Anesthetic management of neonates undergoing palliative operations for congenital heart defects

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Considerable progress has been made in the treatment of congenital heart defects in patients of all ages during the past 2 decades. Further advances have been made in neonates and infants in the past decade. The application of open heart surgical techniques in this group has been enhanced by the use of deep hypothermic circulatory arrest. Palliative or partial corrections have been used in neonates for either of two reasons: (1) mortality associated with complete repair was too high, or (2) definitive repair for many types of defects was not possible. Although these principles are applicable in some cases today, definitive corrective operations have been successfully accomplished for most cardiac defects. Improved techniques and success of deep hypothermic cardiopulmonary bypass in neonates and infants have led many to challenge the traditional concepts of palliative closed heart operations.¹⁻⁴ Since the risks of operations in these patients are related largely to intraoperative events, their anesthetic management is of considerable importance.⁵ Accordingly, we have analyzed our experience with palliative closed heart operations in neonates and infants during the past 20 years.

Patients

From January 1, 1960, to December 31, 1979, 675 patients underwent closed palliative surgical

153

procedures at the Cardinal Glennon Memorial Hospital for Children* (Table 1). Of these, 378 (56%) were younger than one year old at the time of operation and 297 (44%) were older. About one fourth of the total number (169 patients) were neonates younger than one month). We have compared postoperative survival in neonates with that of infants from one to 6 months old and also from 6 months to one year old in order to examine the influence of age on mortality. We have included coarctation repair and pulmonary valvotomy in Table 1 for reasons we will discuss later.

During 20 years' experience, there has been considerable change in the philosophy regarding timing and choice of operation. Figures 1-3 show the fluctuations from year to year in the number of patients undergoing palliative closed heart procedures, in neonates younger than 30 days old, infants from one to 6 months, and from 6 months to one year. For the entire group (newborn to one year old) (Fig. 4) the largest number of operations were performed in the late 1960s. Mortality was considerably higher in children younger than one year. Moreover, there was a striking difference in mortality among the three age groups younger than one year, 42.6% mortality in neonates, 23% mortality in those between one month and 6 months old, and no deaths in those between 6 months and one year old.

Anesthetic management

Just as indications, timing, and choice of operations have changed, a variety of anesthetic techniques have been used during the 20-year period. Many anes-

Table 1. Closed palliative* cardiacsurgery; 1960–1979

Age groups	Patients No.	Deaths No.	Mortality %
>1 yr	297	8	2.7
<1 yr	378	110	29.1
6 mo-1 yr	44	0	0
>1-<6 mo	165	38	23.0
<1 mo	169	72	42.6

* No patent ductus.

thetic agents were administered including cyclopropane, ether, methoxyflurane, halothane (Fluothane), nitrous oxide, muscle relaxants, intravenous narcotics, and in recent years ketamine. The choice of anesthetic has been influenced by the infant's condition upon arrival in the operating room. Many infants were critically ill with low cardiac output, hypoxemia, and severe acidosis, having arrived in the operating room moribund, or having sustained a cardiac arrest before anesthesia. Inappropriate anesthesia under these circumstances may precipitate cardiovascular collapse because of adverse effects on the systemic or pulmonary circulations. Vigorous efforts are necessary to prevent cardiovascular collapse. Successful resuscitation is unlikely if ventricular fibrillation occurs in compromised critically ill infants.

These infants are premedicated with atropine, 0.05 to 0.1 mg, and are taken to the operating room with full support, including oxygen, intermittent positive pressure ventilation, and frequently inotropic drugs.

A femoral venous catheter is usually left in place at the conclusion of cardiac catherization. A plastic percutaneous peripheral cannula is placed if a femoral venous access is not in place when the infant arrives in the operating room. A 19-gauge catheter is frequently intro-

^{*} Saint Louis University Medical Center Children's Hospital.

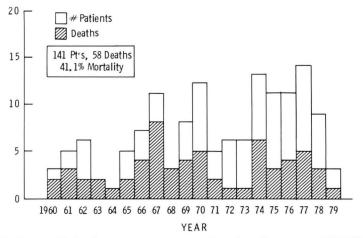


Fig. 1. Closed palliative (no coarctation or patent ductus) cardiac surgery, 1960-1979.

