Hypoplastic Left Heart Syndrome: Experience With Palliative Surgery

WILLIAM I. NORWOOD, MD, FACC*
JAMES K. KIRKLIN, MD*
STEPHEN P. SANDERS, MD†

Boston, Massachusetts

Aortic atresia is a form of hypoplastic left heart syndrome always complicated by associated severe hypoplasia of the ascending aorta and various degrees of mitral valve and left ventricular hypoplasia. At present it is a universally fatal lesion in early infancy. This is a report of a new palliative procedure for hypoplastic left heart syndrome that has resulted in early ongoing survival of two infants with aortic atresia. On the basis of experience with a third patient, an operation for future physiologic correction is proposed.

Prolonged survival after either palliative or reparative surgery for hypoplastic left heart syndrome has not been reported. The anatomic complexity of this cardiac malformation has been considered prohibitive of long survival, with or without surgery, and generally only supportive therapy has been recommended. However, persistent effort to improve the long-term outlook of neonates with hypoplastic left heart syndrome is to be encouraged for at least three reasons: (1) It is the fourth most common congenital cardiac anomaly entering the New England Regional Infant Cardiac Program (after ventricular septal defect, tetralogy of Fallot and transposition of the great arteries) and accounts for nearly 25 percent of cardiac deaths during the 1st week of life and 15 percent of cardiac deaths during the 1st month of life.[1] (2) Unlike many forms of congenital cardiac disease, the syndrome has a very low incidence rate of associated extracardiac anomalies.[1,2] (3) In principle, hypoplastic left heart syndrome is similar to tricuspid atresia and other forms of univentricular heart in which the outlook after surgical intervention has recently improved dramatically.[3,4] This report constitutes our experience with the development of effective palliation for infants with hypoplastic left heart syndrome.

Case Reports

Case 1: This infant, a 2.9 kg product of a full-term, uncomplicated pregnancy, was noted to have cyanosis, tachypnea, hepatomegaly and cardiac enlargement shortly after birth. Cardiac catheterization revealed a hypoplastic left ventricle, aortic atresia and a large patent ductus arteriosus (Fig. 1). Arterial P_{O_2} (partial pressure of oxygen) on 100 percent oxygen was 24 to 33 torr with normal partial pressure of carbon dioxide (P_{CO_2}) and pH. The patient was transferred to an outside hospital but returned 1 month later.

Physical examination revealed a cyanotic infant weighing 3.39 kg. The heart rate was 152 beats/min and the respiratory rate 58/min. Cardiac examination revealed a quiet precordium, single second heart sound and a grade 2/6 systolic murmur at the apex. The electrocardiogram showed an axis of +120° and right ventricular hypertrophy with "strain."

The following operative procedure was performed: November 8, 1977, when the infant was 5 weeks old, with use of deep hypothermia and circulatory arrest, (1) enlargement of the atrial septal defect, (2) a modified Glenn shunt (end to side right pulmonary artery to superior vena cava anastomosis), (3) ligation of

From the Departments of Cardiovascular Surgery* and Cardiology†, Children's Hospital Medical Center and Departments of Surgery and Pediatrics, Harvard Medical School, Boston, Massachusetts. Manuscript received June 25, 1979, accepted July 25, 1979.

Address for reprints: William I. Norwood, MD, Children's Hospital Medical Center, 300 Longwood Avenue, Boston, Massachusetts 02115.

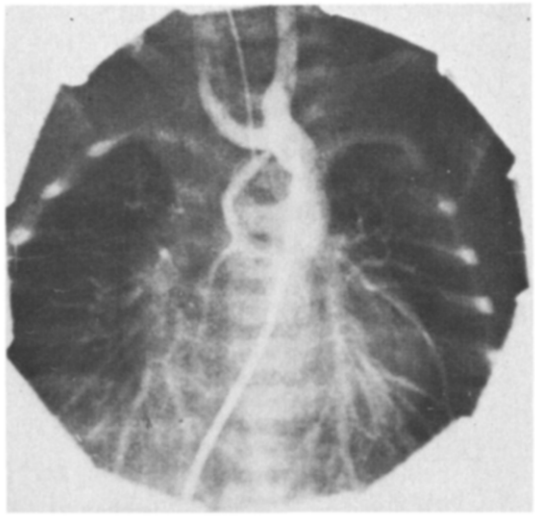

FIGURE 1. Case 1. Frame from anterior-posterior cineangiogram with injection into the main pulmonary artery. Retrograde filling of the diminutive ascending aorta is characteristic of aortic atresia.

