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Corrective Surgery of Congenital Heart Defects in Neonates: the Prague Experience

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Aim. To assess the outcome and mortality trend in newborns undergoing corrective surgery for congenital heart defect.

Method. We reviewed the hospital records of 1,033 neonates under 30 days of life, who had congenital heart defects operated on at the Kardiocentrum, Motol University Hospital in Prague, Czech Republic, during 1977-2001. Early and late mortality and reoperation rates were analyzed.

Results. A total of 1,156 operations were performed in 1,033 neonates. Obstructive lesions were surgically treated in 56%, left-to-right shunts in 21%, and complex conotruncal lesions in 23% of the cases. Total correction has been achieved in 62% of the neonates. Most operations (75%) were performed in the first two weeks of neonate's life. Early 30-day hospital mortality was 13%. Late mortality, after the discharge from the hospital, was 10%. In the last three years, the hospital mortality rate decreased to about 2%. Out of 590 reinterventions in 379 neonates, with the mortality of 6%, 229 were secondary staged corrections and 190 further palliative procedures aimed mostly toward Fontan or Rastelli type of circulation. Residual or recurrent defects were solved in 62 neonates. There were 30 valve replacements, with 18 mechanical valves and 12 pulmonary valve autotransplantations (the Ross procedure). The homograft valved conduit was used in 38 children.

Conclusion. Most newborns with critical congenital heart defects can undergo corrective operation under acceptable risk. Due to complex improvements in perioperative, anaesthetic, surgical, and postoperative care, contemporary hospital mortality can be reduced to 1-3%. Palliative procedures still play an important role in the staged treatment of severe complex heart defects in neonates.

Key words: *aortic coarctation; cardiac surgical procedures; ductus arteriosus, patent; Fontan procedure; heart defects, congenital; heart septal defects; infant, newborn; tetrallogy of Fallot; transposition of great arteries*

The general prevalence of children born with congenital heart defects is 0.5-1.0% (1). In Czech Republic, it is 6.2 per 1,000 live births (1). Since 1977, all children born with congenital heart defects in the Czech Republic have been referred to the Kardiocentrum in Prague for urgent or elective corrective heart surgery. Urgently admitted children with a critical congenital heart defect, presenting with profound hypoxemia or heart failure, or both symptoms simultaneously, represent 35% of children born with congenital heart defects (or 2.36 per 1,000 live born children) (2). Since 1985, approximately 450-500 heart operations in children, including neonates, have been carried out yearly in our Center, similarly as in other centers in Europe. Early correction of critical congenital heart defect in a newborn or infant is required in the case of intractable heart failure caused by volume overload due to left-to-right shunt, pressure overload due to critical valvular stenosis, critical hypoxemia, and pulmonary hypertension. Unless surgically treated, all these conditions are fatal.

Progress in contemporary pediatric cardiac surgery has made the biventricular repair possible even in neonates (3). The goal of surgical correction of congenital heart defect in neonatal age is to establish normal circulation by abolition of pressure, or volume overload of the heart and critical hypoxemia. This can secure further normal growth and development of the child (4,5). Surgical advancements have allowed correction of heart defects even in newborns with low birth weight, after they achieve cardiovascular stability and show acceptable recovery of parenchymal organs compromised by major perinatal insults (6). On the other hand, palliative procedures can control congestive heart failure or hypoxemia in newborns, in which correction of the heart defect is not possible for anatomical reasons, infection, intracranial bleeding, or necrotizing enterocolitis. Palliative procedures are directed towards the establishment of controlled pulmonary blood flow or temporary removal of the obstructive lesions (7).

We have also considered the specific risks of early correction in particular congenital heart defects. The immaturity of tissues can influence the creation of unfavorable scars. The immaturity of liver and renal function under substantial load can lead to multiorgan failure. The reaction of immature immunological system to cardiopulmonary bypass causes general inflammatory reaction with the increased permeability of membranes and decreased resistance to infection. After early correction of the congenital heart defects in the newborn age, reoperation of recurrent or residual defects is more often needed than in children with primary correction at the later age (8).

