The Practice of Emergency and Critical Care Neurology

1. The Presenting Neurologic Emergency
2. Criteria of Triage
The Presenting Neurologic Emergency

• Serious casualty
• Instantaneously deteriorating illness
• Result of major trauma, acute stroke, infection or intoxication
• Urgent referral or walk-in
• Nonspecific symptoms (dizziness, diplopia, twitching, headaches)
The American Board of Emergency Medicine – Core competencies

- Stroke
- Demyelinating disorders
- Acute headache
- Acute Hydrocephalus
- CNS infection
- Guillain-Barre syndrome and myasthenia gravis
- Seizures
- Spinal cord compression and traumatic brain injury
The Emergency Department

• Highly dependent on location (inner-city vs rural)
• Many ED are packed
• Mixed pathology (nonurgent visits, “frequent flyers”)
• Designated critical care area
• Trauma activation (Level 1)
The Neurologic Emergency and its Assessment

- Specific clinical presentation
- Abnormal neuroimaging
- Progression of symptoms
- Neurologic symptoms often fluctuate (VBI)
Signs and Symptoms of Neurologic emergency

- Worsening and changing of neurologic signs
- Abnormal consciousness
- Seizure
- Inability to stand or walk
- Acute cranial nerve deficit/focal neurological deficit
- Severe unexpected headache
Neurologic Tests Available in ED

- CT
- MRI/A
- Cerebrospinal fluid examination
- Electroencephalography
Clinical Judgment in the ED - potential errors

- Missed acute brain injury on CT
- Failure to perform CSF exam
- Missed acute hydrocephalus
- Missed locked-in syndrome
- Missed early signs of brainstem displacement
- Not recognized status epilepticus
- Missed spinal cord compression
- Not recognized neurointerventional options
- Missed narrow windows of therapeutic opportunity
- Failure to recognize brain death and the possibility of organ donation
CONCLUSION

• Patients with critical neurological disorders are often seen in the ED
• The responsibilities of the neurologist are to assist in their evaluation, transfer them to operating room, interventional radiology suite or ICU
• The neurologist should be part of the ED team, facilitate recognition and treatment of unstable neurological patients
Criteria for Admission to the NICU

- Aneurysmal SAH
- Ganglionic, lobar, cerebellar or brainstem hemorrhage
- Major hemispheric ischemic stroke syndromes
- Basilar artery occlusion
- Cerebellar infarct
- Cerebral venous thrombosis
- Acute bacterial meningitis
- Brain abscess
- Myasthenia gravis
- Acute encephalitis
- Acute spinal cord disorders
- Acute white matter disorders
- Acute obstructive hydrocephalus
- Malignant brain tumors
- Status epilepticus
- Traumatic brain injury
- Guillain-Barre syndrome
CONCLUSION

• Any patient with a neurologic disorder and unstable vital signs (pulse rate, blood pressure, respiratory rate, core temperature) or a progressive neurologic presentation SHOULD BE ADMITTED

• Communication between ED and NICU physicians requires special effort.
Evaluation of Presenting Symptoms Indicating Urgency
“Confused and Febrile”
Clinical Assessment

• Debilitated, wasted, underfed (drug abuse, alcoholism, cancer)
• Exposure to ticks and beginning of endemic encephalitis (arboviruses)
• Exposure to wilderness, tropics, animal bite (rabies)
• Exposure to excessive heat (heat stroke)
• Recent travel or immigration from developing country (neurocysticercosis, fungal meningitis)
• Recent vaccination (ADEM)
• Prior transplantation or AIDS (Toxoplasma encephalitis or Aspergillus)
## Systemic Illnesses Producing Fever and Confusion

- Septic shock (any)
- Streptococcal shock syndrome
- Lobar pneumonia
- Acute osteomyelitis
- Abdominal suppuration
- Endocarditis
- Erysipelas
- Measles
- Psyttacosis
- Influenza
- Yellow fever
- Typhoid fever
- Cholera
- Heat stroke
- Thyrotoxicosis
- Anticholinergic drug intoxication
General Clinical Signs Indicating Causes in Confused Febrile Patient