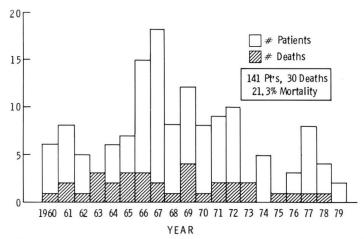


Fig. 2. Closed palliative (no coarctation or patent ductus) cardiac surgery, 1960-1979; infants, 1 to 6 months old.

duced into the internal or external jugular vein over a 0.018-inch flexible guide wire into the superior vena cava for monitoring central venous pressure.⁶

If an endotracheal tube is not already in place, pancuronium, 0.1 mg/kg, is given intravenously, and nasotracheal intubation is performed. Since only three of the 169 neonates in our series weighed less than 2.25 kg (5 pounds), a 3.0- or 3.5-mm endotracheal tube was almost always introduced. The endotracheal tube is initially placed orally to test for ease of insertion and magnitude of any air leak around the tube, and then the correct size is introduced nasotracheally. A small amount of topical 2% lidocaine is sprayed onto the larynx before intubation. The electrocardiogram is monitored with an oscilloscope. A nasograstric tube, esophageal stethoscope, and nasopharyngeal thermistor

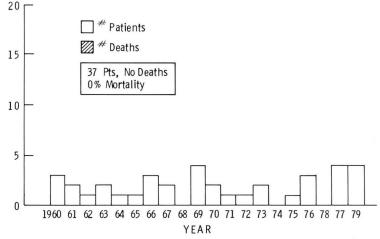


Fig. 3. Closed palliative (no coarctation or patent ductus) cardiac surgery, 1960-1979; infants, 6 months to 1 year old.

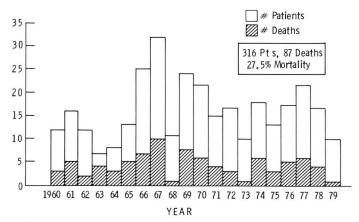


Fig. 4. Closed palliative (no coarctation or patent ductus) cardiac surgery, 1960-1979; newborn to 1 year old.

are appropriately positioned. Temperature control is maintained with a warming mattress and infrared lamps.

Although arterial vascular access often presents a challenge in these neonates, arterial monitoring is mandatory. Percutaneous 22-gauge catheter insertion into the radial or dorsalis pedis artery has been accomplished in 70% of the neonates, and the remainder have required cutdowns. In past years we have cannulated the internal mammary artery following thoracotomy.⁷ More recently we have cannulated the superficial temporal artery. Femoral and brachial artery cannulations are considered as a last resort.

If an intravenous line is not functional when the patient is brought to the operating room, ketamine is frequently injected intramuscularly in a dose of 3 to 5 mg/pound of body weight to facilitate induction of anesthesia and venous access.

Currently, our management of anesthesia combines intravenous analgesia with either morphine, 1 mg/kg, or fentanyl, 25 μ g/kg, as a maximum total dose. The infant is immobilized by the intravenous administration of pancuronium, 0.1 mg/kg. We prefer to use 100% oxygen for patients with systemic arterial unsaturation (cyanosis) or poor preoperative status. If high oxygen concentrations are not considered essential, 50% nitrous oxide is usually employed. Although halothane is not specifically contraindicated in the good risk neonate, it should be given cautiously and in low concentrations (probably less than 0.5%) to avoid its known myocardial depressant effects in neonates. It should be avoided particularly in situations where the need for inotropic stimulation with a catecholamine is anticipated.8

Ventilation during operation is an important aspect of management, and can be performed manually or with a ventilator, although we prefer rapid manual ventilation. It is our impression from clinical evidence that tachypnea, usually present before anesthesia, is the preferred pattern of ventilation. Hypocapnia induced before interruption of the blood flow to one lung during shunt procedures may be beneficial by delaying the onset of acidosis. Intracardiac shunting and diminished pulmonary perfusion may be further accentuated by hypoxia and hypercarbia. Respiratory acidosis may be present even when ventilation appears adequate, due to the reduction of pulmonary perfusion or intracardiac shunting or both. Frequent intraoperative arterial blood gas determinations are essential in guiding therapy.

Cardiovascular collapse is usually preceded by bradycardia and cardiac dilation, followed by asystole or ventricular fibrillation. The anesthesiologist must be prepared to reverse these events by inotropic myocardial stimulants and adjustments in ventilatory dynamics. Since surgical maneuvers to increase pulmonary blood flow immediately may not be possible, initial bolus injections of calcium chloride, 10 to 20 mg/kg; isoproterenol, 0.01 to 0.1 μ g/kg; atropine, 0.1 mg; or sodium bicarbonate, 1 to 2 mEq/kg may reverse the undesirable processes. If this regimen does not improve cardiac rate and contractility, 1 ml of a 1:10 dilution of 2% lidocaine with 1:100,000 epinephrine is given intravenously. These drugs may have to be repeated if bradycardia recurs.

