Chapter 8

Hypoplastic Left Heart Syndrome: Why Use a Hybrid Procedure?

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Additional information is available at the end of the chapter

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1. Introduction

Neonates with hypoplastic left heart syndrome (HLHS) and its variants have diminutive left-sided structures with systemic outflow obstruction and subsequent duct-dependent systemic, cerebral, and coronary circulations. [1]

Bilateral PA banding was described by Norwood based on his early experience of surgical palliation for HLHS before he established the 'Norwood procedure [2] and has been an option to stabilize critically ill neonates with HLHS.[3] This procedure, however, did not become a hybrid procedure until ductal stenting became clinically available.

The Norwood procedure has been the sole option to achieve these purposes for nearly 20 years. [4], [5] Despite a significant improvement in survival after stage I palliation, early mortality remains as high as 20-30% [6], [7] with significant morbidity, including neurologic injury. [8], [9], due to deep hypothermia, cardiocirculatory arrest, prolonged cardiopulmonary bypass, sternum delayed closure, use of extracorporeal membrane oxygenator (ECMO) and prolonged Hospital stay.

Survivors Norwood op. are showing suboptimal neurological development. (Figure 1)

In the last decade the hybrid procedure has emerged as an alternative stage I palliation in neonates with HLHS. (Gibbs et al) [10].: Stenting of the arterial duct combined with banding of the pulmonary arteries and atrial septectomy or septostomy: a new approach to palliation for the hypoplastic left heart syndrome

This review discusses the historical aspect, surgical and interventional techniques, current outcomes and future direction of this procedure.(Figure 2)

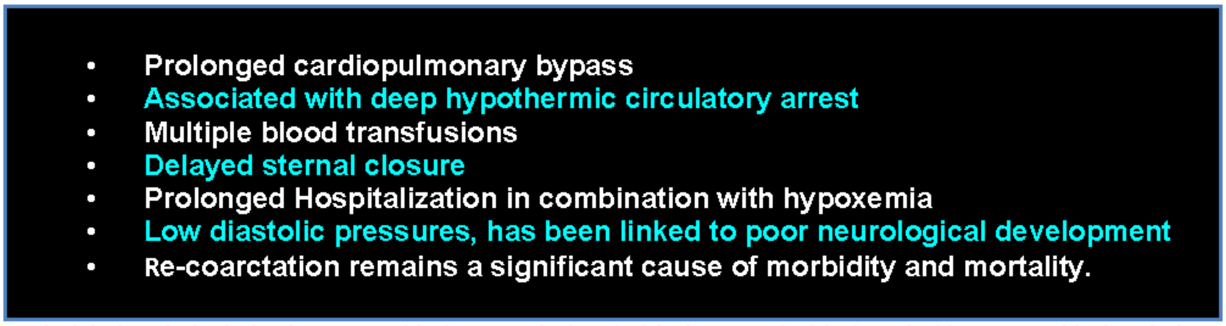

Figure 1. Causes of suboptimal neurologic development

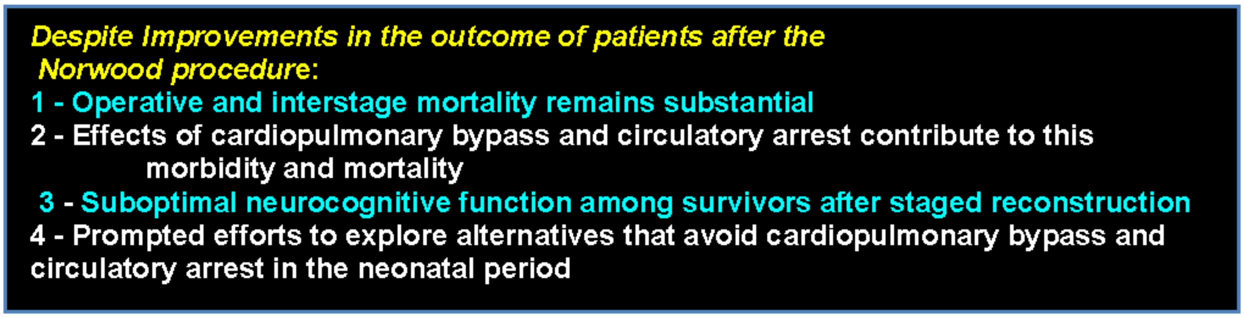

Figure 2. Vantages of Hybrid Procedures in patients with Hypoplastic Left Heart Syndrome