• Skin rash (Rickettsial diseases, vasculitis, aspergillosis)
• Petechiae (Thrombocytopenic purpura, meningococcemia, endocarditis, drug eruption from intoxication, leukemia)
• Splenomegaly (Toxoplasmosis, tuberculosis, sepsis, HIV infection, lymphoma)
• Pulmonary infiltrates (Legionella species, fungi, tuberculosis, mycoplasma, pneumonia, Q fever, tick-borne disease)
Line of Action

Suspicion bacterial meningitis
1. Blood cultures
2. Vancomycin, cefotaxime or ceftriaxone, and dexamethasone
3. CT scan of the brain
4. CSF (Gram stain, cells, protein)
5. MRI/MRV
Suspicion encephalitis

1. CT scan + MRI (FLAIR, DWI, GAD)
2. CBC, platelets, chest x-ray, blood culture
3. CSF (routine PCR, IgM antibodies)
4. Empiric acyclovir
CONCLUSION

• Most confused and febrile patient have an underlying systemic infection
• Confusion may indicate a more specific language disorder
• Multisystem involvement and confusion may indicate certain infectious agents
• Abnormal immune status should be investigated because its presence has a different set of diagnostic possibilities.
“A Terrible Headache”
Warning Signs in Acute Headache

Signs and Symptoms
• Split-second onset, worst, unexpected
• Loss of consciousness, vertigo, vomiting
• Acute cranial nerve deficit (III)
• Carotid bruit in a young person
• Fever and skin rash
• Shock, Addison disease
• Fall and coagulopathy or anticoagulation

Diagnosis to consider
• Aneurysmal subarachnoid hemorrhage
• Cerebellar hematoma
• Carotid artery aneurysm
• Carotid artery dissection
• Meningitis
• Pituitary apoplexy
• Subdural, epidural or intracerebral hematoma
## Acute severe Headache Syndromes from Non-Neurologic Causes

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Location and Time</th>
<th>Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute angle glaucoma</td>
<td>Eye, frontal, acute</td>
<td>Red eye, mid range pupil, decreased vision</td>
</tr>
<tr>
<td>Temporal arteritis</td>
<td>Temporal, frontal, rapidly built up</td>
<td>Painful temporal artery, ESR &gt; 55 mm/h</td>
</tr>
<tr>
<td>Acute sinusitis</td>
<td>Frontal, maxillar, hours</td>
<td>Fever, pressure pain on maxillary or frontal sinus</td>
</tr>
<tr>
<td>Pheochromocytoma</td>
<td>Entire head, rapidly increasing intensity</td>
<td>Sweating, pallor, SBP&gt;200 mm Hg</td>
</tr>
<tr>
<td>Herpes zoster ophthalmicus</td>
<td>Eye, frontal, hours-days</td>
<td>Facial edema, vesicular rash may be delayed, visual loss</td>
</tr>
</tbody>
</table>
Symptomatic Thunderclap Headache Other than SAH

**Diagnosis**
- Hypertensive encephalopathy
- Cerebral venous thrombosis
- Retroclival hematoma
- Pituitary apoplexy
- CSF low pressure syndrome
- Carotid/vertebral a. dissection

**History and Exam**
- SBP > 200 mm/Hg
- Dehydration, BCP
- CSF xanthochromia
- Hypotension, hyponatremia
- Postural headache
- Trauma, DC, Horner’s, carotid bruit, dysarthria

**MRI features**
- T2/flair in parieto-occipital lobes
- Transverse/sagittal sinus-MRV
- Clot posteriorly/clivus level
- Pituitary tumor with hemorrhage
- Meningeal enhancement, SDH, “sagging brain”
- Recent cerebral infarcts; double lumen sign on MRI
### “Benign” Acute Headache Syndrome

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Location/Time/Quality</th>
<th>Characteristic Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Cluster headache</td>
<td>• Oculofrontal/30-90 min/severe</td>
<td>• Rocking, rhinorrhea, Horner</td>
</tr>
<tr>
<td>• Chronic paroxysmal hemicranias</td>
<td>• Unilateral/2-30 min/severe</td>
<td>• Conjunctival inject., lacrimation</td>
</tr>
<tr>
<td>• Acute migraine</td>
<td>• Unilateral/6-30 hr/mod. Severe</td>
<td>• Nausea, photophobia, 80% unil.</td>
</tr>
<tr>
<td>• Trigeminal neuralgia</td>
<td>• Unilateral/seconds/severe</td>
<td>• Triggered by chewing, cold wind, shaving, tooth brushing</td>
</tr>
</tbody>
</table>
Line of Action

