Hybrid Procedures in Congenital Heart Disease

a report by

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Surgery and interventional cardiology have developed greatly during the last decades. For any congenital heart disease a surgical procedure is possible to obtain complete correction or acceptable palliation. Progress in interventional cardiology opens up new directions for the treatment of simple heart defects. Today many simple lesions are suitable for correction in the catheterisation laboratory. Since the beginning of this century long-term follow-up studies and developmental surveys have been published.[1-3] Paediatric patients with congenital heart disease, particularly those who need an intervention in the perinatal period or repeated surgeries, demonstrate different kinds of disability at school age. Although cardiopulmonary bypass is a huge advance, it also has detrimental effects.

Confronted with these observations, new strategies have been developed to shorten cardiopulmonary bypass or to avoid particularly risky surgical approaches. These new strategies are called hybrid procedures because they emerge from the collaborative efforts of cardiac surgeons and interventional cardiologists in an approach to managing congenital heart disease. The idea is to develop therapies that offer the advantages of surgery and interventional techniques in the same setting.[4,5]

Hybrid therapy is not a simple first step. It is a change in approach for surgeons and cardiologists. Both have to admit that joining the two techniques would possibly allow better immediate and long-term results. It also means that cardiologists can enter the operating theatre for other reasons than only to look at the transoesophageal echocardiography (TEE) and that surgeons can enter the catheterisation laboratory not only for emergency purposes. In other words, surgeons and cardiologists have to discuss and collaborate in a positive fashion.

Indications for hybrid therapy are expanding, the most frequent being intraoperative stenting, perventricular ventricular septal defect (VSD) closure, occlusion of vascular structures during surgical repair and percutaneous palliation for single ventricle physiology (such as hypoplastic left heart syndrome [HLHS]). Ideally, these interventions are carried out in a hybrid suite where operating room and catheterisation laboratory facilities are brought together.

Indications, Techniques and Experience

Hypoplastic Left Heart Syndrome

HLHS associates hypoplastic left ventricle (unsuitable for systemic perfusion), aortic valve stenosis and hypoplasia of the ascending aorta. Since the 1980s a three-step surgical palliation can be proposed for these infants. The first step is Norwood stage I, whereby reconstruction of the aortic arch with anastomosis to the pulmonary trunk is performed. At the same time pulmonary blood flow is provided with a modified Blalock-Taussig shunt or a shunt between the right ventricle and pulmonary arteries. The second step, Norwood stage II, is a bidirectional Glenn anastomosis. The third step is the completion of a Fontan circulation, with a connection between inferior vena cava and pulmonary arteries.

HLHS is a typical congenital cardiac malformation with the first surgical palliation occurring in the first days after birth, in which high surgical mortality and high long-term morbidity have been reported. Some authors have demonstrated a striking association for these infants between duration of deep hypothermic circulatory arrest performed during this surgical procedure and developmental outcome at school age.[1]

In an attempt to change the traditional surgical strategy, hybrid therapy combines stent implantation in the ductus arteriosus by the interventional cardiologist and a surgical band for both pulmonary arteries, in order to maintain systemic perfusion and restrict pulmonary blood flow to the lungs. Usually, this procedure is completed with a unique step through a median sternotomy without the need for cardiopulmonary bypass.[6,7] A few months later, with a larger infant, a comprehensive stage I and II Norwood is performed. The main advantage of this technique is to postpone the Norwood I procedure to a later age, allowing a decreasing global risk of this surgical procedure in the neonatal period and avoiding deep hypothermic circulatory arrest. Reconstruction of the aortic arch is then carried out in a larger infant, with a probable decreased risk of brain insult than in the neonatal period. The second advantage is to reduce the number of interventions with cardiopulmonary bypass.

As for any new surgical or interventional procedure there is a learning curve. Initial problems have been identified and can be overcome. For example, tightening of the pulmonary bands (too tight or too loose) and stenting of the ductus arteriosus when aortic coarctation is present have been addressed as a result of published expertise from leading centres.[8] Although long-term follow-up data are still not known, short- and medium-term follow-up is impressive and comparable to surgical results.[7,9–12]

Avoiding cardiopulmonary bypass with hybrid therapy does not mean that these patients are less prone to interstage problems and mortality.