The aim of this study was to overview our experience with correction of congenital heart defects in newborn children surgically treated at our institution in the last 25 years. During that period, operative techniques have advanced and operative mortality in Czech Republic decreased to 2.2% (in last three years) despite the increased complexity of surgical interventions.

Patients and Methods

We reviewed the hospital records and operative protocols of 1,033 neonates surgically treated at the Kardiocentrum, University Hospital Motol, Prague, during the 1977-2001 period. The age of neonates ranged from birth (day 0) to 30 days (median 8 days). The mean body weight of neonates was 3.4 ± 1.6 kg. Out of 155 newborns with low birth weight (< 2.5 kg), 30 weighed less than 1 kg.

Most operations (75%) were performed in neonates in the first (544 or 47%) and second week (323 or 28%) of life, whereas 151 (13%) and 138 (12%) operations were performed in neonates in the third and fourth week of life, respectively.

Operative mortality included deaths occurring within 30 days after the operation or during the same postoperation in-patient treatment period. Late deaths included fatal events after the patients' discharge from the hospital.

We used descriptive statistics to analyze the data. Results were presented either as mean values ± standard deviation or as medians with ranges.

Results

During the 1977-2001 period, a total of 1,156 cardiac operations were performed in 1,033 neonates. The median follow-up period was 12.5 years (range 1-24).

There were three main types of heart defects surgically treated in neonates: obstructive lesions, left-to-right shunts, and cotruncal lesions (Table 1). Out of 637 (56%) obstructive lesions, coarctation of the aorta was surgically treated in 270 neonates, pulmonary atresia in 136, and critical aortic stenosis in 99 neonates. Left-to-right shunts with volume overload of the heart were surgically treated in 240 (20.7%) neonates, with patent ductus arteriosus (90 neonates) and total anomalous pulmonary venous drainage (69 neonates) being the most frequent. Among complex conotruncal lesions surgically treated in 266 (23%) neonates, transposition of the great arteries was the most frequent (182 neonates). There were 13 operations for other cardiac leasions on neonates.

Complete correction was achieved in 717 (62%) neonates by either closed- or open-heart surgery, whereas palliative operations were performed in 439 (37.9%) neonates (Table 1). The most frequently operated heart defect in the newborns was coarctation of the aorta (270 neonates). It was also the most frequent defect among closed-heart surgical interventions. Out of 182 neonates with transposition of the great arteries, 162 underwent corrective open-heart surgery with heart/lung bypass, whereas 20 had palliative shunts. Transposition represents the most frequent defect corrected on open-heart in neonate. Shunts were most frequently constructed in neonates

Table 1. Types of cardiac surgery (N = 1,156) performed for the treatment of congenital heart defects in 1,033 neonates and mortality rates at Kardiocentrum Motol, Prague, in the 1977-2001 period

	No. of surgeries						
	1977-2001					1996-2001	
	corrections				mortality rate	No. of	mortality rate
Heart defect	with CPB[a]	without CPB	palliations	total	(No., %)	surgeries	(No., %)
Coarctation of the aorta		270		270	32 (11.8)	91	0
Transposition of the great arteries	162		20	182	14 (7.6)	112	1 (0.9)
Pulmonary atresia	2		134	136	17 (12.5)	30	0
Aortic stenosis	21		78	99	20 (20.2)	39	3 (7.5)
Patent ductus arteriosus		90		90	3 (3.3)	52	2 (3.8)
Ventricular septal defect	10		62	72	0	5	0
Total anomalous pulmonary venos drainage	69			69	18 (26.0)	17	3 (17.6)
Tetralogy of Fallot	2		38	40	5 (12.5)	24	2 (8.3)
Interrupted aortic arch	21		18	39	18 (46.1)	7	0
Pulmonary stenosis			37	37	4 (10.8)	20	0
Single ventricle			32	32	0	19	0
Hypoplastic left heart syndrome	24			24	2 (8.3)	24	2 (8.3)
Double outlet right ventricle	4		18	22	3 (13.0)	10	0
Persistent truncus arteriosus communis	13			13	0	7	0
Aortico-left ventricular tunnel	6			6	0	1	0
Atrioventricular septal defect	5			5	1 (20.0)	2	0
Double aortic arch		4		4	0	1	0
Congenital tumor of the heart	5			5	0	2	0
Atrial septal defect	2			2	0	0	0
Aorto pulmonary window	3			3	0	1	0
Ebstein's anomaly			2	2	1 (50.0)	0	0
Cor triatriatum	2			2	0	1	0
Hemitruncus	2			2	2 (100.0)	0	0
Total	353	364	439	1,156	140 (12.1)	465	13 (2.7)