• CT scan
• CSF (pressure, clarity, color before-after centrifuge, cell count, protein)
• MRI/A, possibly MRV
• Cerebral angiogram
# Abortive Therapies

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Therapy Options</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine</td>
<td>• Sumatriptan (6mg SC), repeat after 1 hour</td>
</tr>
<tr>
<td></td>
<td>• Droperidol (2.75-8.25 mg IM)</td>
</tr>
<tr>
<td></td>
<td>• Meperidine (100 mg IM) and hydroxyzine (50 mg IM)</td>
</tr>
<tr>
<td></td>
<td>• Valproate (1 g IV in 50 ml NS over 1 hour)</td>
</tr>
<tr>
<td></td>
<td>• Dihydroergotamine (1-3 mg IV) &amp; metoclopramide (100 mg IM)</td>
</tr>
<tr>
<td></td>
<td>• Prochlorperazine 10 mg (in 10 ml NS infused in 2 min)</td>
</tr>
<tr>
<td></td>
<td>• Oxygen therapy (7L/min face mask), metoclopramide (10 mg IM), sumatriptan (6 mg SC), nasal butorphanol (1 mg), intranasal lidocaine 4% (4 sprays)</td>
</tr>
<tr>
<td>Cluster headache</td>
<td>• Fosphenytoin IV loading (15-20 mg/kg IV)</td>
</tr>
<tr>
<td>Trigeminal neuralgia</td>
<td>• Carbamazepine (1200 mg/day)</td>
</tr>
<tr>
<td></td>
<td>• Lamotrigine (50-100 mg/day)</td>
</tr>
<tr>
<td></td>
<td>• Topiramate (50-100 mg/day)</td>
</tr>
</tbody>
</table>
CONCLUSION

• The evaluation of severe headache in ED is common
• The distinctive nature of a “thunderclap headache” needs to be recognized. CT will show a SAH (in most instances)
• Acute headache syndromes may be refractory migraine or cluster headache (good treatment options exist in the ED)
• Acute severe headache may have a non-neurologic cause
“Blacked Out and Stumped Down”
Causes of Syncope

- Naturally mediated (reflex)
- Orthostatic hypotension
- Cardiac arrhythmias
- Structural cardiac or cardiopulmonary disease
Naturally mediated (reflex) syncope

• Vasovagal syncope (common faint)
• Carotid sinus syncope
• Situational syncope (cough, sneeze, swallow, defecation, visceral pain, micturition, after exercise, after a meal)
• Glossopharyngeal neuralgia
Orthostatic hypotension

- Autonomic failure (primary autonomic failure - pure autonomic failure, MSA, Parkinson plus syndrome, secondary autonomic failure - diabetic neuropathy, amyloid neuropathy), drugs and alcohol, after exercise, after a meal
- Volume depletion (hemorrhage, diarrhea, Addison disease)
Cardiac arrhythmias

• Sinus node dysfunction (bradycardia-tachycardia syndrome)
• Atrioventricular conduction system disease
• Paroxysmal SVT and VT
• Congenital syndromes (long QT syndrome)
• Malfunction of an implanted device (pacemaker, defibrillator)
• Drug-induced arrhythmias
Structural cardiac or cardiopulmonary disease

- Obstructive cardiac valvular disease
- Acute myocardial infarction
- Obstructive cardiomyopathy
- Atrial myxoma
- Acute aortic dissection
- Pericardial disease
- Pulmonary embolus and pulmonary hypertension
CONCLUSION

• Cardiac arrhythmias can mimic seizures or be caused by seizures
• Patient with ictal bradycardia likely need a pacemaker
• Syncope rarely heralds an acute neurologic disease but may be part of an undiagnosed chronic neurologic illness
“See Nothing, See Double, See Shapes”
Degree of visual loss

- 20/200  Legal blindness
- 20/800  Finger counting
- 2/1000  Arm movements
- 20/∞   No light perception
Ophthalmologic Disorders

Diagnosis
• Central retinal artery occlusion
• Retinal vein occlusion
• Retinal detachment
• Ischemic optic neuropathy
• Optic neuritis
• Vitreous hemorrhage