the patent ductus arterious, (4) left pulmonary arterial banding, and (5) side to side anastomosis of the aorta and main pulmonary artery using the right pulmonary artery to extend the angioplasty. During the early postoperative period, left atrial and right atrial pressures were 9 to 10 mm Hg and arterial pressures were 80 to 90 mm Hg with normal sinus rhythm and a heart rate of 130 beats/min. Marked arterial desaturation persisted, however, and the patient died with progressive acidosis and hyperkalemia 7 hours after operation.

Case 2: This infant, a 2.9 kg product of a full-term uncomplicated pregnancy, was referred on January 19, 1979 at age 36 hours because of severe cyanosis and acidosis. Physical examination revealed a cyanotic infant with a pulse rate of 160 beats/min, respiratory rate of 80 to 100/min and a systolic blood pressure in all limbs of 50 mm Hg. There were diffuse rales bilaterally. The chest roentgenogram showed cardiac enlargement with moderate pulmonary vascular congestion. The electrocardiogram revealed an axis of +120°, right atrial enlargement and right ventricular hypertrophy.

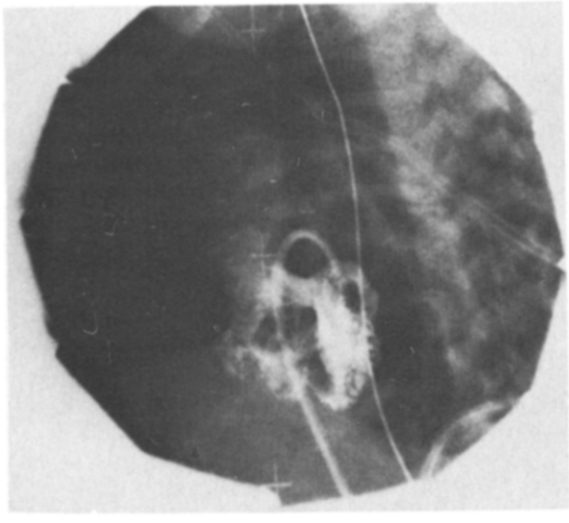

FIGURE 2. Case 2. Frame from left anterior oblique cineangiogram with injection into the left ventricle filling a large infundibuloventricular septal defect.

Cardiac catheterization demonstrated aortic atresia, a large patent ductus arteriosus, left superior vena cava to coronary sinus, mild hypoplasia of the left ventricle with a large ventricular septal defect and no significant mitral stenosis or regurgitation (Fig. 2).

Operations: On January 20, at age 3 days, with use of both a left thoracotomy and median stenotomy, a 10 mm Dacron® graft was placed from the proximal main pulmonary artery to the descending aorta. The patent ductus arteriosus was ligated and a band was placed around the main pulmonary artery distal to the graft. A second cardiac catheterization on January 31, performed because of persistent respiratory insufficiency, revealed occlusion of the origin of the right pulmonary artery by the band (Fig. 3). On February 6, with the infant under deep hypothermia and total circulatory arrest, a 12 mm valved conduit was placed from the right ventricular outflow tract to the descending aorta and the pulmonary artery band was moved to the repaired proximal main pulmonary artery.

Postoperatively, while hemodynamically stable, she required prolonged intermittent ventilator support and had nutritional insufficiency with gastrointestinal feeding intolerance. Prolonged intravenous hyperalimentation was required, with consequent Staphylococcus epidermidis septicemia requiring long-term antibiotic therapy. The patient is now 5 months old and has a satisfactory hemodynamic, respiratory and nutritional status.