[a]CPB – cardio-pulmonary bypass (extracorporeal circulation).

with pulmonary atresia (134 neonates). Critical aortic stenosis was most often treated by percutaneous balloon valvuloplasty (in 78 out of 99 neonates). Ductus arteriosus was closed in 70 premature babies out of 90 newborns. Other frequent heart defects solved by open-heart surgery were total anomalous pulmonary venous drainage in 69 neonates, hypoplastic left heart syndrome in 24 neonates, interrupted aortic arch in 21 neonates, and persistent truncus arteriosus in 13 neonates.

During the past 10 years, we have surgically treated around 400 children per year, with mortality rate constantly decreasing (Fig. 1). The number of surgical interventions in newborns constantly increased, from 34 in 1990 to 96 in 2001 (Fig. 1).

Early mortality after surgery in the whole group of 1,033 neonates was 12.9%, declining to 3.6% in last 5 years and to 2.2% in last 3 years (Fig. 2). The late mortality in the whole group of operated neonates was 10.1%.

After undergoing surgical intervention in neonatal age, 379 children needed reoperation. There were 590 reoperations performed during their first year of life and after, with 6% mortality rate (Table 2). Secondary stage corrections after initial palliative procedure in the newborn age were performed in 229 children. Reoperations of residual or recurrent defects were performed in 59 children (62 reoperations). There were 109 reoperations performed on the valves. In 150 children, 190 additional palliative procedures were performed on pulmonary arteries. There were 54 arterial shunts performed, 36 cavopulmonary shunts, 16 angioplasties of the pulmonary artery, and 84 pulmonary artery bandings.

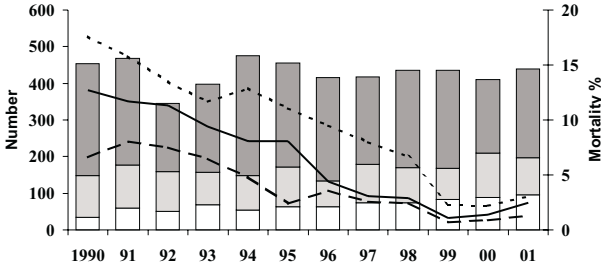

Figure 1. Cardiac surgery for congenital heart defects performed per year in children (age 0-18 years) at the Kardiocentrum Motol, Prague, and changes in their mortality rates during the 1990-2001 period. Total number of operations in children (N = 5,150) includes operations in newborns (n = 808) and infants (n = 2,021). All children (0-18 years) – dark gray bars; newborns (0-30 days) – white bars; infants (0-1 year) – light gray bars. Mortality rates: all children – dashed line; newborns – dotted line; infants – full line.