Findings
• Afferent pupillary defect, retinal edema, optic disc pallor, cherry-red spots
• Extensive intraretinal hemorrhage
• Translucent gray wrinkled retina
• Pale optic nerve, swelling, scalp tenderness and absent temporal artery pulsation (temporal arteritis)
• Normal findings (“patient sees nothing, doctor sees nothing”)
• Diabetes, hypertension
## Diplopia Due to Cranial Nerve Palsy

<table>
<thead>
<tr>
<th>Cranial Nerve</th>
<th>Position of eye</th>
<th>Symptoms &amp; Signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>III</td>
<td>Down and out</td>
<td>Crossed diplopia, ptosis, dilated fixed pupil</td>
</tr>
<tr>
<td>IV</td>
<td>Higher</td>
<td>Vertical diplopia, head tilted away from affected side, chin down</td>
</tr>
<tr>
<td>VI</td>
<td>Inward</td>
<td>Uncrossed diplopia, head turned to affected side</td>
</tr>
</tbody>
</table>
Urgent Disorders in Acute Diplopia

**Acute III nerve palsy**
- Basilar a. aneurysm, PCOM aneurysm
- Pituitary apoplexy
- Acute midbrain infarct or hemorrhage
- Carotid cavernous fistula
- Granulomatous inflammation (Tolosa-Hunt)
- Diabetic microvascular disease

**Acute IV nerve palsy**
- Trauma, Carotid aneurysm, Meningitis (infectious or neoplastic)
- Herpes zoster ophthalmicus

**Acute VI nerve palsy**
- Cavernous sinus thrombosis, Nasopharyngeal carcinoma, Increased intracranial pressure
Line of Action

Acute oculomotor palsy

- Proptosis, Lid retraction, Red eye
- MRI/A, Cerebral angiogram, Neurosurgery consult

Monocular or binocular visual loss

- Optic disc lesion or edema
- Ophthalmology consult
- Sedimentation rate
- Temporal a. biopsy, MRI/CSF
CONCLUSION

• Neurologic causes of acute monocular (optic neuropathy) or binocular (occipital lobes) vision loss are less common than ophthalmologic causes
• Painful ophthalmoplegia has a broad spectrum of causes and need urgent evaluation
• Positive visual phenomena rarely indicate acute neurologic disease and more often are associated with neurotoxicity
“Spinning”
## Vertigo and Otologic Emergencies

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Clues</th>
<th>Therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Herpes zoster oticus</td>
<td>Ear lobe vesicles, hearing loss, facial palsy</td>
<td>Acyclovir 10-12 mg/kg IV q 8 X 10 days</td>
</tr>
<tr>
<td>Bacterial labyrinthitis</td>
<td>Acute deafness, prior cholesteatoma, meningitis</td>
<td>Surgical management, specific antibiotics</td>
</tr>
<tr>
<td>Malignant external otitis</td>
<td>Extreme ear pain, facial palsy (Pseudomonas aeruginosa)</td>
<td>Ciprofloxacin 750 mg po bid or gentamicin 1.7 mg/kg q 8</td>
</tr>
<tr>
<td>Perilymph fistula</td>
<td>Tinnitus, hearing loss, positional vertigo, barotrauma</td>
<td>Conservative first, then surgery</td>
</tr>
<tr>
<td>Labyrinth hemorrhage</td>
<td>Nausea, vomiting, hearing loss, trauma</td>
<td>Correct coagulopathy</td>
</tr>
</tbody>
</table>
“Can’t Walk or Stand”
Acute or Subacute Ataxia

• Intoxication and poisoning
• Acute occlusion of PICA
• Acute demyelination or multiple sclerosis
• Acute cerebellar ataxia (vaccinations, varicella zoster virus)
• Normal pressure hydrocephalus
• Paraneoplastic disease
# Acute paraplegia