All drugs must be prepared in advance and the intravenous administration system arranged so that drug injection results in immediate introduction into the venous circulation. We use a Tee connector with an extension (Abbott Laboratories, Inc, North Chicago, Illinois) attached to the plastic intravenous cannula. Ten-milliliter syringes of the appropriate drugs are connected to scalp vein sets (Butterfly, Abbott Laboratories, Inc) and introduced into the Tee. As many as four different scalp vein needles can be introduced into the Tee. Multiple stopcock manifolds are also available but are somewhat more cumbersome. The necessity of washing the drug into the patient from the manifold and its connecting link to the intravenous cannula delays the onset of drug effect. Intravenous lidocaine in boluses of 1 mg/kg before cardiac instrumentation may prevent or reduce the incidence of ventricular fibrillation. For prolonged inotropic support postoperatively, we prefer to use continuous intravenous infusions of isoproterenol or dopamine, frequently in combination with the peripheral dilator, nitroprusside.

Care must be taken in administering intravenous fluids and blood to avoid inadvertent circulatory overload and the resultant interstitial pulmonary edema, which further impairs oxygen transfer across alveolar membranes. This can be avoided by the use of volume infusors, which facilitates programmed administration of fluids. We replace blood loss in excess of 10% with 5% albumin in lactated Ringer's solution. In infants with polycythemia we use 5% albumin in lactated Ringer's solution as long as the hematocrit value is above 45%.

Respiratory support before, during, and after surgery may determine its success or failure. If respiratory insufficiency is present preoperatively, it usually persists postoperatively. Postoperative respiratory support is required for most neonates.⁹ Continuous positive airway pressure and intermittent mandatory ventilation are frequently used during weaning from controlled mechanical ventilation. Extubation is accomplished when there is evidence of satisfactory ventilatory dynamics, no evidence of excessive work, and satisfactory arterial blood gases on 3 cm H₂O continuous positive airway pressure.

Since the type of defect and operation performed may alter the anesthetic technique, specific comments regarding anesthetic management for the various palliative procedures will be discussed for each entity. Although a variety of cardiac anomalies were present in our 675 patients, the palliative procedures were designed to (1) increase pulmonary blood flow, (2) decrease pulmonary blood flow, (3) promote mixing between the pulmonary and systemic circuits, (4) decrease the afterload on a specific cardiac chamber, or (5) correct the lesions that were correctable in infants with multiple defects.

Increasing pulmonary blood flow

Shunt procedures to increase pulmonary blood flow were performed in 50 neonates with an overall mortality of 40% (Table 2). Since 60% of those who had a Waterston shunt died, it has not been used in recent years. The infants who died had excessive shunting, pulmonary congestion, and frank tracheal bleeding because of difficulty in controlling orifice size. More recently, the use of magnification and microsurgical techniques have made performance of the Blalock-Taussig shunt possible in neonates.¹⁰ The use of polytetrafluoroethylene (Impra, Inc, Tempe, Arizona) grafts allow control of the orifice size and provide an alternative to the Blalock-Taussig shunt when the subclavian anatomy is not suitable^{1, 11} or when a short-term shunt is required for palliation before definitive correction.

The intravenous infusion of prostaglandin E_1 before and during shunt procedures may increase flow through a closing ductus arteriosus and allow survival until a shunt can be established.^{12, 13} In a few cases we have used temporary intraluminal silastic shunts to reduce afterload during coarctation repair or to increase pulmonary flow during systemic to pulmonary artery anastomosis. We routinely administer heparin, 1 mg/kg, intravenously before occlusion of the pulmonary artery to prevent thrombus formation and shunt occlusion.

Although the Glenn shunt has been useful in our overall experience, it proved fatal in neonates and infants younger than 6 months.¹⁴ Although no patients died during operation, five of six neonates died during the immediate postoperative period. Mortality was related to the high pulmonary vascular resistance in the neonates producing

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Type of procedure	Age							
	<1 mo		>1-6 mo		>6 mo-1 yr		Total	
	Patients No.	Deaths No.	Patients No.	Deaths No.	Patients No.	Deaths No.	Patients No.	Deaths No.
Blalock-Taussig	18	3	18	4	13	0	49	7
	16%		22%		0%		14%	
Waterston	15	9	2	0			17	9
	57%		0%		0		53%	
Systemic PA	11	3	2	0			13	3
(Impragraft)	27%		0%		0		23%	
Glenn	6	5	22	6	3	0	31	11
	83%		27%		0%		35%	
Total	50	20	44	10	16	0	110	30
	40%		23%		0%		27%	

