Cardiac Surgery in Infants
Less than Six Weeks of Age

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SUMMARY
Sixty-one critically ill infants with congenital heart disease had operative treatment within 6 weeks of birth. Anatomic and physiologic diagnoses were established preoperatively by cardiac catheterization and angiography in all. A variety of palliative or corrective operations were performed to relieve specific hemodynamic burdens. Thirty-seven (61%) survived hospitalization, but 18 had one or more postoperative complications. At the present time, 29 (48%) are alive 9-45 months after operation. Although operative mortality and morbidity rates are high, none of these patients was expected to survive early infancy without operation.

EIGHT TO 10 babies per 1000 live births have congenital heart disease\(^1\)\(^2\) and without treatment about 30-40% of them die in childhood.\(^1\)\(^2\) Most of these deaths occur very early: 14-22% of children born with congenital heart disease die within the first week after birth\(^1\)\(^3\) and 19-27% are dead by 1 month of age.\(^1\)\(^3\) In general, infants who succumb in early infancy have complex cardiac malformations or simple cardiac anomalies associated with severe circulatory dysfunction or serious disease of other organs; therefore, they are not able to adapt their fetal circulation to that of infancy. Since many children with congenital heart disease die in infancy, meaningful improvement in therapy requires improved results in infants who cannot be managed successfully by nonoperative means during the first few days and weeks of extrauterine life.

During the 3 calendar years 1968-70, 163 infants less than 2 years of age had cardiac operations at the University of California, San Francisco, and 74% of these patients survived hospitalization. Of the 163 infants, 61 were 6 weeks or less in age at the time of operation, and none was expected to survive early infancy without operation. This report summarizes our experience with these 61 infants and reports follow-up data in all except one patient as of September 1971 (9-45 months after operation).

Clinical Material
The numbers of each operation, and the hospital (within 30 days) and late deaths are presented in table 1. The results for each year are presented in table 2.

Interruption of Patent Ductus Arteriosus
Symptoms and signs of severe congestive heart failure with or without associated respiratory disease prompted interruption of a patent ductus arteriosus in 13 infants. Eight had birth weights less than 1650 g, and four of these infants had associated idiopathic respiratory distress of the newborn. Eleven were cyanotic, 12 had apneic spells, eight had episodic bradycardia, and one had three cardiac arrests. A systolic murmur was
heard over the left upper chest at 4–21 days of age in all. At preoperative cardiac catheterization a patent ductus arteriosus was the major cardiac anomaly in 10, and three patients had additional lesions: transposition of the great vessels with ventricular septal defect, pulmonary valvular stenosis, and double-outlet right ventricle (Taus-sig-Bing anomaly).

A large ductus arteriosus (in relation to the aortic arch) was ligated in seven patients and divided in six. Seven patients weighed less than 1500 g at operation (900–1480 g). The ductus tore during ligation in one and was difficult to oversew because of friable tissue in three. In addition to ligation of the ductus arteriosus, the main pulmonary artery was banded in the infant with double-outlet right ventricle.

One premature infant with severe respiratory distress died of progressive pulmonary disease 24 hours after operation. The remaining 21 infants survived, and in seven the postoperative course was not complicated (table 3).

The infant with transposition of the great vessels died of pneumonia at 3.5 months of age. The child with double-outlet right ventricle (now 21 months) had chronic congestive heart failure. One premature infant developed Dandy Walker syndrome and is retarded. Another premature infant has a mild hemiplegia. The remaining eight infants are well, have satisfactory somatic growth, and apparently normal mental development for age.

**Systemic-Pulmonary Arterial Anastomoses**

Fifteen infants who were severely cyanotic (arterial pO₂ 11–35 mm Hg) and three infants who had hypoplasia of the left heart had systemic-pulmonary arterial anastomoses. (Ten of these infants have been reported previously.4) Four patients had episodes of bradycardia preoperatively, and two infants with lesions of the left heart had congestive heart failure. All infants were acidic preoperatively, and 14 required repeated doses of sodium bicarbonate and tromethamine (THAM-E) for progressive acidosis (arterial pH 6.95–7.28). Diagnoses found at cardiac catheterization are listed in table 4. The two patients with complex lesions had pulmonary atresia with double aortic arch and additional somatic anomalies.

Two of the three patients with hypoplasia of the left heart had Waterston aortic-right pulmonary arterial anastomoses, and one had a Potts anastomosis. All died during (one patient) or shortly after (two patients) operation, and at the present time we do not operate for hypoplasia of the left heart.