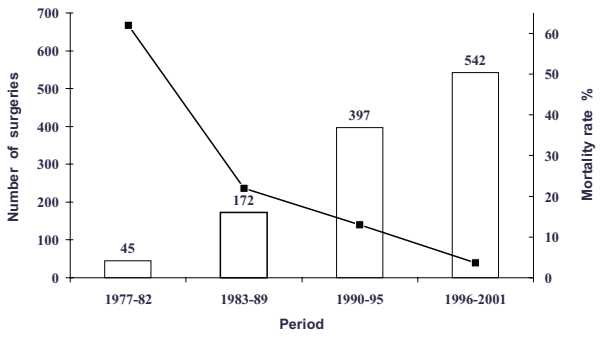

Figure 2. Decrease in mortality of newborns (N = 1,033) after cardiac operations (N = 1,156) at the Kardiocentrum Motol, Prague, during the 1977-2001 period. Bars – number of surgeries; line – morality trend.

Table 2. Reoperations (N = 590) performed after the first stage of surgery in 379 neonates at the Kardiocentrum Motol, Prague, in the 1977-2001 period

Type of reoperations	No. of reoperations
Second stage corrections	229
Reoperations for residual/recurrent defect:	62
– recoarctation	49
secondary resection	16
ballon dilatation	33
– intracardiac shunt	3
– subaortic stenosis	5
– coronary artery stenosis	3
– vascular ring	2
Reoperations on valves:	109
– valve replacement	30
mechanical valve	18
pulmonary autograft	12
– homograft valved conduit	38
– balloon valvuloplasty	41
aortic valve	26
pulmonary valve	15
Secondary palliative procedures:	190
– arterial shunt	54
– cavopulmonary shunt	36
– pulmonary artery plasty	16
– pulmonary artery banding	84
Total	590

Out of 62 reoperations of residual defects, 49 were recoarctations, treated with 16 repeated resections and 33 percutaneous balloon catheter angioplasties. Recoarctation rate decreased to 9.6% in the last 5 years.

Out of 13 other children with recurrent defects, 2 had residual ventricular septal defect with left-to-right shunt, 5 had recurrent subaortic stenosis, 3 developed coronary artery stenosis after switch procedure, and 2 had recurrent vascular ring.

Out of 109 reoperations on the valves, 30 were late valve replacements. Eighteen mechanical valves were implanted into aortic, mitral, and tricuspid orifices, and 12 pulmonary valve autotransplantations were performed to construct a new aortic root according to the Ross procedure (9). Aortic or pulmonary homografts for the reconstruction of the right ventricular outflow tract were used in 51 children. Thirteen neonates had primary homograft implantation and 38 children underwent homograft exchange. In this group, 41 additional balloon catheter reinterventions were performed for 26 aortic valve restenosis and 15 pulmonary valve restenosis.

Discussion

The most common critical heart defect in the group of 270 newborns was the coarctation of the aorta. Today, most neonates with coarctation do not have to be operated on emergency basis. Their hemodynamic status can be stabilized before repair by prostaglandin E_2, which allows depressed abdominal

organ function to recover under stabilized ductus-dependent circulation (6). We prefer extended resection of coarctation in the neonate, with shifting of the anastomosis either below the left carotid artery if the hypoplastic aortic arch is present, or directly into the ascending aorta. In such operations, complete removal of all ductal tissue, tension-free repair, and precise suturing technique become particularly important. The anastomosis on the aorta is long and oblique, and sutured by very thin monofilament continuous polypropylene stitch 7/0 or 8/0, or resorbable monofilament material 7/0. The hypoplastic aortic arch can be enlarged by the reversed Hart and Waldhausen (10) or Amato (11) technique. In complex forms of coarctation with ventricular septal defect, we prefer resection of coarctation and pulmonary artery banding. Most ventricular septal defects later become restrictive or close spontaneously. We perform elective closure of large ventricular septal defects during the second month of the child's life.

The hospital mortality in neonates with coarctation decreased to zero in the last years. The weight of the child and type of repair did not correlate with the risk of recoarctation in our group of neonates. Among 23 neonates with body weight less than 2.5 kg, only a single recoarctation developed (4.3%). This corresponds to the findings of McElhinney et al (12). Most recurrences appeared within a year after repair, and less frequently within the period of 3 to 5 years after initial operation in neonatal age. Our results did not support the findings of Bacha et al (13), who found the child's body weight less than 1.5 kg at the time of repair to be the only independent predictor of recurrent aortic arch obstruction. Recurrent coarctations can be solved with catheter balloon dilatation quite effectively in most cases (14). Stents are not used in children during their growth. Reoperation is recommended in long narrow or bent segment of isthmus or hypoplasia of the aortic arch.