Case 3: This infant, a 3.7 kg product of a full-term pregnancy by cesarean section, was transferred at age 2 days with findings of a heart murmur, cyanosis, decreased peripheral pulses and an echocardiographic diagnosis of hypoplastic left heart. Physical examination revealed a cyanotic infant with a heart rate of 150 beats/min, a respiratory rate of 80/min, mean blood pressure of 55 mm Hg in the arms and diminished peripheral pulses. There was a single second heart, a ventricular gallop sound and a grade 3/6 systolic ejection murmur. The chest roentgenogram revealed mild cardiac enlargement with increased pulmonary blood flow. The electrocardiogram disclosed a QRS axis of +50°, right atrial enlargement and right ventricular hypertrophy. Cardiac catheterization demonstrated aortic atresia, mitral atresia, a patent ductus arteriosus and a 20 mm Hg pressure gradient from the right ventricular sinus to descending aorta. The patient was resuscitated with a continuous infusion of prostaglandin E_1 and intermittent doses of sodium bicarbonate.

On March 3, at age 3 days, a 12 mm valved external conduit was anastomosed to the descending aorta through a left thoracotomy. Through a median sternotomy, using deep hypothermia and circilatory arrest, the proximal end of the conduit was anastomosed to the right ventricular outflow tract (Fig. 4). The ductus arteriosus was ligated and the main pulmonary artery banded. Postoperatively the infant received intravenous infusion of dopamine (5 μg/kg per min) and nitroprusside (5 μg/kg per min) for 3 days and was extubated on the 5th postoperative day. She was released from the hospital on the 24th postoperative day, feeding well, gaining weight and clinically well.

Discussion

Hypoplastic left heart syndrome: Aortic atresia or stenosis, atresia or interruption of the aortic arch, severe

stenosis or atresia of mitral valve and a small left ventricle compose a constellation of defects initially termed "hypoplasia of the aortic tract complexes" by Lev[5] in 1952 and later called hypoplastic left heart syndrome by Noonan and Nadas.[1] Among these, aortic atresia is uniquely complex and although numerous proposals have been reported, no effective management protocol has been established.[6–8] The central anatomic feature of each of the three patients in this report is aortic atresia, which is always complicated by associated severe hypoplasia of the ascending aorta and various degrees of mitral valve and left ventricular hypoplasia. When a large ventricular septal defect is present, as in Case 2, mitral valve and left ventricular hypoplasia may be mild. In a review of 172 patients with aortic atresia, Moodie et al.[7] identified 12 patients (7 percent) with an associated large ventricular septal defect. However, in 60 percent of cases the ventricular septum is intact and the mitral valve hypoplastic, allowing no egress of blood from the left ventricle, which remains diminutive with vacuolization of the subendocardial layer and development of large myocardial sinusoids. Aortic atresia and mitral atresia with no discernible left ventricle occur in combination in about one third of patients.

Prognosis: Perinatal survival is dependent on maintenance of the systemic circulation through a patent ductus arteriosus and on adequate pulmonary venous return, most commonly through an interatrial communication (usually a patent foramen ovale). Neonatal death usually results from inadequate coronary and systemic perfusion secondary to high pulmonary to systemic flow ratios with severe congestive heart failure or closure of the ductus arteriosus, or both. Less commonly, death results from hypoxemia and pulmonary venous congestion secondary to a restrictive interatrial communication. While a lone child with aortic atresia survived to age 3.5 years,[7] the reported mean survival time ranged from 4 to 23 days.[1,7]

Because of this high neonatal mortality rate, any systematic approach aimed at improving long-term survival for neonates with hypoplastic left heart syndrome requires early recognition and definition of pertinent anatomic features with cardiac catheterization and echocardiography (including the presence of aortic atresia, the presence or absence of a large ventricular septal defect, the size of the interatrial communication, the status of the mitral valve and the presence and location of coarctation of the aorta[9]). This should be followed, if necessary, by resuscitation with balloon atrial septotomy and prostaglandin E_1 infusion to maintain ductal patency in preparation for early surgical intervention.