Table 2.	Closed palliative cardiac surgery; 1960-1979 (systemic to pulmonary
	artery shunt; mortality by procedure and age)

% = percent mortality, PA = pulmonary artery.

cerebral edema and central nervous system damage, and the frequent occurrence of chylothorax.¹⁴ Accordingly, Glenn shunts are no longer used in infants younger than 6 months. Continuous superior vena caval pressure monitoring and the administration of nitroprusside into the superior vena cava to reduce pulmonary vascular resistance have decreased morbidity in older infants.

Decreasing pulmonary blood flow

During the early years of this experience, pulmonary artery banding was the mainstay of therapy for infants with severe pulmonary congestion due to leftto-right shunting.¹⁵ Of 110 patients who underwent pulmonary artery banding, 18 were neonates (*Fig. 5*). In 11 of these 18, pulmonary artery banding was the only surgical procedure performed. Mortality in the entire group of 110 was only 18%, but in the neonates it was 82%, which has led us to avoid banding in this age group whenever possible. In a recent review of pulmonary artery

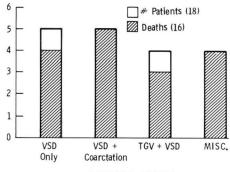




Fig. 5. Closed palliative cardiac surgery, 1960– 1979; pulmonary artery banding (infants <30 days old). VSD = ventricular septal defect, TGV = transposition of great vessels, misc = miscellaneous.

banding for a variety of indications, mortality for those without simple ventricular septal defect was high and the band often led to outlet reconstruction problems at the time of later definitive repair.^{16, 17} Diagnoses and mortality in the 18 neonates who were banded are shown in *Figure 5*. Presently, we prefer to perform complete repair during car-

diopulmonary bypass for ventricular septal defect alone, atrioventricular canal, and transposition with ventricular septal defect. Complete repair or other palliative techniques are used for truncus arteriosus.

Early in our experience, the combined lesion of ventricular septal defect and coarctation was treated by a combination of coarctation repair and pulmonary artery banding. Because six of eight infants died, we abandoned this combined approach. Only one of seven infants with ventricular septal defect and coarctation died after repair of coarctation alone. Patients with the combination of coarctation and ventricular septal defect, with or without a patent ductus arteriosus, are now treated by initial coarctation repair and patent ductus ligation.¹⁸ If severe heart failure persists, ventricular septal defect closure during cardiopulmonary bypass is performed. Presently, our indications for pulmonary artery banding in a neonate are multiple ventricular septal defects, some cases of univentricular heart, or double outlet right ventricle.

During pulmonary artery banding, the anesthesiologist must carefully coordinate his activities with those of the surgeon. We monitor pressure in a systemic artery, either radial or mammary, in the pulmonary artery distal to the band; and in the pulmonary or "common" ventricle. This technique has been previously reported.¹⁵ An increase in systemic pressure of 20% with a decrease in pulmonary artery pressure of 30% to 40% is usually considered a sufficient amount of constriction. Distal pulmonary arterial pressure should not be lowered to less than 30 mm Hg. Arterial blood gases must be checked immediately thereafter and at intervals to determine whether excessive right-to-left Vol. 48, No. 1

shunting has been produced. We obtain at least one determination of PaO_2 at an F_IO_2 of 0.4 or less. Ventricular irritability may occur during the pericardiotomy, and surgical retraction is necessary to isolate the pulmonary artery before surgical constriction. Lidocaine, 1 mg/kg, intravenously will reduce ventricular irritability.

Procedures to promote mixing between the systemic and pulmonary circulation

The Blalock-Hanlon atrial septectomy was performed in 57 neonates, with an overall mortality of 32% (Table β). The mortality in neonates with simple transposition was 31%. In those with complex transposition or other complex cardiac lesions the mortality was 35%. In the early years of this review, before the introduction of the Rashkind atrial septostomy, the Blalock-Hanlon septectomy was the procedure of choice. Presently, it is considered only when Rashkind atrial septostomy is not successful. In our past experience, the Blalock-Hanlon procedure prolonged palliation and allowed us to delay the Mustard operation¹⁹ until 9 months to one year of age to avoid the higher risk of late vena caval obstruction, which has been common when surgical correction is performed in infancy.²⁰ However, we and those at other centers have successfully performed Mustard's operation¹ or the modified Senning operation²¹ in infants for whom the Rashkind atrial septostomy did not provide palliation. As open heart surgical techniques improve, the Hanlon-Blalock procedure may soon become unnecessary in most patients with transposition of the great arteries.

