Hybrid cardiac surgery for hypoplastic left heart syndrome

Although children with congenital heart disease have been managed jointly by paediatric cardiologists and cardiac surgeons as a collaborative team, competition was introduced between these teams in the 1980s with the advent of interventional techniques. Although interventional treatment for pulmonary valve stenosis and patent arterial duct was accepted by the surgeons, treatment of other defects was not. It became clear that many interventional techniques, rather than replacing surgery, actually facilitated it and so these were accepted. Interventional procedures were performed either before or after surgery as a complementary procedure. A natural development was surgeons and cardiologists working together in the catheter laboratory or in the theatre. It is this team effort which is given the name of 'hybrid' surgery. One area in which this has been an important development is in the treatment strategy for the hypoplastic left heart syndrome.

Hypoplastic left heart syndrome

Hypoplastic left heart syndrome is a complex congenital heart defect, with a mortality in excess of 90% in the first month of life if left untreated. Although cardiac transplantation is a treatment option, in most countries this is not realistic. Since the 1980s, staged palliative treatment with Norwood operations has been accepted. The first stage of the surgery in the neonatal period involves reconstruction of the aortic arch to widen the hypoplastic ascending aorta using the native main pulmonary artery for the anastomosis. The pulmonary arterial blood flow is established by constructing a Blalock–Taussig shunt and an atrial septectomy is performed to achieve unrestrictive blood flow from the left to the right atrium. The second stage, at about 6 months of age, involves taking down the Blalock–Taussig shunt and constructing a Glenn shunt (an anastomosis between the superior vena cava and the right pulmonary artery). The third stage of the strategy, at 18 months to 3 years of age, involves completion of the total cavo-pulmonary connection (Fontan circulation).

Rationale for hybrid cardiac surgery

There have been marked improvements in survival of the first stage Norwood operation for hypoplastic left heart syndrome in recent years. The average mortality for the first stage is about 20%, for the second stage less than 5% and for the third stage less than 2%. Nevertheless, the operation requires prolonged cardiopulmonary bypass, which, in the neonatal period especially in the setting of hypoxaemia and low diastolic pressures, may be associated with poor neurological development and higher risks.

The risk factors for mortality and complications include initial presentation with cardiogenic shock, birth weight of less than 2.5 kg, prematurity (34 weeks' gestational age), less than a 2 mm ascending aorta, poor ventricular function, tricuspid regurgitation, intact or restrictive atrial septum, additional cardiac anomalies, or additional severe extracardiac genetic malformations (Gaynor et al, 2002). In these babies, the first stage Norwood operation may have a mortality of 20–50% and possibly higher.

An alternative method, that could palliate these babies for a few months, may allow their growth and so they may tolerate the effects of cardiopulmonary bypass better than in the neonatal period, hence the need to develop strategies to lower this mortality.

In 1990s, in the hypoplastic left heart syndrome, bilateral branch pulmonary artery banding combined with stenting of the arterial duct was performed as a bridge to cardiac transplantation (Gibbs et al, 1993). Banding of the branch pulmonary arteries was performed in the theatres and stenting of the arterial duct in the catheterization laboratory. This was a novel collaborative approach rather than a true hybrid, but can be considered as a precursor to the current hybrid surgery. Although the technique fell out of favour because of poor survival, it showed its technical feasibility. Some centres revived the technique and started to obtain improved results, but with the aim of performing a later Norwood operation rather than cardiac transplantation (Akinturk et al, 2002, 2007).

The approach was modified further into a truly hybrid procedure by performing ductal stenting via a sternotomy through a sheath in the main pulmonary artery, thus avoiding percutaneous access, arrhythmias, and valvar regurgitation associated with percutaneous procedures (Bacha et al, 2006). Ultimately, this strategy could potentially lead to a percutaneous Fontan completion, with one major operation performed on cardiopulmonary bypass flanked by two interventional procedures (Galantowicz and Cheatham, 2005).

Technique of hybrid procedure

The hybrid procedure is performed under general anaesthesia in the catheter laboratory. If performed in the theatre, adequate quality fluoroscopy is needed during stenting of the arterial duct. The surgeon's task is to perform a midline sternotomy and perform banding of both the pulmonary artery branches. Banding should result in a drop of the oxygen saturation levels to approximately 80% and there should be a small rise in the sytemic pressure. After completing the banding, the surgeon makes a small purse-string suture in the main pulmonary artery just above the pulmonary valve. A 6 French short vascular sheath and dilator are inserted through this and the dilator is removed while the sheath is held in place.

The cardiologist's task is to stent the arterial duct by performing angiography through the side-arm of the sheath to define the anatomy of the arterial duct, assess the distal and transverse aortic arch and measure the maximum diameter of the arterial duct, the descending aorta and the length of the duct to just beyond the coarctation site. A stent premounted on a balloon or a self-expanding stent of the appropriate diameter and length is selected on the basis of the measurements, aiming to cover the whole length of the duct including the site

of coarctation. On most occasions, the stent needs to be 18–24 mm long and dilated to 6–8 mm in diameter. Prostaglandin infusion is stopped just before the stent is deployed. The surgeon then closes the sternotomy and the patient is transferred to the intensive care unit with a view to early extubation. It is rare for inotropes to be needed if preoperatively the baby is stable.

After the hybrid procedure, a combined comprehensive first and second stage Norwood operation is required 3–6 months later. This involves removal of the bands on the pulmonary arteries, removal of the stent in the arterial duct, extensive repair of the hypoplastic aortic arch and construction of a Glenn shunt and, if needed, an atrial septectomy.