Surgical considerations: Except in the presence of an associated large ventricular septal defect and two well developed ventricles, physiologic correction of aortic atresia is possible only by the separation of the pulmonary and systemic circulations using the right ventricle as the systemic pumping chamber. The feasibility of utilizing the right atrium as the pulmonary pumping chamber has been established both experimentally and clinically,[10,11] but requires near normal

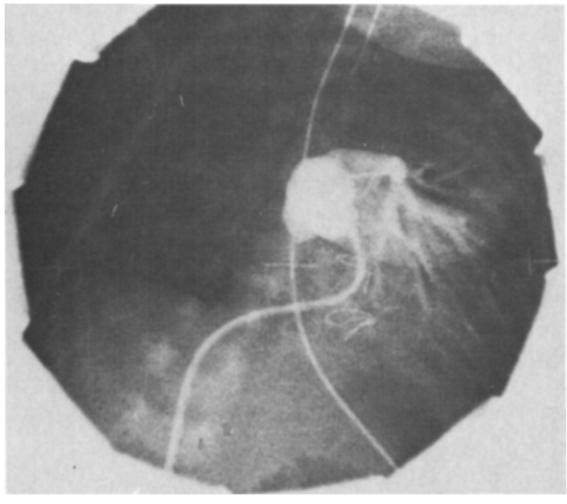

FIGURE 3. Case 2. Frame from anterior-posterior postoperative cineangiogram with injection into the main pulmonary artery filling the conduit and the left pulmonary artery. The right pulmonary artery is occluded by the pulmonary arterial band.

adult pulmonary vascular resistance with a mean pulmonary arterial pressure of less than 20 mm Hg and normal ventricular end-diastolic pressure. In the newborn period, however, pulmonary vascular resistance is elevated and may not decrease to adult levels until the infant is 6 to 8 weeks of age.[2] Furthermore, a postmortem quantitative pulmonary morphometric analysis by Heidelberger et al.[12] of neonates with the hypoplastic left heart syndrome demonstrated pulmonary vascular

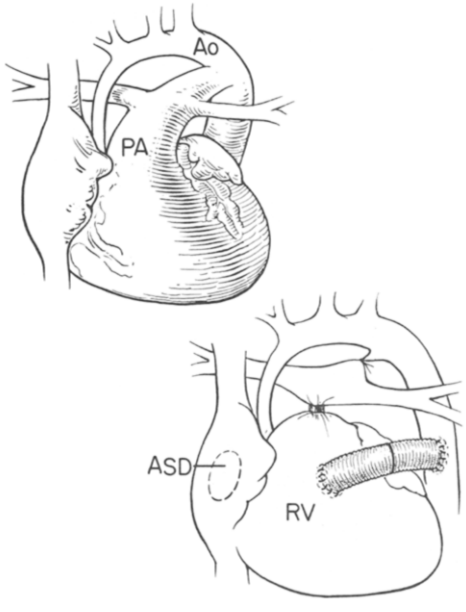

FIGURE 4. Case 3. Artist's conception of aortic atresia (**top**) and palliative procedure (**bottom**). Lower rendition illustrates ligation of patent ductus arteriosus, band on the main pulmonary artery (PA) and valved conduit between the right ventricular (RV) outflow tract and the descending aorta. Ao = aorta; ASD = atrial septal defect.

changes (including extension of smooth muscle into distal pulmonary arteries, increased wall thickness of distal arteries and slightly decreased arteriolar to alveolar ratio) that are likely to result in elevated pulmonary vascular resistance. Because pulmonary vascular resistance is expected to be high in the neonate with hypoplastic left heart syndrome, we now consider a multistage approach most likely to produce long-term survival.

Multistage surgical approach: *The principles of the first stage include* (1) establishment of a permanent communication between the right ventricle and aorta, (2) limitation of pulmonary blood flow to attenuate the pulmonary vascular changes secondary to elevated pulmonary blood flow and pressure, and (3) insurance of a satisfactory interatrial communication in the presence of mitral valve hypoplasia or atresia. Two attempts, by Cayler et al.[13] and Freedom et al.,[13] at surgical palliation incorporated these principles using a Waterston shunt in a neonate with critical aortic stenosis[13] and a Potts shunt in a child with aortic atresia and ventricular septal defect.[8] Both procedures were followed by right and left branch pulmonary arterial banding. The palliative effects lasted 7 and 16 months, respectively. However, the difficulty in achieving limited and bilaterally balanced pulmonary blood flow and the risk of anatomic distortion of the distal pulmonary arteries make branch pulmonary artery banding undesirable.

