Hemodynamic assessment after palliative surgery for hypoplastic left heart syndrome

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ABSTRACT Ten patients with hypoplastic left heart syndrome underwent cardiac catheterization to assess the effectiveness of palliative surgery designed to prepare them for a modified Fontan procedure. The objectives of palliation were to establish unobstructed systemic blood flow, normalize pulmonary blood flow and pressure, and relieve pulmonary venous obstruction. In the first four patients, systemic blood flow from the right ventricle was established by means of a conduit from either the right ventricular free wall or the proximal main pulmonary artery to the thoracic aorta. Pulmonary blood flow was limited by pulmonary artery banding and ligation of the ductus arteriosus in three patients and by a Blalock-Taussig shunt in one. Conduit obstruction of systemic blood flow developed in two of these infants, and pulmonary vascular abnormalities precluded reparative surgery in the other two. In the other six patients, systemic blood flow was established by direct anastomosis of the proximal main pulmonary artery to the ascending aorta and aortic arch. The pulmonary vasculature was protected by providing pulmonary flow through a central or Blalock-Taussig shunt. The interatrial communication was enlarged by atrial septectomy or balloon atrial septotomy. One infant had progressive tricuspid regurgitation necessitating valve replacement. One, in whom balloon atrial septotomy had been performed, developed an obstructive interatrial communication necessitating late atrial septectomy. These six patients were candidates for physiologic correction. To date, three patients have undergone a modified Fontan procedure; two are clinically well at 9 and 12 months after surgery. Three patients await this procedure.


NEWBORNS with hypoplastic left heart syndrome (HLHS) have one effective ventricle and depend on patency of the ductus arteriosus for maintenance of the systemic circulation. Physiologic closure of the ductus arteriosus in the neonatal period is the most common cause of death. Any significant improvement in long-term outlook for infants with HLHS is based on the feasibility of separating the pulmonary and systemic circulations, which is analogous to Fontan’s physiologic correction of tricuspid atresia, another form of congenital heart disease in which there is only one effective ventricle. The use of such a procedure is predicated on the observation that pulmonary circulation can be maintained without a ventricular pump if pulmonary vascular resistance is low, as it is in the normal mature lung. Pulmonary vascular resistance of the newborn infant, however, is physiologically high, and a staged surgical approach is therefore necessary for definitive treatment of infants with HLHS. The initial palliation must establish permanent unobstructed communication between the right ventricle and the aorta, limit the pulmonary blood flow and pressure to near normal levels, and ensure a large interatrial communication; the first objective is necessary for adequate systemic perfusion and to preserve ventricular function, while the second two are essential for normal development of the pulmonary vasculature. Our initial results with developing surgical techniques to meet these objectives have been reported. In this article we present hemodynamic data from cardiac catheterization of 10 survivors of palliative surgery to assess their suitability for a modified Fontan procedure.

Methods

Between September 1979 and December 1982, 35 infants with HLHS (ages 1 day to 6 months, median 5 days) underwent various forms of palliative surgery at the Children’s Hospital Medical Center, Boston. The surgical procedures used in these patients have been previously described. Briefly, in our study group of 10 patients, the first four underwent procedures in which a conduit was placed in the systemic circulation, either from the free wall of the right ventricle to the thoracic aorta or

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from the main pulmonary artery to the aorta. In three infants, pulmonary blood flow was limited by pulmonary artery banding and ligation of the ductus arteriosus. In the fourth, pulmonary blood flow was limited by a Blalock-Taussig shunt (table 1). The last six patients in the study group were palliated by an operation that evolved from our previous experience (figure 1). Output from the right ventricle to the aorta was established through the proximal main pulmonary artery, which was used to reconstruct the diminutive ascending aorta and aortic arch. Pulmonary blood flow was established by a central or Blalock-Taussig shunt. Pulmonary venous obstruction was relieved by balloon atrial septotomy or atrial septectomy. All procedures were performed during a period of circulatory arrest, with body temperature at 20°C.