Fourteen patients with inadequate pulmonary arterial blood flow had Waterston anastomoses, and one had a Potts anastomosis. Four had balloon septostomy during preoperative catheterization; now this procedure is performed in all infants with pulmonary or tricuspid atresia who do not have large intraatrial communications. The two patients with complex lesions, and one infant with tricuspid atresia (Potts anastomosis) died during (two patients) or shortly after (one patient) operation. One patient with pulmonary atresia died postoperatively with an occluded anastomosis, and another died 6 weeks after operation following an attempt to band both pulmonary arteries because of an excessive shunt.
Table 3

Postoperative Complications in Hospital Survivors

<table>
<thead>
<tr>
<th>Operation</th>
<th>No. pt</th>
<th>None</th>
<th>Atelectasis</th>
<th>Pneumonia</th>
<th>Pneumothorax</th>
<th>Chylothorax</th>
<th>Acute obstruction endotracheal tube</th>
<th>CHF</th>
<th>Severe arrhythmias</th>
<th>Persistent acidosis</th>
<th>Bleeding reoperation</th>
<th>Temp oliguria</th>
<th>Paroxysmal hypertension</th>
<th>Wound</th>
<th>Temp seizures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Interruption of patent duct</td>
<td>12</td>
<td>7</td>
<td>2</td>
<td>1</td>
<td></td>
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<tr>
<td>ductus arteriosus</td>
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</tr>
<tr>
<td>Systemic-pulmonary arterial</td>
<td>10</td>
<td>4</td>
<td>4</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
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<td>1</td>
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<tr>
<td>shunt</td>
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<tr>
<td>Resection coarctation</td>
<td>6</td>
<td>4</td>
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<td></td>
<td>2</td>
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<td></td>
<td></td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Blalock-Park anastomosis</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td></td>
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<td></td>
<td></td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Blalock-Hanlon atrial septostomy</td>
<td>4</td>
<td>2</td>
<td>1</td>
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<td>1</td>
<td>1</td>
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<tr>
<td>Pulmonary valvulotomy</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td></td>
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<tr>
<td>Pulmonary arterial banding</td>
<td>1</td>
<td></td>
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</tr>
<tr>
<td>Totals</td>
<td>37</td>
<td>19</td>
<td>8</td>
<td>3</td>
<td>4</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>

*Some patients had more than one complication; each complication in surviving patients is listed.
Four of 10 hospital survivors did not have postoperative complications (table 3).

Four patients died 6 weeks to 8 months after operation. One infant died of chronic congestive heart failure, and two died of respiratory infections. One died unexpectedly at home. The six surviving patients are generally well but are cyanotic and tire easily. All have apparently normal mental development but are below the third percentile in physical growth. Four have had one or more episodes of pneumonia. Three have been recatheterized at 18–27 months of age, and in two of them systemic pressures are present in the pulmonary circulation.

**Resection of Aortic Coarctation**

Nine patients had resection of coarctation of the aorta for severe congestive heart failure unresponsive to medical management. At cardiac catheterization one had tubular hypoplasia of the aortic arch, two had small ductus arteriosus, and three had ventricular septal defects. The coarctation was resected with division or ligation of the ligamentum or ductus arteriosus in all.

Three patients died, one of bradycardia and arrest during operation when the aorta was occluded. One had postoperative oliguria (aorta clamped 19 min) but died of acute obstruction of the endotracheal tube 2 days after operation. A third infant died of pneumonia 14 days postoperatively.

Two other infants survived postoperative complications (table 3). One developed paroxysmal hypertension (blood pressure over 300 mm Hg) the night of operation. Another had postoperative bleeding and pneumonia.

Two survivors developed recurrent coarctations which were first apparent 2 and 3 months after operation, and were successfully resected at 11 weeks and 2.5 years of age, respectively. One patient was lost to follow-up 1 year after operation but did not have residual cardiac disease at that time. The remaining five patients have normal growth and development; the ventricular septal defect has closed in one patient.

**Blalock-Park Subclavian-Aortic Anastomosis**

Seven infants had anastomosis of the left subclavian artery to the ascending or descending aorta. Three infants had interruption of the aortic arch distal to the left subclavian (type 1), and two had interruption of the aortic arch between the left carotid and subclavian arteries (type 2). Two infants had severe coarctation: in one, this was associated with a Taussig-Bing anomaly, and in the other with aortic atresia and hypoplasia of the left ventricle and ascending aorta. All infants had severe congestive heart failure, and five required operation during the first week after birth. In all, femoral pulses were weak or absent, and a large ductus arteriosus was the principal or exclusive source of blood to the lower body.