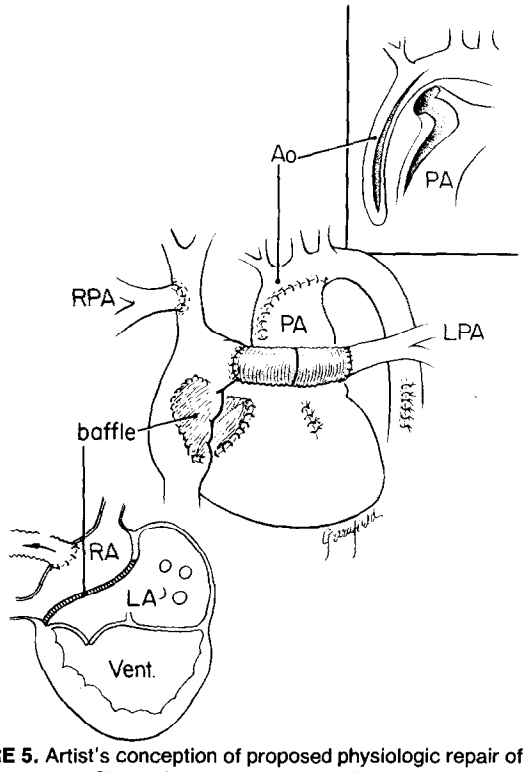

FIGURE 5. Artist's conception of proposed physiologic repair of aortic atresia (stage 2). The main pulmonary artery (PA) and the aorta (Ao) are anastomosed using the proximal stump of the right pulmonary artery (RPA) (**upper panel**). The right pulmonary artery is anastomosed end to side with the superior vena cava and a valved conduit placed between the right atrium (RA) and left pulmonary artery (LPA). An interatrial baffle ensures filling of the right ventricle (Vent.) with pulmonary venous blood from the left atrium (LA). A schematic drawing of the baffle and conduit is shown in the **lower panel.**

Litwin et al.[14] avoided branch pulmonary arterial banding by placement of a nonvalved conduit from the main pulmonary artery to the descending aorta and placing a band on the main pulmonary artery distal to the conduit in four infants with an interrupted aortic arch. The one survivor manifested not only severe distortion of the pulmonary architecture by the distal pulmonary arterial band but also pulmonary vascular obstructive disease.

On the basis of the experience reported here, we propose that a more satisfactory means of accomplishing the palliative goals outlined is placement of a valved conduit from the right ventricular free wall to the descending aorta allowing ample room for the standard placement of a pulmonary arterial band on the main pulmonary artery (Fig. 4). This can be accomplished by end to side anastomosis between the valved conduit and descending aorta through a left thoracotomy incision using a partially occluding clamp. After placing the conduit entirely within the left chest, the thoracotomy incision is closed and the patient repositioned for a median sternotomy. Cardiopulmonary bypass is established by cannulation of the main pulmonary artery for arterial inflow through the ductus arteriosus and cannulation of the right atrial appendage for venous return. The branch pulmonary arteries are occluded during the cooling phase and, with use of deep hypothermic circulatory arrest the proximal end of the conduit is anastomosed to the right ventricular outflow tract. The atrial septal defect may be enlarged if necessary. The ductus arteriosus is ligated and the main pulmonary artery is banded after discontinuation of cardiopulmonary bypass. This approach was used on a 2 day old boy (Case 3) with aortic and mitral atresia who was released from the hospital on the 24th postoperative day and is now clinically well at age 3 months.

The second stage of the surgical approach, subsequent physiologic correction of this lesion, is dependent on separation of the systemic and pulmonary circulations. Although many procedures have been proposed,[6,8,15,16] we believe that utilization of autologous valve and vascular tissue in the systemic circulation and a valved external conduit for the pulmonary circuit maximize both simplicity of technique and potential for growth (Fig. 5). As demonstrated in Case 1, anterograde flow through the ascending aorta can be established by side to side anastomosis of the pulmonary artery and diminutive ascending aorta using a portion of the right branch pulmonary artery (upper panel, Fig. 5). An interatrial baffle is constructed to provide continuity between the left atrium and tricuspid valve through an atrial septal defect. The pulmonary arterial circulation can be completed by anastomosis of the distal right pulmonary artery to the superior vena cava and placement of a conduit from the right atrium to the left pulmonary artery (Fontan procedure). When a large ven-

tricular septal defect and left ventricle coexist, the left ventricle can be used for the systemic circulation by connecting the ventricular septal defect and pulmonary artery with an interventricular baffle while the palliative valved conduit on the right ventricular outflow tract is transferred from the descending aorta to the distal pulmonary artery. In these cases the atrial septal defect is merely closed.