The critical period in the anesthetic management of neonates undergoing

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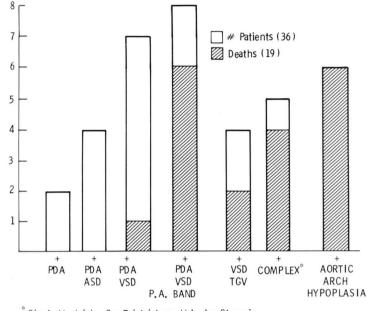
	Age							
	<1 mo		>1-6 mo		>6 mo-1 yr		Total	
Type of procedure	Patients No.	Deaths No.	Patients No.	Deaths No.	Patients No.	Deaths No.	Patients No.	Deaths No.
Blalock-Hanlon	57	20	27	8	3	0	87	28
	32%		30%		0%		31%	
Blalock-Taussig	18	3	18	4	13	0	49	7
	16%		22%		0%		14%	
Waterston	15	9	2	0	()	17	9
	60%		0%				53%	
Systemic PA (Impragraft)	11	3	2	0	0)	13	3
	27%		0%				23%	
Glenn	6	5	22	6	3	0	31	11
	83%		27%		0%		35%	
PA banding	11	9	63	8	13	0	87	17
ů,	82%		13%		0%		20%	
Coarctation	21	12	15	8	5	0	41	20
	57%		53%		0%		49%	
Pulmonary	20	10	8	1	3	0	31	11
valvulotomy	50%		13%		0		35%	
Total	159	71	157	35	40	0	356	106
	45%		27%		0%		30%	

Table 3. Closed palliative cardiac surgery; 1960-1979 (mortality by procedure and age)

Hanlon-Blalock atrial septectomy occurs when the right pulmonary artery and vein are occluded as the transatrial clamp is applied. However, as soon as an adequate septectomy is accomplished, the clamp can be reapplied to the atrial edges, allowing blood to flow through the created defect. The pulmonary vascular occlusive tapes can then be released restoring perfusion to the right lung. Inotropic drugs, calcium chloride, atropine, and sodium bicarbonate are frequently required during this critical period. Three of the neonates who died during operation experienced cardiovascular collapse during clamping or shortly after atrial septectomy.

Procedures that relieve obstruction and decrease ventricular afterload

Coarctation repair may be considered "curative" rather than palliative when performed in patients with no other congenital cardiac defect. We noted no difference in surgical mortality associated with correction of coarctation in neonates as compared with infants from one to 6 months of age. In our series of 36 infants younger than 6 months, only two had an isolated coarctation (patent ductus as the only other associated anomaly) (*Fig. 6*). These two patients and four others who had associated atrial septal defects survived. Of 15 infants younger than 6 months with coarc-



 \degree Single Ventricle, Cor Triatriatum, Valvular Stenosis

Fig. 6. Closed palliative cardiac surgery, 1960–1979; coarctation of aorta (mortality related to associated defects in infants <6 months old). PDA = patent ductus arteriosus, ASD = atrial septal defect, VSD = ventricular septal defect, PA = pulmonary artery, TGV = transposition of great vessels.

tation and a ventricular septal defect, seven died. As noted, six of these had a concomitant pulmonary artery banding. All six neonates with coarctation and aortic hypoplasia died. Four of five neonates with other complex associated anomalies failed to survive. The intraoperative management of these patients with coarctation is greatly influenced by their associated defects²² and their condition upon arrival in the operating room. Since congestive heart failure refractory to medical therapy is the indication for coarctation repair in most neonates, pulmonary congestion and respiratory insufficiency should be anticipated. Many have low cardiac output with metabolic acidosis and impaired renal function.

