrosis)

Osteochondritis (or osteochondrosis) of a metatarsal head is probably a type of traumatic osteonecrosis of the subarticular bone in a bulbous epiphysis (akin to osteochondritis dissecans of the knee). It usually affects the second metatarsal head (rarely the third) in young adults, mostly women. The patient complains of pain at the MTP joint. A bony lump (the enlarged head) is palpable and tender and the MTP joint is irritable. X-rays show the head to be flattened and wide, the neck thick and the joint space apparently increased. If discomfort is marked, a walking plaster or moulded sandal will help to reduce pressure on the metatarsal head. If pain and stiffness persist, operative synovectomy, debridement and trimming of the metatarsal head should be considered. Pain relief is usually good and the range of dorsiflexion is improved.

Stress fracture

Stress fracture, usually of the second or third metatarsal, occurs in young adults after unaccustomed activity or in women with postmenopausal osteoporosis. The dorsum of the foot may be slightly oedematous and the affected shaft feels thick and tender. The x-ray appearance is at first normal, but later shows fusiform callus around a fine transverse fracture. Long before x-ray signs appear, a radioisotope scan will show increased activity. Treatment is either unnecessary or consists simply of rest and reassurance.

Interdigital nerve compression (Morton's metatarsalgia)

Morton's metatarsalgia is a common problem, with neuralgia affecting a single distal metatarsal interspace, usually the third (affecting the third and fourth toes), sometimes the second (affecting the second and third toes), rarely others. The patient typically complains of pain on walking, with the sensation of walking on a pebble in the shoe, or of the sock being rucked-up under the ball of the foot. The pain is worse in tight footwear and often has to be relieved by removing the footwear and massaging the foot. Activities that load the forefoot (running, jumping, dancing) exacerbate the condition, which often consists of severe forefoot pain and then a reluctance to weightbear. In Morton's metatarsalgia the pain is typically reproduced by laterally compressing the forefoot whilst also compressing the affected interspace – this produces the pathognomic Mulder's click as the 'neuroma' displaces between the metatarsal heads. This is essentially an entrapment or compression syndrome affecting one of the digital nerves, but secondary thickening of the nerve creates the impression of a 'neuroma'. The lesion, and an associated bursa, occupy a restricted space between the distal metatarsals, and are pinched, especially if footwear also laterally compresses the available space.

Treatment A step-wise treatment programme is advisable. Simple offloading of the metatarsal heads by using a metatarsal dome insole and wider fitting shoes may help. If symptoms do not improve with these measures then a steroid injection into the interspace will bring about lasting relief in about 50 per cent of cases. Surgical intervention is often successful; the nerve should be released by dividing the tight transverse intermetatarsal ligament; this can be done through either a dorsal longitudinal or a plantar incision; most surgeons will also excise the thickened portion of the nerve. This is successful in about 90 per cent of patients; the remaining 10 per cent will continue to experience varying degrees of discomfort.

TARSAL TUNNEL SYNDROME

Pain and sensory disturbance in the medial part of the forefoot, unrelated to weightbearing, may be due to compression of the posterior tibial nerve behind and below the medial malleolus. Sometimes this is due to a space-occupying lesion, e.g. a ganglion, haemangioma or varicosity. The pain is often worse at night and the patient may seek relief by walking around or stamping the foot. Paraesthesia and numbness may follow the characteristic sensory distribution, but these symptoms are not as well defined as in other entrapment syndromes.

The diagnosis is difficult to establish but nerve conduction studies may show slowing of motor or sensory conduction.

Treatment To decompress the nerve it is exposed behind the medial malleolus and followed into the sole; sometimes it is trapped by the belly of adductor hallucis arising more proximally than usual.