Ten patients underwent cardiac catheterization 2 weeks to 16 months after surgery (median 6 months). Four of these patients underwent catheterization for clinical indications and six were electively studied to assess effectiveness of the palliative surgery. The objectives of the hemodynamic studies were to determine (1) the pressure difference between the right ventricle and the thoracic aorta, (2) right ventricular end-diastolic pressure, (3) systemic arterial oxygen saturation, (4) pulmonary vascular resistance, and (5) left and right atrial pressures.

Results

Of the 13 patients discharged from the hospital after palliative surgery, 10 underwent cardiac catheterization (table 1). As noted, the first four patients underwent closely related surgical procedures that established systemic circulation by means of a conduit. Patient M. W. was catheterized in extremis and had a right ventricular pressure of 135/19 mm Hg. A right ventricular angiogram demonstrated thrombotic obstruction of the conduit from the right ventricle to the thoracic aorta. Patient J. M. was also seriously ill at the time of cardiac catheterization. There was a 30 mm Hg systolic pressure gradient of a main pulmonary artery to aortic arch nonvalved conduit at the distal anastomosis. Patient N. D. had no obstruction to systemic output but had complete occlusion of the right pulmonary artery secondary to a main pulmonary artery band placed distal to a conduit.3,4 There was no obstruction to systemic output or derangement of the pulmonary artery architecture in the last patient in this group (J. L.), but there was elevated pulmonary artery pressure secondary to an obstructive interatrial communication.

The remaining six patients were palliated by the procedure we now favor (figure 1). None of these patients had a significant systolic pressure gradient between the right ventricle and the thoracic aorta. One patient who underwent a preoperative balloon atrial septotomy had a restrictive interatrial communication with a 19 mm Hg gradient between the left and right atria. Another patient had progressive tricuspid regurgitation that was treated by replacement of a structurally abnormal tricuspid valve. Postoperatively, all patients had low right ventricular end-diastolic pressure and pulmonary resistance.

Discussion

The term “hypoplastic left heart syndrome” has been used to describe various degrees of underdevelopment of the left heart structures. In the most complex form of HLHS, the central anatomic feature is atresia of the aortic valve, which is associated with severe hypoplasia of the ascending aorta. In over 90% of the patients with aortic atresia there is coexisting atresia or severe hypoplasia of the mitral valve and a diminutive or absent left ventricle.

HLHS is not rare. It is the fourth most common cardiac defect reported in the New England Regional Infant Cardiac Program (NERICP) and accounts for nearly 25% of cardiac deaths during the first week of life. Between 1969 and 1979 there were 223 infants reported to the NERICP having HLHS with aortic atresia. None survived a year. Unlike infants with many other forms of congenital heart disease, newborns with HLHS are most often of normal birth weight and have a low incidence of accompanying extracardiac anomalies.

With the prospect of achieving significant physiologic improvement in patients with all forms of single ventricle by the use of techniques originally introduced by Fontan, we were stimulated in 1979 to examine the feasibility and efficacy of surgical treatment of infants with HLHS. The history of surgical management of this defect is not extensive and has been reviewed previously.

The focus of this report is the hemodynamic assessment of patients surviving surgery; however, the mortality in this series requires comment. The infants in this study were treated with a diverse and evolving set of techniques, with higher mortality early in our experience compared with more recent results. The majority of deaths were attributable to technical difficulties in the perioperative period, including compromise of coronary perfusion, excessive or insufficient pulmonary blood flow, obstructed systemic conduit, restrictive interatrial communication, and obstructed endotracheal airway. These deaths were considered to be related to technical problems that should be amenable to refinements in surgical technique and perioperative management. Three patients died of a hemorrhagic diathesis, one neonate had occlusion of the right coronary artery by an embolus on the second postoperative day. Some infants could not be weaned from cardiopulmonary bypass.
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On the basis of the hemodynamic outcome documented here, the surgical palliation we currently favor provides antegrade flow from the right ventricle to the ascending aorta and aortic arch all demonstrated growth of the reconstructed aorta (figure 2). Of theoretical concern is the eventual limitation of coronary flow by the native severely hypoplastic aortic root. Postoperative angiograms, however, demonstrate growth of the aortic root and coronary arteries, and there have been no instances of late or progressive coronary insufficiency.