Transposition of the great arteries in newborns varies from simple forms of transposition to forms with restrictive foramen ovale defects or aortic arch obstruction that often lead to profound hypoxemia and acidosis of the newborn. In most cases, the initial resuscitation includes balloon atrial septostomy and administration of prostaglandin E_2 to ensure that ductus arteriosus remains open. Two-dimensional echocardiography provides rapid and complete noninvasive diagnosis, showing coronary anatomy in detail, making catheter investigation in neonate with transposition of the great arteries rarely necessary (4). We use angiocardiography in coronary anomalies not clearly visible on echocardiography or in cases of complex transposition of the great arteries when combined with interruption of the aortic arch or total anomalous pulmonary venous drainage. Correction of the transposition of the great arteries has been shifted from previous atrial repair in infant age towards the arterial switch operation mostly performed during the first two weeks of life as a method of choice (15). We performed 90 switch operations during the last 4 years and none of the operated newborns died. The anatomic variations of the origin and course of coronary arteries, including the intramural course of left coronary artery, as well as the weight of the newborn child (down to 2 kg) are not contraindications for arterial switch operation (16). Out of 9 newborns with weight below 2.5 kg, none died after arterial switch operation. Transposition with ventricular septal defect, coarctation of aorta, and low birth weight are reasons for staged procedure, with resection of coarctation being done first and later followed by arterial switch operation. Late trivial aortic regurgitation occurs in 10% of patients after arterial switch operation.

Rare late complications also require attention. Occluded coronary arteries have to be reoperated for recanalization or revascularization of the area supplied by the affected coronary artery (17). Out of three revisions for coronary artery ostial stenosis or secondary atresia among 168 arterial switch operations, we performed one successful ostioplasty of the left coronary artery and one aortocoronary venous bypass grafting of the occluded right coronary artery in a 2-year-old child. The revascularization was still functional after a 2-year follow-up. The second child underwent reimplantation of stenotic left coronary artery 5 days after the primary arterial switch operation. Only a single child died after the revision of stenotic left coronary artery performed two months after the arterial switch operation.

Shunts are optimal palliative treatment of pulmonary atresia in neonates. Staged correction is shifted to infant and/or later age. Because of the complicated spectrum of anatomical defects with pulmonary atresia, it is necessary to follow a particular plan of staged correction from the very beginning of surgical treatment (18). In cases with pulmonary atresia and ventricular septal defect, further treatment depends on the size and morphology of intrapericardial pulmonary artery branches and pulmonary segmental blood supply. It is necessary to unifocalize the multifocal big collateral arteries which supply the lungs and temporary connect them by arterial shunt to control the growth of the reconstructed pulmonary artery branches at about 6 months of age. Total correction of this complex heart defect is performed in the second stage, after satisfactory capacity of the pulmonary vascular bed has been reached. The development of pulmonary arteries is measured against the McGoon ratio and/or Nakata index (cf. 19). Total correction is performed in older children, in whom the outflow tract of the right ventricle can be reconstructed with a fresh frozen homograft of the pulmonary valve (20).

Depending on the size of tricuspid valve anulus and the right ventricular cavity with the initial arterial shunt, children with pulmonary atresia and intact ventricular septum can later undergo one of the following treatments: a) biventricular correction with pulmonary artery reconstruction, realized if the right ventricular size and tricuspid valve anulus diameter are 60% greater than diameter normal for the age; b) correction with leaving the small right ventricle in circulation and construction the bidirectional cavopulmonary shunt if right ventricle or tricuspid anulus are 60% smaller than normal; and c) modified Fontan op-

eration of univentricular heart if the right ventricle capacity's is very small or coronary sinusoides are present.

