Cardiovascular Surgery in Newborn Infants:
Results in 1,050 Patients Less than One Year Old

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THE highest mortality from congenital heart disease occurs during the first year of life. More than one fourth of infants born alive with anomalies of the heart and great vessels die before they reach one month of age, and 60 per cent die before one year if not operated upon.[23, 24] A variety of anomalies are capable of causing heart failure and hypoxemia, the physiologic derangements that constitute the main indications for operation. Medical therapy is employed initially but often provides inadequate relief. In these cases survival depends on surgical alteration of cardiac anatomy and function, and operation must frequently be performed on an urgent or emergency basis.[3-5, 7-9]

Clinical Material

Between January, 1954, and October, 1970, 1,050 infants less than one year of age underwent palliative or corrective surgical procedures for congenital cardiovascular anomalies. Sixty per cent were 3 months old or less, and 80 per cent were 6 months old or less (Table 1).

The main indications for operation were refractory heart failure (as in patients with left-to-right shunts or valvular obstructions), hypoxemia (as in patients with decreased pulmonary blood flow and right-to-left shunts), and tracheoesophageal compression (due to vascular ring anomalies). When symptoms were not relieved by medical therapy with oxygen, digitalis, and diuretics, palliative or curative operation was scheduled, frequently as an emergency. In many cases the patient's critical condition precluded cardiac catheterization and angiocardiography, so operation was performed on the basis of clinical diagnosis alone.

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Results

Of the 1,050 patients in this series who underwent palliative or curative operations for congenital heart disease, 291 (28 per cent) died (Table 2). Postoperative improvement in the 759 (72 per cent) survivors was gratifying, with significant relief of cardiac failure or hypoxemia. Since total correction of certain complex anomalies during the newborn period is associated with formidable mortality, these infants were treated in two stages; a palliative operation was performed during the first year of life and total correction deferred until later years when open-heart technics could be applied with greater safety.

Anomalies Treated in One Stage

Total correction was accomplished in one stage in 397 patients, most of whom had anomalies correctable by closed technics, although some required cardiopulmonary bypass or caval inflow occlusion. Mortality for this group was 21 per cent (Table 2). One hundred and ninety patients had tricuspid atresia or miscellaneous anomalies that could not be surgically corrected and therefore were treated by one-stage palliative operations. Ninety-three of these patients died after operation.

TABLE 1. *Age Distribution in 1,050 Infants Undergoing Cardiovascular Surgery*

Age (Months)	No. Patients	Per Cent
0–3	627	60%
4–6	202	20%
7–9	125	11%
10–12	96	9%
	1,050	100%

Patent Ductus Arteriosus. Most of these patients had large ductus and low pulmonary vascular resistance causing a high-volume left-to-right shunt that produced refractory cardiac failure. Through a left posterolateral fourth interspace incision the ductus was interrupted either by suture-ligature (usually sufficient in small infants) or division and suture (sometimes necessary in larger infants). Of the 168 patients with this lesion, 13 (8 per cent) died, usually as a result of associated, more complex cardiac anomalies.

Coarctation of the Aorta. Indication for operation in the 98 patients with coarctation of the aorta was cardiac failure. The coarctation was resected and end-to-end anastomosis performed through a left posterolateral incision in the fourth interspace. Interrupted sutures were used in many patients on the accessible ventral surface of the anastomosis to favor subsequent growth. An associated patent ductus, when present, was closed as the coarctation was resected. If an associated ventricular septal defect had been diagnosed by catheterization and the pulmonary artery remained tense after repair of the coarctation, a constricting band was placed around the pulmonary artery. Most of the 22 (22 per cent) patients who died had preductal coarctation or associated intracardiac lesions, or both.

Aortic Stenosis. Thirty-two patients, mostly in the first few weeks of life, had aortic stenosis producing severe heart failure that responded poorly to medical therapy. Cardiopulmonary bypass enabled aortic valvotomy under direct vision. Ten (31 per cent) of these patients did not survive operation.

Pulmonary Stenosis. Pulmonary stenosis with intact ventricular septum produced severe heart failure requiring pulmonary valvotomy in 30 patients. Both closed transventricular valvotomy and open operation with caval inflow occlusion have been used in the past, but the preferred technic in recent years has been open valvotomy with cardiopulmonary bypass. Most of the nine (30 per cent) patients who died after operation had a small right ventricle.

Vascular Ring. Compression of the trachea and esophagus by anomalies of the aortic arch system (such as double aortic arch, right arch with left ligamentum arteriosum, and retroesophageal right subclavian artery) caused symptoms severe enough to warrant operation during the first year of life in 18 patients. A left posterolateral incision was used and the offending vessel was divided. Vessels were removed from the retroesophageal area when feasible, and the ductus or ligamentum was divided. The esophagus and trachea were dissected to release any constricting fascial bands.[15, 16] In most patients the anomaly causing compression was a double aortic arch. All 18 patients with vascular ring survived operation.