### TABLE 1
Cardiac catheterization data

<table>
<thead>
<tr>
<th>Patient/age at cath. (mo.)</th>
<th>Procedure</th>
<th>RVP (mm Hg)</th>
<th>AP (mm Hg)</th>
<th>PAP (mm Hg)</th>
<th>PVR (mm Hg/l/min/m²)</th>
<th>Left-to-right ATP gradient (mm Hg)</th>
<th>Arterial O₂ saturation (%)</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>M. W./3</td>
<td>RV to descending aorta valved conduit; MPA band; PDA ligation</td>
<td>135/19</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>69</td>
<td>Thrombotic obstruction of RV to aortic conduit, pH 7.0</td>
</tr>
<tr>
<td>J. M./5</td>
<td>Proximal MPA to aortic arch nonvalved conduit; transection distal MPA; PDA ligation; right BT shunt</td>
<td>115/9</td>
<td>85/42</td>
<td>—</td>
<td>4</td>
<td>72</td>
<td>Severe low output at cath.</td>
<td></td>
</tr>
<tr>
<td>N. D./0.5</td>
<td>Proximal MPA to descending aorta nonvalved conduit; distal MPA band; PDA ligation</td>
<td>85/9</td>
<td>85/33 56</td>
<td>—</td>
<td>2</td>
<td>80</td>
<td>Distal pulmonary artery band with occlusion of right pulmonary artery</td>
<td></td>
</tr>
<tr>
<td>J. L./2</td>
<td>RV to descending aorta valved conduit; MPA band; PDA ligation</td>
<td>109/8</td>
<td>101/60 72</td>
<td>10</td>
<td>2.8</td>
<td>4</td>
<td>50</td>
<td></td>
</tr>
<tr>
<td>J. L./13</td>
<td>Transection distal MPA; proximal MPA to ascending aorta anastomosis; RV to distal MPA valved conduit (banded)</td>
<td>107/10</td>
<td>103/57 74</td>
<td>19</td>
<td>2.3</td>
<td>6</td>
<td>75 Subsequent modified Fontan procedure</td>
<td></td>
</tr>
<tr>
<td>J. P./7</td>
<td>Proximal MPA to ascending aorta anastomosis; transection distal MPA; neoaoorta to distal MPA shunt; PDA ligation; atrial septectomy</td>
<td>115/2</td>
<td>108/41 69</td>
<td>—</td>
<td>—</td>
<td>3</td>
<td>67 Subsequent right BT shunt</td>
<td></td>
</tr>
<tr>
<td>J. H./7</td>
<td>Proximal MPA to ascending aorta anastomosis; transection distal MPA; neoaoorta to distal MPA shunt; PDA ligation; atrial septectomy</td>
<td>116/6</td>
<td>116/58 80</td>
<td>11</td>
<td>2.7</td>
<td>1</td>
<td>71 Subsequent modified Fontan procedure</td>
<td></td>
</tr>
<tr>
<td>S. K./16</td>
<td>Proximal MPA to ascending aorta anastomosis; transection distal MPA; neoaoorta to distal MPA shunt; PDA ligation; atrial septectomy</td>
<td>94/8</td>
<td>92/53 61</td>
<td>7</td>
<td>0.9</td>
<td>0</td>
<td>79 Primum type atrial septal defect noted at surgery; subsequent modified Fontan procedure</td>
<td></td>
</tr>
<tr>
<td>G. A./14</td>
<td>Proximal MPA to ascending aorta anastomosis; transection distal MPA; neoaoorta to distal MPA shunt; PDA ligation; atrial septectomy; right BT shunt (2 wk later)</td>
<td>130/4</td>
<td>118/96 77</td>
<td>12</td>
<td>1.8</td>
<td>1</td>
<td>72 Occluded central shunt; subsequent modified Fontan procedure</td>
<td></td>
</tr>
<tr>
<td>D. J./6</td>
<td>Proximal MPA to ascending aorta anastomosis; transection distal MPA; right BT shunt; PDA ligation; atrial septectomy</td>
<td>130/18</td>
<td>126/56 86</td>
<td>28/14 20</td>
<td>2.0</td>
<td>0</td>
<td>72 Severe tricuspid regurgitation; subsequent tricuspid valve replacement</td>
<td></td>
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<tr>
<td>P. T./2</td>
<td>Balloon atrial septotomy; aortic valvotomy; proximal MPA to ascending aorta anastomosis; transection distal MPA; right BT shunt; PDA ligation</td>
<td>70/14</td>
<td>71/45</td>
<td>—</td>
<td>—</td>
<td>19</td>
<td>65 Subsequent atrial septectomy</td>
<td></td>
</tr>
</tbody>
</table>