Critical aortic stenosis presents with a circulatory shock and low cardiac output usually immediately after closure of patent ductus arteriosus in the newborn. The procedure of choice at our Center is urgent percutaneous balloon valvuloplasty of critical aortic stenosis, after which the intractable heart failure usually quickly falls back. Surgery in very ill newborns, especially those with low birth weight, carries high risk of death (21). Occasionally, severe residual aortic valve regurgitation appears as a complication of the valvuloplasty after tear off of the thick dysplastic aortic valve leaflet. Neonates usually tolerate severe aortic valve regurgitation if supported by cardiotonics and diuretics. The hemodynamically severe aortic regurgitation can be managed during the first year of life with the Ross procedure – the aortic root replacement with pulmonary autograft even in the neonate (22). We have done two neonatal Ross operations and another seven during the first year of life. The pulmonary autograft in the position of the aortic root grew along with the heart (23).

Patent ductus arteriosus often loads the circulation of premature newborn with respiratory problems due to immature lung parenchyma. The very low systemic diastolic pressure with the reversal flow in the aorta during the diastole is compromising the perfusion and oxygen supply to the brain and abdominal organs. The method of choice for closure of patent ductus arteriosus in premature child is the farmacological administration of indomethacin or ibuprofen. If this fails, the surgical closure is indicated. We prefer performing the operation directly in the incubator at the intensive care units for premature neonates at obstetric departments rather than moving neonates to pediatric cardiac surgical unit. Such preterm babies are critically ill, mechanically ventilated, and have several venous lines introduced. The preterm neonate with the lowest birth weight surviving patent ductus arteriosus closure in our series of 70 operations had 400 g.

The isolated ventricular septal defect usually does not compromise circulation in the neonate. It is the most common component of complex heart anomalies and requires closure together with the correction of the complex defect or, in some newborns, palliative pulmonary artery banding until the total correction can be performed.

Total anomalous pulmonary venous drainage has to be corrected in neonates because no palliative alternative is available. The volume overload of the heart in neonates has to be urgently treated with heart-lung bypass. We have operated on 69 neonates with a total anomalous pulmonary venous drainage. The mortality of the intervention was 26% and decreased to 17.6% in last five years. One of the most severe early risk factors is pulmonary hypertensive crisis after the correction of total anomalous pulmonary venous drainage. Later complication of the operation is the development of progressive pulmonary venous obstruction, which occurred in 7 cases in our series. The reoperation of this complication is one of the most difficult surgical tasks.

Single ventricle defects require the staged surgical treatment with initial palliation, arterial shunt, or pulmonary artery banding in neonates, followed by bidirectional cavopulmonary shunt at 6 months and modified Fontan procedure at the age of two. Neonates with hypoplastic left heart syndrome, with aortic and/or mitral valve atresia or stenosis undergo palliative treatment with Norwood (24) or Damus-Kaye-Stansel operation (25). The single functioning right ventricle is connected through the pulmonary trunk directly with the aortic arch and descending aorta. The pulmonary artery is supplied from the right subclavian artery through a modified arterial Blalock shunt or through the shunt from the right ventricular outflow tract. Neonates elective for this procedure profit from this first-stage operation, followed by a bidirectional cavopulmonary shunt at 6 months of age and completed by modified Fontan operation – the total cavopulmonary connection at the age of two. Since 1998, we have surgically treated 24 neonates with hypoplastic left heart syndrome. The 30-day mortality rate was 8.3%. Before 1998, children with hypoplastic left heart syndrome had not been indicated for operation due to generally poor outcome of Norwood procedure. After initial Norwood procedure we performed second-stage bidirectional cavopulmonary shunt in 13 children and in 5 children completion of total cavopulmonary connection. There were 3 late deaths. The hypoplastic left heart syndrome is the most challenging defect for surgical treatment and a measure of cardiac team capability (4). The indications for staged univentricular Norwood procedure and/or heart transplantation are still to be solved (26,27).