Hybrid palliation yields equivalent but not superior stage I palliation survival and comparable 1-year survival to conventional Norwood palliation, comparable pre-stage II hemodynamics and pulmonary artery growth, and preserved ventricular function in stage II palliation. Hybrid palliation utilizes significantly less resource and shortens postoperative recovery. In comprehensive stage II palliation the impact of pulmonary artery reconstruction on subsequent pulmonary artery growth has not been determined and should be further investigated. A prospective, randomized trial is warranted to compare these two surgical strategies for neonates with hypoplastic left heart syndrome.

The high mortality and morbidity stems from the following two components: an essentially unstable "in-parallel" circulation of Norwood physiology with a systemic-to-pulmonary shunt, and surgical stress driven by cardiopulmonary bypass (CPB), deep hypothermic circulatory arrest (DHCA), and the subsequent systemic inflammatory response.

2. Hybrid procedure

2.1. Indications

In High-risk patients:

- Aortic atresia

- Severe non-cardiac anomalies
- Low body weight (<2.5 kg)
- Intact or highly restrictive atrial septum
- Prematurity
- Poor ventricular function
- Diminutive aortic arch
- Stenotic aortic isthmus

The HLHS variants, such as tricuspid atresia with transposed great arteries, unbalanced atrioventricular septal defect with aortic arch obstruction, or double inlet left ventricle with transposed great arteries, are all considered an indication for the hybrid procedure. [11]

Staged surgical palliation with the standard Norwood approach, hybrid approach, and primary transplantation are equally offered to the patient's family by the cardiologist, and no specific decision-making protocol is applied

In patients with aortic atresia, the coronary and cerebral circulations are entirely dependent on retrograde blood flow through the aortic isthmus. If there are any signs of pre-operative obstruction at the aortic isthmus or retrograde aortic arch, deployment of a ductal stent can result in acute or chronic obstruction of the aortic isthmus, leading to coronary and cerebral ischemia.[12] This specific anatomic feature can be a relative contraindication for a 'conventional' hybrid procedure [13] unless specific measures are applied to secure the coronary and cerebral circulations:

Surgical procedures proposes

- Prophylactic main PA to innominate artery shunt during stage I palliation: Reverse Blalock-Taussig (BT) shunt
- Stent placement in the stenotic aortic isthmus
- Avoidance of the hybrid procedure in favor of the Norwood procedure or transplantation.
- Operative Techniques
- Hybrid procedure for the treatment of HLHS is preferably indicated in the following situations (Figure 3)

2.2. Stage I palliation

2.2.1. Hybrid stage I

The first hybrid procedure for the palliative treatment of HLHS,was held in São Paulo Federal University, was held in 1995 in a patient 30 days, weight 3,800 grams by reason of the refusal of parents to the procedures offered: Op Norwood or heart transplant [14]

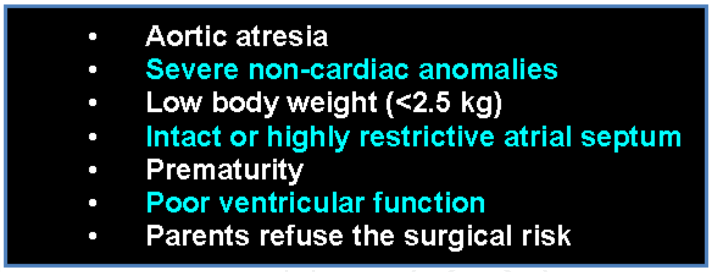

Figure 3. Indication Hybrid procedure

Palliation has been previously described. [9, 10] Herein, the current techniques used in The Hospital for Sick Children are described. The procedure is performed in the catheterization laboratory, which is referred to as the 'hybrid suite'. Under general anesthesia, a median sternotomy is made and the thymus is resected. Bilateral PA banding is achieved using a 3.5 mm polytetrafluoroethylene graft, which is divided longitudinally and wrapped around the branch PAs for a width of approximately 3 mm.