Particularly in patients with aortic atresia and intact ventricular septum, this physiologic repair requires both low pulmonary vascular resistance and minimal secondary changes of the pulmonary architecture from palliative surgery. Available experimental and clinical evidence suggests that early smooth muscle alterations in pulmonary arterioles, secondary to a large left to right shunt, are reversible after adequate pulmonary arterial banding.[17] Because normal pulmonary vascular resistance is expected after several months and because secondary changes from the pulmonary band may occur after 1 year, the current plan is to perform repair in these two children before they are 1 year of age. The ultimate efficacy of these procedures remains to be demonstrated, but early results of stage I palliation in our two patients are encouraging.

References

1. **Noonan JA, Nadas AS.** The hypoplastic left heart syndrome: an analysis of 101 cases. Pediatr Clin North Am 1958; 5:1029–56.
2. **Rudolph AM.** Congenital Diseases of the Heart. Chicago: Yearbook Medical Publishers, 1974:29–48, 550.
3. **Norwood WI, Castaneda AR.** Tricuspid atresia and Ebstein's anomaly. In: Ravitch MM, Welch K, Benson C, Aberdeen E and Randolph J, eds. Pediatric Surgery, 3rd ed. Chicago: Yearbook Medical Publishers, Chicago, 1979:741–4.
4. **Gale AW, Danielson GK, McGoon DC, Wallace RB, Mair DD.** Modified Fontan operation for univentricular heart and complicated congenital lesions (abstr). J Thorac Cardiovasc Surg, in press.
5. **Lev M.** Pathologic anatomy and interrelationship of the hypoplasia of the aortic tract complexes. Lab Invest 1952; 1:61–70.
6. **Doty DB, Knotts HW:** Hypoplastic left heart syndrome: experience with an operation to establish functionally normal circulation. J Thorac Cardiovasc Surg 1977; 74:624–30.
7. **Moodie DS, Gallen WJ, Friedberg DZ.** Congenital aortic atresia: report of long survival and some speculations about surgical approaches. J Thorac Cardiovasc Surg 1972; 63:726–31.
8. **Freedom RM, Culham JAG, Rowe RD.** Aortic atresia with normal left ventricle distinctive angiocardiographic findings. Cathet Cardiovasc Diagn, 1977; 3:283–95.
9. **Von Rueden TJ, Knight L, Moller JH, Edwards JE.** Coarctation of the aorta associated with aortic valvular atresia. Circulation 1975; 52:951–4.
10. **Sade RM, Castaneda AR.** The dispensable right ventricle. Surgery 1975; 77:624–31.
11. **Fontan F, Choussat A, Coqueran J, Chauve A, Baudet E.** Repair of tricuspid atresia: surgical considerations and results (abstr). Circulation 1974; 50:Suppl III:III-72.
12. **Heidelberger KP, Neumann MS, Dick M, Rosenthal A.** Fetal pulmonary vascular changes associated with hypoplastic left ventricle syndrome (abstr). Am J Cardiol 1979; 43:364.
13. **Cayler GC, Smeloff EA, Miller GE.** Surgical palliation of hypoplastic left side of the heart. N Engl J Med 1970; 14:780–3.
14. **Litwin SB, Van Praagh R, Bernhard WF.** A palliative operation for certain infants with aortic arch interruption. Ann Thorac Surg 1972; 14:369–75.
15. **Freedom RM, Williams WG, Dische MR, Rowe RD.** Anatomical variants in aortic atresia: potential candidates for ventriculoaortic reconstitution. Br Heart J 1976; 38:821–6.
16. **Thiene G, Gallucci V, Macartney FJ, Del Torso S, Pellegrino PA, Anderson RH:** Anatomy of aortic atresia: cases presenting with a ventricular septal defect. Circulation 1979; 59:173–8.
17. **Rabinovitch M, Haworth SG, Castaneda AR, Nadas AS, Reid LM:** Lung biopsy in congenital heart disease: a morphometric approach to pulmonary vascular disease. Circulation 1978; 58:1107–22.