<table>
<thead>
<tr>
<th>Disorder</th>
<th>History</th>
<th>Suggest</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myelitis</td>
<td>Vaccination, febrile illness, optic neuritis, travel, tick bite, immunsuppression, AIDS</td>
<td>Postvaccination myelopathy, postinfectious transverse myelitis, MS or Devic disease, schistosomiasis, cysticercosis, Lyme disease, TB, syphilis</td>
</tr>
<tr>
<td>Myelopathy</td>
<td>Cancer, AA or recent catheterization, low back pain, connective tissue disease (SLE), anticoagulation</td>
<td>Acute necrotic myelopathy, cord infarction, vasculitis, radiation myelopathy, paraneoplastic myelopathy, epidural hematoma, spinal AVM, dural AV fistula</td>
</tr>
<tr>
<td>Polyradiculopathy</td>
<td>Diarrhea, URI, CMV, HSV, EBV, DM, leukemia, sarcoidosis</td>
<td>GBS, acute diabetic polyradiculopathy</td>
</tr>
<tr>
<td>Neoplastic meningitis</td>
<td>Carcinoma, lymphoma</td>
<td>Infiltrative leptomeningeal spread</td>
</tr>
<tr>
<td>Neuromuscular Junction Disorders</td>
<td>Dysphagia, diplopia, ptosis, small lung cancer</td>
<td>Myasthenia gravis, Lambert-Eaton syndrome, botulism</td>
</tr>
<tr>
<td>Myopathy</td>
<td>Autoimmune disorder, thyrotoxicosis, exercise intolerance</td>
<td>Polymyositis, dermatomyositis, metabolic myopathy</td>
</tr>
</tbody>
</table>
Major Acute Spinal Cord Syndromes

Complete
Central
Hemisection
Anterior
CONCLUSION

• Acute gait disorder may be caused by viral infections or recent vaccinations in children and acute cerebellar infarction or medication overdose in the elderly

• Acute paraplegia may be a consequence of spinal cord compression, and neuroimaging with MRI is urgently needed

• Acute chest pain and paraplegia requires immediate evaluation for aortic dissection
“Short of Breath”
### Three Major Causes of Respiratory Failure in Acute Neurologic Disease

<table>
<thead>
<tr>
<th>Abnormal Respiratory Drive</th>
<th>Abnormal Respiratory Conduit</th>
<th>Abnormal respiratory mechanics</th>
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<td>Abnormal Respiratory Drive</td>
<td>Abnormal Respiratory Conduit</td>
<td>Abnormal respiratory mechanics</td>
</tr>
<tr>
<td>Sedatives (opioids, barbiturates)</td>
<td>Upper airway obstruction</td>
<td>Spinal cord lesion (trauma, MS, ALS)</td>
</tr>
<tr>
<td>Pontomedullary lesion (hemorrhage)</td>
<td>Massive aspiration</td>
<td>Absent or decreased neuromuscular junction traffic (MG, botulism, organophosphates)</td>
</tr>
<tr>
<td>Hypercapnia</td>
<td>Neurogenic pulmonary edema</td>
<td>Diaphragm weakness (myopathies, phrenic nerve lesion) or associated trauma</td>
</tr>
<tr>
<td>Hypothermia</td>
<td>Pneumothorax</td>
<td></td>
</tr>
<tr>
<td>Hypothyroidism</td>
<td></td>
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</tr>
</tbody>
</table>
Clinical Features of Imminent Neuromuscular Respiratory Failure

- Dyspnea at low levels of activity
- Restlessness
- Tachycardia (HR>100/min)
- Tachypnea (respiratory rate>20/min)
- Use of sternocleidomastoid, scalene muscles
- Forehead sweating
- Hesitant, constantly interrupting speech
- Asynchronous (paradoxical) breathing
Pulmonary Function Tests in Monitoring Neuromuscular Respiratory Failure

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Normal value</th>
<th>Critical value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vital capacity</td>
<td>40-70 ml/kg</td>
<td>20 ml/kg</td>
</tr>
<tr>
<td>Maximum inspiratory pressure</td>
<td>Male &gt;-100 cm H$_2$O</td>
<td>-30 cm H$_2$O</td>
</tr>
<tr>
<td></td>
<td>Female&gt;-70 cm H$_2$O</td>
<td></td>
</tr>
<tr>
<td>Maximum expiratory pressure</td>
<td>Male&gt;100 cm H$_2$O</td>
<td>40 cm H$_2$O</td>
</tr>
<tr>
<td></td>
<td>Female&gt;40 cm H$_2$O</td>
<td></td>
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</table>