Total Anomalous Pulmonary Venous Drainage. Fifty-one patients with cardiac failure required operation for total anomalous pulmonary venous drainage. Supracardiac, cardiac, infracardiac and mixed types were encountered and all required cardiopulmonary bypass. The common pulmonary vein was anastomosed to the left atrium, the patent foramen ovale or atrial septal defect was closed, and the connection between the pulmonary vein and systemic venous system was ligated.[10, 12, 22] Twenty-nine patients (57 per cent) died after operation.

Tricuspid Atresia. Tricuspid atresia was surgically treated in 66 newborn patients.

TABLE 2. *Results of Cardiovascular Surgery in 1,050 Newborn Infants*

Anomalies Treated in One Stage	No. Pts.	No. Deaths	Mortality
Patent ductus arteriosus	168	13	8%
Coarctation of aorta	98	22	22%
Aortic stenosis (corrective)	32	10	31%
Pulmonary stenosis	30	9	30%
Vascular ring	18	0	0
Total anomalous pulmonary return	51	29	57%
Subtotal	397	83	21%
Tricuspid atresia (palliative)	66	13	20%
Miscellaneous anomalies	124	80	65%
Subtotal	190	93	49%
Anomalies Treated in Two Stages			
Tetralogy of Fallot	144	24	17%
Transposition of great vessels	171	43	25%
Ventricular septal defect	116	33	28%
Pulmonary valve atresia	32	15	47%
Subtotal	463	115	25%
Total	1050	291	28%

Most had decreased pulmonary flow with severe hypoxemia and underwent systemic-pulmonary shunt for palliation.[19] The few patients who had increased pulmonary flow due to associated ventricular septal defect or transposition underwent pulmonary artery banding. Occasional use of cavopulmonary shunts yielded discouraging results. Thirteen (20 per cent) of the 66 patients with this lesion died after operation.

Miscellaneous Anomalies. Complex, often incurable anomalies such as truncus arteriosus, atrioventricularis communis, single ventricle, and endocardial fibroelastosis were found in 124 patients referred for operation during the first year of life. Surgical efforts in these patients were generally palliative. In those with hypoxemia, systemic-pulmonary artery shunts were used to increase pulmonary blood flow. Atrial septal defects were created to promote intracardiac mixing in patients with transposition of the great vessels, and pulmonary artery banding was employed where increased pulmonary blood flow was producing heart failure. In a few cases clinical or laboratory diagnosis was doubtful and exploratory thoracotomy was performed as a a final, desperate effort. Even palliative procedures were not possible in many of these patients because of unfavorable anatomy. Mortality for this heterogenous group of 124 patient was 65 per cent (80 deaths).

Anomalies Treated in Two Stages

Four hundred and sixty-three patients had major anomalies that would require cardiopulmonary bypass for total correction (Table 2). To avoid the excessive risk associated with open heart procedures in the newborn period, we utilized a two-stage approach, with a closed palliative operation performed first, deferring total correction until later in life.

Tetralogy of Fallot. Severe hypoxemia was the indication for operation in 144 patients with tetralogy of Fallot. Since an effective and safe palliative procedure is

available for this lesion, total correction in these patients was deferred to an older age, and all underwent systemic-pulmonary artery shunts. We used the Blalock [2] and Potts [26] technics in earlier cases, but have recently preferred an intrapericardial anastomosis between the posterior aspect of the ascending aorta and the subjacent right pulmonary artery.[6, 20] Mortality in this group of patients was 17 per cent (24 deaths).

Transposition of the Great Vessels. Patients with transposition of the great vessels, the most common cyanotic anomaly requiring operation during infancy, exhibited severe hypoxemia with or without congestive failure. Here again, a first-stage, palliative procedure was chosen to avoid the risk of open-heart repair during the neonatal period. We used a modified version of the Blalock-Hanlon [1] technique that improved intracardiac mixing by creation of an atrial septal defect.[11, 17, 18] However, since 1967 most infants requiring intervention for transposition have been treated by balloon atrial septostomy, as described by Rashkind,[27] obviating the need for the Blalock-Hanlon operation. In patients with an associated ventricular septal defect causing increased pulmonary flow and pressure, the pulmonary artery was banded after the atrial septum was excised or balloon atrial septostomy performed. Some infants had pulmonary stenosis and also received a systemic-pulmonary artery anastomosis. Of the 171 patients who underwent operations for transposition, 43 (25 per cent) died.

Ventricular Septal Defect. Intractable heart failure caused by a ventricular septal defect warranted operation in 116 patients. Open-heart repair was used between 1956 and 1959 in 31 of these patients, and 13 (42 per cent) died. This high mortality prompted us to adopt a staged approach. Since 1959, 85 patients have undergone pulmonary artery banding with a mortality of 23 per cent. Mortality for the entire group of 116 patients with ventricular septal defect was 28 per cent (33 deaths).