The bands are then secured to the adventitia of the main PA with 5-0 or 6-0 non-absorbable polypropylene sutures to avoid distal dislodging. A vascular clip is placed at the proximal edge of the left PA band to guide the interventionist for ductal stenting.

The patient is partially heparinized, a purse-string suture is placed on the main PA, and a 6-Fr sheath is inserted. The ductal stent is deployed through the sheath under fluoroscopic guidance, achieving stage I palliation (Figs. 4, 5, 6, 7 and 8). We currently use a self-expanding stent, 20 mm in length and 8-10 mm in diameter

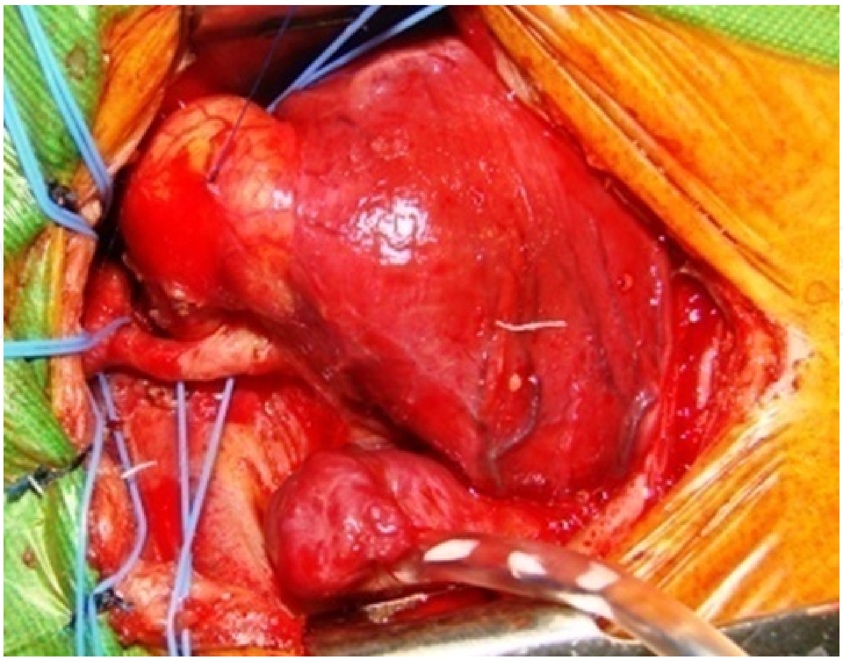

Figure 4. Surgical view – Case with Hypoplastic Left Heart Syndrome (HLHS)

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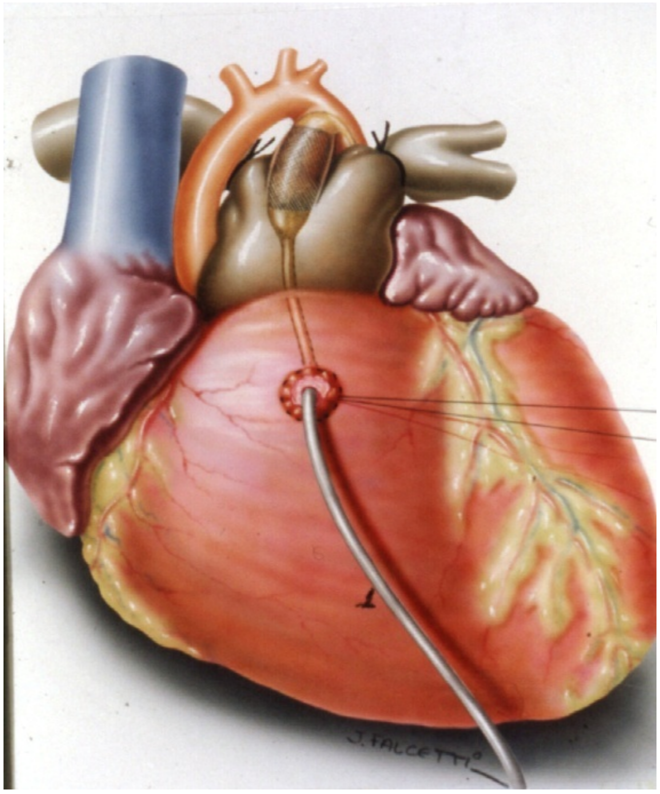