The hybrid surgery may reduce the operative risk in babies of low birth weight such as those less than 2.5 kg. However, the mortality risk is transferred from the conventional first stage Norwood to the combined first and second stage Norwood operation after a hybrid procedure, because it is technically more demanding. Nevertheless, in the group of babies for whom the hybrid procedure is intended, the net effect should be improved survival.

The group from Giessen have used the strategy developed by Gibbs et al (1993). Akinturk et al (2007) reported on 58 newborns in whom modified hybrid surgery (stenting the duct in the catheterization laboratory, followed a few days later by surgical bilateral pulmonary artery banding) was performed. The combined first and second stage surgery was performed at a median age of 4.8 months and total cavopulmonary connection (the third stage) at a median of 3.1 years. Nine of the patients were listed for heart transplantation. Overall, 8 of 58 patients (13.8%) treated by the transcatheter-surgical combined approach died.

Bacha et al (2006) reported their experience of the learning phase in 14 high-risk newborns in whom a truly hybrid approach was adopted. The presence of various factors would have excluded most of these babies from undergoing the conventional Norwood operation. The hospital mortality was 22%. There were two interstage deaths and two of the eight patients, who had second stage surgery, died.

Galantowicz and Cheatham (2005) reported their experience of the evolution of the hybrid surgery in 34 patients. Initial hybrid surgery from 2001 to 2004 included 29 newborns of 1.8–4.2 kg. There were five (17%) hospital deaths and three (10%) interstage deaths. Comprehensive second stage surgery was performed in 18 patients with four (22%) deaths and transcatheter Fontan completion was achieved in five patients.

Calderone et al (2007) reported their hybrid experience in 18 patients, 11 of whom were as an alternative to Norwood operation. There were two (18%) deaths. Comprehensive first and second stage surgery was performed in nine patients with one death. Among Norwood-alternative survivors, intubation times and lengths of stay in the intensive care unit and in the hospital tended to be shorter than Norwood survivors. These are encouraging results for a high-risk group of babies.

Other indications

During development of the hybrid procedure for the hypoplastic left heart syndrome, other important indications have emerged. Newborn babies with critical aortic stenosis but with a borderline left ventricle may form a small group of patients that often cause difficulties of management. Conventional treatment involves balloon dilation or surgical aortic valvotomy. Afterwards, if the left ventricle cannot cope, then first stage Norwood operation is performed, but with high risks. The disadvantage of this approach is that if the left ventricle recovers, then it may be possible to take down the Norwood operation and revert to a two-ventricle circulation, but the patient is subjected to high-risk strategies on several occasions.

With the hybrid approach, the duct is stented after pulmonary artery banding and at the same operation, a sheath is inserted into the ascending aorta. Through this, a guidewire can be passed across the aortic valve and using an appropriate sized balloon, dilation of the valve is performed to decompress the left ventricle. This strategy may allow time for the left ventricle to develop. If it increases in size and its function improves, it is easier to take down this circulation and convert it to a two-ventricle circulation.

Conclusions

Hybrid cardiac surgery in hypoplastic left heart syndrome is an innovative procedure which could offer an alternative management strategy. This technique has been used in a very limited number of patients so long-term evaluation is needed. **BJHM**

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Akinturk H, Michel-Behnke I, Valeske K et al (2002) Stenting of the arterial duct and banding of the pulmonary arteries. Basis for combined Norwood stage 1 and 2 repair in hypoplastic left heart. *Circulation* **105:** 1099–103

Akinturk H, Michel-Behnke I, Valeske K et al (2007) Hybrid transcatheter-surgical palliation: basis for univentricular or biventricular repair: the Giessen experience. *Pediatr Cardiol* **28:** 79–87

Bacha EA, Daves S, Hardin J et al (2006) Single-ventricle palliation for high-risk neonates: The emergence of an alternative hybrid stage I strategy. *J Thorac Cardiovasc Surg* **131:** 163–71

Caldarone CA, Benson L, Holtby H, Li J, Redington AN, Van Arsdell GS (2007) Initial experience with hybrid palliation for neonates with single-ventricle physiology. *Ann Thorac Surg* **84:** 1294–300

Galantowicz M, Cheatham JP (2005) Lessons learned from the development of a new hybrid strategy for the management of hypoplastic left heart syndrome. *Pediatr Cardiol* **26:** 190–9

Gaynor JW, Mahle WT, Cohen MI et al (2002) Risk factors for mortality after the Norwood procedure. *Eur J Cardio-thorac Surg* **22:** 82–9

Gibbs JL, Wren C, Watterson KG, Hunter S, Hamilton JR (1993) 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. *Br Heart J* **69:** 551–5

KEY POINTS

- Hybrid surgery for hypoplastic left heart syndrome is new and is being performed in only a few centres.
- Hybrid surgery may be an alternative to the conventional Norwood stage I operation in babies with a low birth weight or babies of normal birth weight but with preoperative neurological insult.
- After a hybrid procedure, the second stage of removing the duct stent, the pulmonary artery bands, repairing the aortic arch and constructing a Glenn shunt is more complex and is associated with higher mortality. However, the net effect may be improved survival.
- In babies with critical aortic stenosis but with borderline left ventricle, a hybrid procedure, performing aortic balloon dilation, bilateral pulmonary artery banding and stenting of the arterial duct, allows time for the left ventricle to grow and may ultimately lead to a two-ventricle circulation.