Arterial pressure should be measured in the right radial artery to avoid errors due to "streaming" in the left subclavian artery and to allow performance of left subclavian aortoplasty. We frequently place a Doppler on a dorsalis pedis artery to determine leg blood pressures intraoperatively and to measure the radial-dorsalis pedis artery pressure gradient after repair. If dorsalis pedis Doppler sounds are completely abolished by aortic cross-clamping, an intraluminal silastic shunt is placed during subclavian aortoplasty. Although paraplegia rarely occurs, the risk may be increased in neonates with low cardiac output or cyanosis or both due to associated defects. Additionally, some advantage may be gained from the shunt by allowing decompression of the proximal aorta, thereby reducing afterload. It may be expected that increased left ventricular afterload resulting from aor-

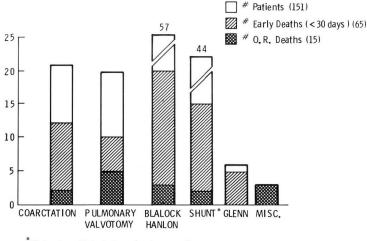
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tic cross-clamping will decrease left ventricular function in patients without ventricular septal defect and increase left-to-right shunting in those with ventricular septal defect. Two patients in our series died during the operation (Fig. 7). One died after division of the patent ductus before coarctectomy, and the other death occurred in a 21-day old, 1757 g (3 pounds, 14 ounces), premature infant with a hypoplastic aortic arch. If the aorta is clamped and a shunt is not used, nitroprusside infusion may reduce proximal aortic hypertension. Heparin, 1 mg/kg, is given before clamping or shunt insertion. Sodium bicarbonate, 1 mEq/kg, is frequently given at the time of reperfusion of the lower half of the body, if indicated by blood gas analysis.

Although closed pulmonary valvulotomy may be considered "corrective", we believe that it is an imprecise, incomplete correction, and that some neonates (if they survive closed valvulotomy) may require a second operation later in life.

Closed pulmonary valvulotomy was performed in 20 neonates; ten died. This is to be compared with a mortality of 9.1% in 11 infants between one month and one year of age. Moreover, five of the ten died in the operating room during or shortly after valvulotomy (Fig. 7). Several factors may have contributed to this high mortality. Closed pulmonary valvulotomy provides inadequate relief of right ventricular obstruction if there is an associated infundibular stenosis or severe valvular dysplasia. Pulmonary outflow tract patch or complete valvular resection is not possible with the closed approach. Ventricular fibrillation during the manipulations necessary to perform closed valvulotomy was the most common precipitating cause of intraoperative death. Postoperative deaths were most commonly due to severe infundibular obstruction or spasm, the "suicidal right ventricle" phenomenon. In view of the high mortality associated with closed pulmonary valvulotomy in neonates, we no longer



^{*}Waterston, Blalock-Taussig, Impragraft

Fig. 7. Closed palliative cardiac surgery, 1960–1979; early and operative deaths in infants <30 days old. Misc = miscellaneous.

perform this procedure. All neonates and infants younger than 6 months with pulmonary valvular stenosis undergo repair utilizing cardiopulmonary bypass and deep hypothermic circulatory arrest.

The importance of the intraoperative management of neonates during closed palliative cardiac surgery is accentuated by the fact that 16 of 65 died in the operating room and all 16 were neonates (Fig. 7). Although infants who are moribund preoperatively may not survive, there are specific, critical periods during the operation when the risk is highest. In our series the time of highest risk of intraoperative death was during pulmonary artery occlusion for systemicpulmonary shunts, atrial clamping and pulmonary vascular occlusion for Blalock-Hanlon procedures. maximal banding of the pulmonary artery, during or shortly after pulmonary valvulotomy, and during aortic cross-clamping for coarctation. These critical periods should be anticipated by both surgeon and anesthesiologist. The surgeon must complete all preparatory steps in order to work expeditiously during these periods. The anesthesiologist must have all bolus drugs prepared and assure that conditions are optimal prior to occlusion. Immediately after release of the clamps and restoration of normal blood flow, there may be washout acidosis, which requires administration of sodium bicarbonate. Vigorous ventilation of the lungs may increase oxygen availability and restore paCO₂ to normal levels.

Summary

We have noted a higher mortality associated with palliative cardiac surgery in neonates compared with the mortality in older infants. The differentiation of neonates from infants at 30

days is an arbitrary dividing point. Most deaths in infants one month to 6 months of age occurred in infants older than one month but younger than 3 months. Operating room deaths occurred exclusively in neonates, most of whom were moribund preoperatively. The neonatal mortality associated with pulmonary banding, closed pulmonary valvulotomy, and the Glenn procedure appears to be prohibitive. The advances in surgical techniques and the increased survival in corrective operations have led us to abandon closed palliative operations for many entities. However, we prefer systemic-pulmonary artery shunts and later total correction for palliation of infants with tetralogy of Fallot. Palliative operations will continue to be the only hope for survival for infants with lesions that are noncorrectable in infancy, such as univentricular heart, double outlet right ventricle, and tricuspid atresia.

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