Figure 5. Surgical draw of hybrid procedure: A purse-string suture is placed on the main PA, and a 6-Fr sheath is inserted. The ductal stent is deployed through the sheath under fluoroscopic guidance, achieving stage I palliation

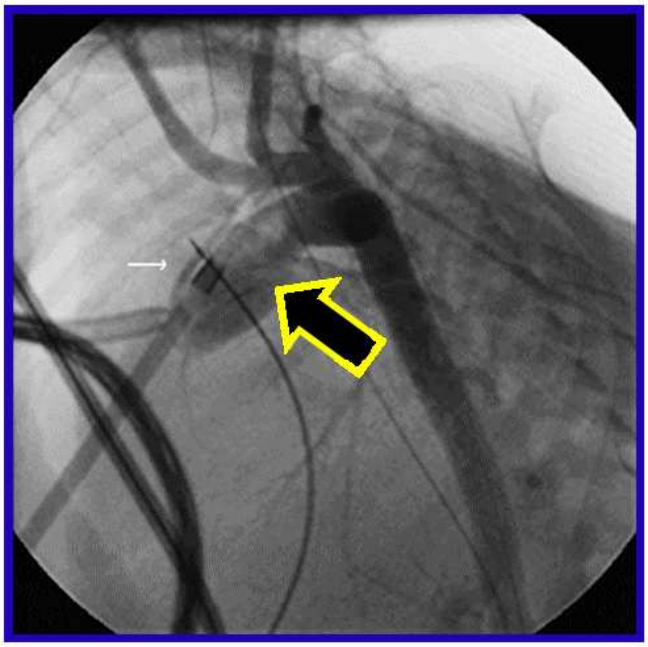

Figure 6. Angiographic study during stent deployment in hybrid stage I palliation. The lateral view showing the sheath and guide wire inserted via the main PA through the Arterial Duct. (arrow)

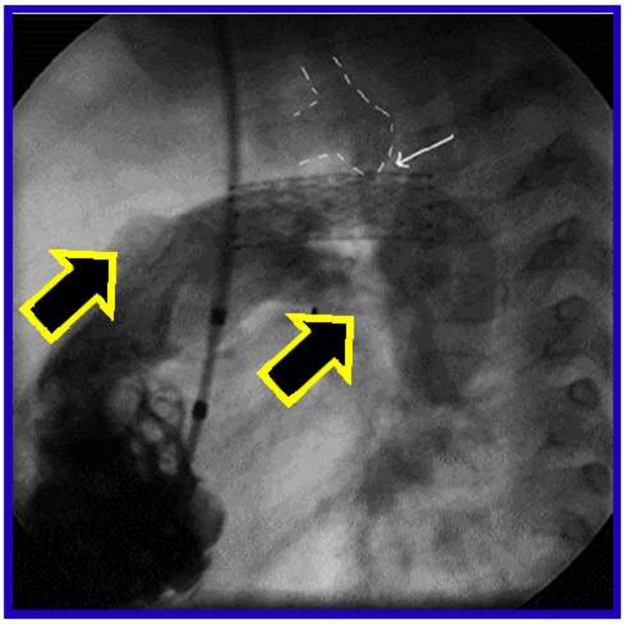

Figure 7. Angiographic sequence of stent deployment in hybrid stage I palliation. The lateral view showing the stent into arterial duct (thin arrow) and bilateral banding of the Pas (large arrow).