The operation was inappropriate in the patient with aortic atresia since this feature was not recognized until after angiograms were developed and emergency operation was completed. The left subclavian artery was anastomosed to the descending aorta in the three infants with type 1 arch interruption and in the infant with Taussig-Bing anomaly. In the patients with type 2 arch interruption, the left subclavian artery was anastomosed to the ascending aorta in one and to the left carotid in the other. The large ductus arteriosus was divided after the anastomosis was completed in three infants. The main pulmonary artery was banded in two patients, and both left and right pulmonary arteries were banded in two others.

The infant with Taussig-Bing anomaly died of hemorrhage into the lungs when the ductus arteriosus was divided after the subclavian-aortic

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**Table 4**

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Pt</th>
<th>Hospital deaths</th>
<th>Late deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary atresia</td>
<td>10</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Intact ventricular septum (7)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hypoplasia of pulmonary ventricle (7)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transposition of the great arteries (2)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Complex lesions</td>
<td>2</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Aortic or mitral atresia</td>
<td>3</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>with hypoplasia of left ventricle</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>and ascending aorta</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Totals</td>
<td>18</td>
<td>8</td>
<td>4</td>
</tr>
</tbody>
</table>

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*Circulation, Volume XLVI, August 1972*
anastomosis. Three infants, including the baby with aortic atresia, died of progressive congestive heart failure during the first postoperative day.

One infant with type 1 aortic arch interruption, corrected transposition of the great vessels, and bilateral pulmonary arterial bands died 3 months after operation of atelectasis and pneumonia caused by dilated pulmonary vessels. One infant with type 1 arch interruption and associated transposition of the great arteries with ventricular septal defect developed mild left hemiparesis 15 months after operation, but is otherwise well at age 21 months. (Details of this case have been reported previously.\textsuperscript{5}) The infant with type 2 arch interruption and ventricular septal defect is alive and well at 14 months of age.

**Blakow-Hanlon Atrial Septostomy**

Eight infants with transposition of the great vessels had an atrial septostomy before 6 weeks of age. All were deeply cyanotic and all required therapy for metabolic acidosis before cardiac catheterization. Two infants also had congestive heart failure and respiratory difficulty. A small ventricular septal defect was present in three patients. A Rashkind balloon septostomy was performed in five, but did not sufficiently improve atrial mixing and systemic arterial oxygen saturation in any. The main pulmonary artery was banded after completion of the atrial septostomy in two patients who had large ventricular septal defects.

Four patients, all of whom were operated upon in 1968, died during initial hospitalization. One infant died of inferior vena caval thrombosis 7 months after operation. Two infants who had pulmonary arterial bands are well and are developing satisfactorily 10 and 25 months after operation. One patient had successful insertion of an intraatrial baffle at 42 months of age.

**Pulmonary Valvotomy**

Two infants with pulmonary atresia, intact ventricular septum, and apparently adequate right ventricular development had pulmonary valvotomy during inflow occlusion for severe cyanosis and acidosis. One infant died of progressive hypoxia from right ventricular insufficiency during the first postoperative day. The other patient, who had bilateral pneumothorax immediately after operation, is well without medication at 10 months of age.

**Pulmonary Arterial Banding**

One infant with type 1 truncus arteriosus, absent right pulmonary artery, and hypoplasia of the right lung had banding of the left pulmonary artery as the only procedure performed. One infant had attempted banding to control an excessive Waterston aortic-pulmonary shunt. In seven other patients (table 5) pulmonary arterial banding was performed in addition to other operative procedures.

The patient with the excessive Waterston shunt and a patient with type 1 interrupted aortic arch died shortly after operation. Another patient with type 1 arch interruption and the patient with hemitruncus died 3 and 2 months after operation. The five surviving patients are well, have apparently normal development, but are below the third percentile in physical growth.

**Cardiopulmonary Bypass**

Three infants had cardiopulmonary bypass, two for relief of severe aortic stenosis, and one for correction of total anomalous pulmonary venous connection. Neither infant with aortic stenosis could be resuscitated after bypass. The infant with total anomalous pulmonary venous connection died 6 weeks after operation of acute tubular necrosis.