Pulmonary Valve Atresia. Hypoxemia was extreme in the 32 patients with pulmonary valve atresia. Systemic-pulmonary artery shunts were used in most of this group, since, except in rare instances, the usual small size of the right ventricle and pulmonary annulus made valvotomy impossible or impractical. Special care was taken to construct the anastomoses small enough to lessen the chance of cardiac failure. Fifteen patients (47 per cent) died after operation.

Discussion

Rapid advances in surgical treatment of congenital heart disease during the last few decades have enabled correction or palliation of many cardiovascular malformations previously considered incurable. Accurate clinical diagnosis is more important today than ever before, since we now have an array of effective surgical procedures that, if performed promptly and appropriately in the newborn child, may save lives otherwise doomed at infancy. Moreover, improvement in technics of general anesthesia for small infants has lowered significantly the risk of thoracotomy in this age group.[28]

The poor prognosis in general for infants with congenital heart disease not surgically treated holds true for individual lesions as well. Seventy-five per cent of infants showing symptoms of coarctation of the aorta die before they are one year old.[25] An average survival of only 5½ months has been reported for a group of unoperated newborn patients with transposition of the great vessels.[1] Mortality during the first year of life for patients with tricuspid atresia is 66 per cent without operation; for tetralogy of Fallot it is 55 per cent.[21] All patients with pulmonary atresia and intact ventricular septum, and 80 per cent of those with total anomalous pulmonary ve-

nous drainage die during the first few months of life if not operated upon.[13, 14]

If initial medical therapy is successful in patients with severe symptoms of congenital heart disease, operation may be deferred until an older, more favorable age is attained. But when medical treatment is ineffective in ameliorating these symptoms, as it so often is, operation should be performed without delay, and on a clinical basis alone if the patient is too ill to tolerate cardiac catheterization. Although hypoxemia and cardiac failure are the principal indications for neonatal surgery, experience has shown that other findings may signify the need for operation; for instance, syncope, increasing cardiomegaly, electrocardiographic evidence of left ventricular strain, or discovery on cardiac catheterization of significant gradients, shunts, or pulmonary hypertension.[9]

The best surgical procedure for an infant with congenital heart disease is that which will offer the most physiologic benefit with the least operative risk. Lesions such as patent ductus arteriosus or coarctation of the aorta can be safely corrected with closed technics. Total correction of anomalies such as tetralogy of Fallot or ventricular septal defect, on the other hand, requires extracorporeal circulation, an added risk in neonates. These anomalies are best treated in two stages using a palliative operation first (when medical therapy is not successful) and doing the corrective procedure later in life. Usually the palliative operation will salvage the patient until a more favorable age and size for open-heart surgery is reached. For some lesions such as total anomalous pulmonary venous drainage no consistently effective palliative procedure exists, and complete correction with cardiopulmonary bypass must be attempted. Other complex anomalies such as tricuspid atresia are not surgically correctable and palliative measures must suffice.

Most deaths in this series of 1,050 patients occurred within 12 to 24 hours after operation; yet patients who survived usually did so with, and because of, dramatic improvement in hypoxemia and cardiac failure. This suggests that surgical intervention in these seriously ill infants must be definitive and provide major physiologic benefit if the patient is to survive more than a few hours.

This series included many patients who were extremely poor operative risks and many desperate cases of exploratory operations performed on the remote chance that a correctable or palliable lesion might be found. Had selection of patients been limited to those with lesions susceptible to correction or palliation, survival rate would have been greater. In view of the high mortality during the first year of life in unoperated infants with cardiovascular disease, we believe that the 72 per cent survival reported in this paper justifies aggressive surgical treatment in the newborn period when symptoms do not respond to medical therapy.

Summary

Maximum mortality in patients with cardiovascular anomalies occurs during the first year of life when 60 per cent die unless operated upon. A variety of anomalies can cause hypoxemia and/or cardiac failure and when symptoms do not respond to medical therapy, operation must be performed to prevent fatal outcome.

Between 1954 and 1970, 1,050 infants underwent non-elective operations during the first year of life for major cardiovascular anomalies including patent ductus, coarctation, aortic stenosis, pulmonary stenosis, vascular ring, total anomalous pulmonary venous drainage, tricuspid atresia, tetralogy of Fallot, transposition of the great vessels, ventricular septal defect, pulmonary valve atresia, and miscellaneous anomalies. When feasible, corrective operations were performed. In patients with complex anomalies such as transposition of the great vessels a palliative operation was chosen, deferring

total correction until a more favorable age for open-heart surgery was reached. Survival of 72 per cent in this unselected series demonstrates the value of surgical treatment in patients with cardiovascular anomalies producing intractable symptoms during the critical first year of life.

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