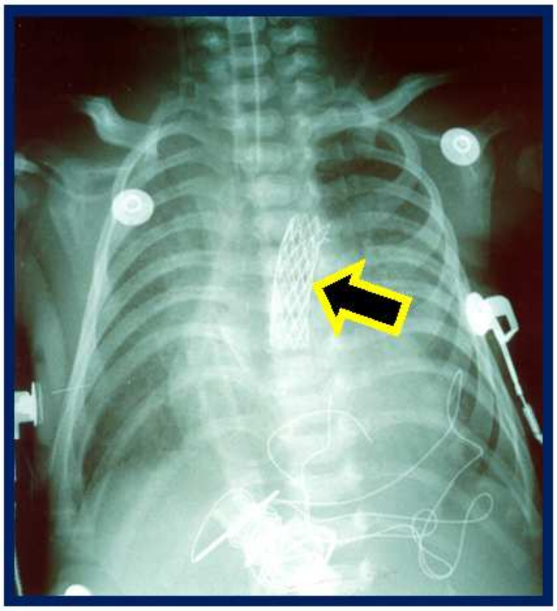

Figure 8. Post-operative Chest X Ray showing the stent into the Arterial Duct (arrow)

- Milrinone is commonly used in post-operative care to optimize cardiac output and provide systemic vasodilatation. This management strategy stems from our early observation that patients who underwent a hybrid procedure had diminished cardiac output, high systemic vascular resistance, and a high pulmonary-to-systemic flow ratio (Qp/Qs) despite avoiding CPB/DHCA.

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- In our early experience, atrial septectomy was typically performed as part of the same procedure; currently atrial septectomy and stenting are deferred until the intra-atrial communication becomes restrictive.

Reverse Blalock-Taussig shunt

- In patients with aortic atresia or those thought to have severely restricted prograde aortic flow, a revered BT shunt is prophylactically placed from the main PA to the innominate artery.. After bilateral PA banding is achieved, a side-biting clamp is placed on the main PA and proximal anastomosis is made using a 3.5 or 4 mm polytetrafluoroethylene graft on the anterior wall of the main PA. Distal anastomosis is then performed to the proximal innominate artery with a standard anastomotic technique. [11].The flow in the shunt is directed from the main PA to the innominate artery. Ductal stenting is then performed.

Interstage monitoring

- Close monitoring with weekly or biweekly clinic visits and echocardiograms are performed. The arm-leg blood pressure difference is measured at every clinic visit to exclude any signs of aortic arch obstruction. Echocardiographic follow-up is particularly focused on the presence or absence of flow acceleration in the aortic isthmus, indicating progression of retrograde aortic arch obstruction. The status of the atrial septum is also carefully monitored, determining the timing of atrial septectomy. There are no certain criteria for intervention on the atrial septum. The decision is made based on the balance between the patient's clinical status and the pressure gradient across the intra-atrial communication. A mild-to-moderate pressure gradient up to 8-10 mmHg is acceptable as long as the patient is thriving. Prompt intervention is required when the patient has symptoms of left atrial hypertension, such as poor feeding, an increased respiratory rate, and a decrease in arterial saturation.

- Ventricular function and atrioventricular valve regurgitation are of great importance during the follow-up echocardiographic examination. Depressed ventricular function can be a result of retrograde aortic arch obstruction in the setting of minimal or no antegrade aortic flow, or stenosis of a reverse BT shunt. The patency and flow pattern of the reverse BT shunt is documented. Anti-platelet and/or anti-coagulation is indicated only if the patient has a reverse BT shunt. Anti-platelet therapy is initiated if the patient has a stent across the atrial septum. Pre-stage II cardiac catheterization is electively performed at 3-4 months of age unless intervention for the atrial septum is necessary at an earlier age.

3. Conclusion

- Thus, it remains of outmost importance to determine the outcome of children treated with hybrid procedure in comparison to that of the Norwood procedure to establish the best treatment strategy for this most vulnerable population.

- This can only be achieved by performing a large multicenter randomized trial, in which the effect size can be based on the results of our study and in which all potential risk factors such as preoperative delayed brain development and brain injury, intra- and postoperative factors and socio-demographic factors need to be considered.

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