### Table 5

**Pulmonary Arterial Banding in Infants Less than 6 Weeks of Age**

<table>
<thead>
<tr>
<th>No. pt</th>
<th>Banded pulmonary artery</th>
<th>Associated operation</th>
<th>Diagnosis</th>
<th>Hospital deaths</th>
<th>Late deaths</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 left</td>
<td>Left</td>
<td>None</td>
<td>Hemitruncus arteriosus, absent right pulmonary artery</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>1 main</td>
<td>Main</td>
<td>Division of ductus arteriosus</td>
<td>Taussig-Bing anomaly</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Main</td>
<td>Blalock-Park</td>
<td>Interrupted aortic arch</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Blalock-Hanlon</td>
<td>Transposition of great vessels with ventricular septal defect</td>
<td>0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>Right and left</td>
<td>Waterston</td>
<td>Pulmonary atresia</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Right and left</td>
<td>Blalock-Park</td>
<td>Interrupted aortic arch</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>2</td>
<td>2</td>
</tr>
</tbody>
</table>

*Circulation, Volume XLVI, August 1972*
Discussion

In early infancy, mortality rates of different congenital heart lesions vary, and reliable figures, particularly for complex anomalies, are difficult to obtain. Nearly all deaths from patent ductus arteriosus, 87% from coarctation of the aorta, and 64% from ventricular septal defect occur before 1 month of age. Forty percent of deaths from transposition of the great arteries occur within 1 month, and approximately 95% die within 1 year of birth. More than half the babies born with pulmonary or tricuspid atresia and 75% of infants with interrupted aortic arch die before 1 month of age.

In our judgment, none of the patients in this series would have survived early infancy without operation. Although we operated upon a few patients with uncorrectable lesions (e.g., aortic atresia with hypoplasia of the ascending aorta, pulmonary atresia with associated cardiac and somatic anomalies), other lesions that were previously thought to be inoperable in newborns (type II interrupted aortic arch, patent ductus arteriosus in premature with respiratory distress, Taussig-Bing anomaly, and truncus arteriosus) proved to be operable. Because neonates with severe circulatory dysfunction have little hope, we operated upon all in whom a specific operative procedure might theoretically improve the hemodynamic and/or respiratory condition. The high incidence of hospital complications and deaths reflects this positive philosophy and the critical condition of most patients.

Several features in the management of these infants deserve emphasis. All required specially trained nurses, frequent examination by physicians, precise drug and fluid dosages, and continuous cardiac, respiratory, and temperature monitors. Accurate anatomic and physiologic diagnoses were essential, and cardiac catheterization and angiography always preceded operation. During operation arterial (umbilical or radial) and venous catheters permitted accurate measurement of blood pressure and blood gases (PaO₂, PaCO₂, and pH) and immediate infusion of required blood, fluids, or drugs. Postoperative pulmonary problems occurred frequently but were satisfactorily managed as outlined previously.

The present series is consecutive, includes all operations that we performed for congenital heart disease in newborns, includes follow-up data, and excludes the better results that are usually obtained in older infants. A comparable series has not been published, but some reports include data of operations in newborns or report small numbers of newborns that have had specific procedures. In this series, we applied to newborns both palliative and corrective operations that are well established for older children. Experience reduced hospital mortality, but when palliative operations were applied to more complex lesions late mortality rose (table 2). Our hospital results compare favorably with those published by others for newborns, and the data principally evaluate the contribution of procedures that exclude cardiopulmonary bypass to the treatment of critically ill newborns.

Twenty-nine of the 61 patients are presently alive, and all except one have apparently normal development. The majority of the survivors had extracardiac lesions (patent ductus arteriosus, coarctation) which were completely corrected by operation. Some patients with operable intracardiac lesions had good results from palliative procedures, but others developed irreversible pulmonary vascular disease or died of early or late postoperative complications. In these patients, the contribution of palliative operations must be questioned since corrective operations can now be applied to newborns.

Although only palliative operations are available for some congenital heart lesions, total correction of operable intracardiac defects in the neonatal period may further improve results in newborns that cannot be managed by nonoperative means.

Acknowledgment

The authors are indebted to the surgical and pediatric house officers and to the nurses who participated in the care of these infants and to Miss Beverly Hill who helped assemble the data.
References


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Circulation. 1972;46:250-256
doi: 10.1161/01.CIR.46.2.250
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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