Interrupted aortic arch is characterized by the absence of an anatomic continuity within the aortic arch or the aortic isthmus. Associated anomalies include ventricular septal defect, left ventricular outflow tract obstruction, persistent truncus arteriosus, transposition of the great arteries, aortopulmonary window, and double-outlet right ventricle. Careful echocardiographic evaluation of coexisting anomalies is essential. The presence of interrupted aortic arch is an indication for urgent surgical intervention (28). Intensive preoperative treatment includes prostaglandin infusion, anticongestive medication, and ventilation as the newborn may develop congestive heart and renal failure with metabolic acidosis. One-stage operation through median sternotomy includes the reconstruction of the aortic arch by the anastomosis of descending to ascending aorta and the repair of all intracardiac anomalies, which is the method of choice. Severe risk factor in postoperative morbidity is concomitant DiGeorge syndrome for the development of pneumonia or sepsis. Out of 7 operations in the last 5 years, no newborn with interrupted aortic arch died in our cohort.

Persistent truncus arteriosus in neonates presents with congestive heart failure, which needs urgent correction by heart-lung bypass, especially if additional regurgitation of the dysplastic truncal valve is present

(29). The large escape of blood volume into the lung circulation decreases the systemic diastolic pressure, which compromises coronary artery perfusion and leads to myocardial ischemia and left ventricular dysfunction. There is no effective palliation available. The reconstruction of the right ventricular outflow tract requires a small fresh human pulmonary allograft or xenograft Hancock conduit with the diameter of 12 mm. These small human allografts sometimes undergo early accelerated fibrocalcific degeneration, which is most probably the result of the antigenic immunity reaction (30-32). Therefore, we use the allograft matched with ABO blood group to diminish the immunity reaction of the host. Since the child outgrows the original small allograft or xenograft, it has to be replaced with large-sized allograft (16-20 mm) at the age of four or five years. This allograft assures sufficient diameter of the pulmonary valve until the adult age.

The substantial improvement of survival of neonates with critical heart defects was made possible by improvement in complex perioperative care, ie, improved early recognition of newborns with critical heart defect in neonatal units, early and reliable echocardiographic diagnosis with prenatal screening of heart defects, and perinatal intensive care with administration of prostaglandin E_2 in ductus-dependent heart defects, which enables the neonate to reach the pediatric cardiosurgical department in time and in general good condition (5). Mechanical ventilation can be adjusted to achieve a carbon dioxide tension considered optimal to manipulate the pulmonary vascular resistance (6). Also, there has been a substantial progress in the choice and timing of the most suitable operation for correction of particular critical heart defect. Furthermore, microsurgical technique is being constantly developed, the heart-lung machines have been miniaturized, optimal technique of deep hypothermia adapted, hemodilution and priming volume diminished, and ultrafiltration made more effective (7,33,34). The specific and highly effective postoperative care concentrated on the stabilization of circulatory and respiratory problems, renal functions, prevention of the capillary leak syndrome, prevention of hypertensive crises, and early nutrition of the newborn child after surgery (5).

Effective treatment of newborns with congenital heart defects depends on the teamwork of pediatric cardiologists, anesthesiologists, cardiac surgeons, and intensive care unit personnel. This team should cover the whole health care of such children, starting with an early and reliable recognition of congenital heart defects in newborns and the precise diagnosis of the heart defect. Perinatal intensive care should provide support to the failing heart and reduce effects of hypoxemia in a child with ductus-dependent circulation before operation, as well as stabilize the vital functions of the child.

Most newborns with critical congenital heart defects can be operated today with an acceptable risk. Complex improvements in perioperative, anaesthetic, surgical, and postoperative care make feasible the decrease of contemporary hospital mortality to 1-3%. Although palliative procedures are being replaced by corrective procedures today more than ever, they still play an important role, especially in the staged treatment of severe complex heart malformations.

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