RVP = right ventricular pressure; AP = aortic pressure; ATP = atrial pressure; PAP = pulmonary artery pressure; PVR = pulmonary vascular resistance; RV = right ventricle; MPA = main pulmonary artery; PDA = patent ductus arteriosus; BT = Blalock-Taussig.
As a result of unobstructed outflow and limited pulmonary blood flow, right ventricular function has been well preserved during the follow-up period. The ultimate fate of the right ventricle functioning at systemic pressure remains a hypothetical concern. Three patients had tricuspid regurgitation on presentation as neonates. One patient could not be weaned from cardiopulmonary bypass; the second infant developed progressive regurgitation from a structurally abnormal valve, which was later replaced. The third patient's tricuspid regurgitation was mild to moderate when she presented at 2 weeks of age and it abated shortly after palliative surgery, suggesting that the tricuspid valve regurgitation was at least in part related to preoperative ventricular dysfunction. At present we consider tricuspid regurgitation a relative contraindication to surgical management.

Our experience suggests that the pulmonary vasculature is best preserved and protected by the use of a shunt to provide pulmonary artery blood flow. The pulmonary vascular resistance is extremely dynamic in these neonates perioperatively, and the appropriate pulmonary blood flow is often difficult to establish. Recently, the use of a Blalock-Taussig shunt has been most effective.

Pulmonary blood flow as well as maturation of the

FIGURE 1. Procedure for palliation of HLHS. A, The heart with aortic atresia has a diminutive ascending aorta and aortic arch and a large patent ductus arteriosus. B, The main pulmonary artery is transected and an incision is made in the ascending aorta and aortic arch. C, The distal main pulmonary artery is oversewn and the distal anastomosis of the 4 mm shunt is established. D, The main pulmonary artery is anastomosed to the ascending aorta and aortic arch and the ductus arteriosus is ligated.
pulmonary vasculature are influenced by venous obstruction arising from a restrictive interatrial communication. Most often the interatrial communication in HLHS is a foramen ovale. Although a balloon atrial septotomy was effective in creating a large interatrial communication in three patients, the septum primum is often abnormally thick in HLHS. In one neonate the balloon septotomy did not permanently augment the atrial septal defect. Thus atrial septectomy is an important feature of the palliative surgery.

Von Reuden et al. noted that seven of 41 patients with aortic atresia had coexisting coarctation of the aorta. A mild coarctation was the cause of the small gradient noted on the catheterization of one patient (G. A.). Preoperative evaluation of the aortic isthmus by cineangiography is important so that coarctations may be repaired.

Four of the 10 patients studied underwent a modified Fontan procedure, in which the atria was partitioned so that the left atrium was associated with the tricuspid valve and the right atrium was anastomosed to the pulmonary arteries. The first patient died of progressive right heart failure, with evidence of increased pulmonary vascular resistance. He had a moderately restrictive interatrial communication documented at cardiac catheterization before corrective surgery. The second patient died of sudden and unexplained hyperkalemia several days after surgery. The last two patients, who had separation of the pulmonary and systemic circulations at 16 and 13 months of age, had an unremarkable postoperative convalescence and are thriving with normal development 12 and 9 months after surgery.

On the basis of these data, we believe that the features of successful palliative surgery for HLHS include the use of autologous valve and vascular tissue to reconstruct the aorta, the use of a small nondistorting shunt from the systemic circulation to the pulmonary artery, and the creation of a large interatrial communication. Palliative surgery can be successfully performed in neonates with HLHS with good hemodynamic results. Once this has been accomplished, a modified Fontan procedure can then be performed. The advisability of surgical management of HLHS, however, will depend on improving early results and extensive long-term follow-up.

References
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