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These infants remain high-risk patients. Strict ambulatory follow-up is mandatory to anticipate potential complications. For example, insufficient stenting of the ductus arteriosus, obstruction of one or even both pulmonary arteries and dysfunction of the right ventricle in systemic position are potential acute problems that can occur at any time between the first and second intervention.[11]

Isolated Ventricular Septal Defect or in Association with Other Congenital Malformations

VSD is one of the most frequent congenital heart diseases. It can be isolate, multiple or associated with more complex congenital malformation such as double outlet right ventricle or transposition of the great arteries. Most of the VSDs are localised in the perimembranous area of the interventricular septum, and only 20% are in the muscular part. Specific devices have been developed for interventional closure of perimembranous and muscular VSDs. Interventional closure of muscular VSDs is one of the most challenging percutaneous interventions because of high anatomical variability, different thickness of the muscular septum and the presence of trabeculations on the right ventricular side. In addition, infants with symptomatic VSD demonstrate signs of congestive heart failure and failure to thrive and are potentially too unstable for a long catheterisation procedure. Thus, most of them are not suitable for percutaneous closure. On the other hand, surgical muscular VSD closure could be difficult for the same anatomical reason (localisation of the VSD in the muscular septum and visibility of this area in an arrested heart).

To overcome these problems and find a better strategy, a hybrid therapy has been developed. The procedure is performed under TEE guidance and after median sternotomy or mini-sternotomy. A purse ring suture is placed on the right ventricular free wall opposite to the VSD, allowing the VSD to be crossed directly with a needle. An appropriately sized sheath is then advanced through the VSD. Before the device is released the location of the device is checked with TEE. The same procedure could be performed, for example, in the case of repair of transposition of great arteries with VSD or repair of a double outlet ventricle. The advantages are the shortened time of cardiopulmonary bypass and simplification of an already complex surgery. In cases of multiple muscular VSD (so-called Swiss cheese presentation) the procedure can be repeated, allowing every significant defect to be closed.[12-16]

Overall results are excellent and the avoidance of cardiopulmonary bypass for these sick infants is a great advance.

Intraoperative Stenting of Pulmonary Arteries

Congenital heart disease with right ventricular obstruction is frequently associated with stenotic or hypoplastic peripheral pulmonary arterial branches. Recurrent stenosis of the pulmonary arteries is usual and impairs normalisation of right ventricular pressure. Redo surgery has the disadvantage that dissection and recognition of structures are made more difficult. On the other hand, percutaneous intervention is possible with balloon angioplasty (standard balloon, high-pressure balloon or cutting balloon) or with stent deployment. The main issues in these cases are either vascular access or reaching the lesion in a satisfactory fashion for the intervention. Tortuous pulmonary branches and cardiac conduit between right ventricle and pulmonary artery are some of the obstacles for cardiologists.

Again, hybrid therapy offers possibilities to overcome these challenging situations. Usually, the procedure is monitored by fluoroscopy and performed at the time of another planned surgery, such as a conduit change. According to the type of lesions, the surgeon either provides access to the lesion (through the main artery purse ring suture, for example) or the main pulmonary is opened and direct stenting of pulmonary arteries achieved under direct visualisation.[12,17-19]

In these complex patients with different levels of right ventricular outflow tract obstruction, any effort to reduce the number of interventions and their associated morbidity is of great help. In addition, improving in fewer stages the obstruction and so decreasing right ventricular pressure will hopefully contribute to improved long-term outcome.

Occlusions of Vascular or Surgical Structures During Surgery

In some particular settings, pulmonary perfusion is dependent on abnormal vascular structure or surgical shunts. Surgical ligation of these alternative ways of pulmonary flow could be difficult and time consuming for the surgeon. A stepped approach with closure of devices of these structures followed by surgery is almost impossible without a significant decrease of arterial saturation, leading to haemodynamic instability of the patient.

With a hybrid procedure, occlusion of vascular or surgical structures is feasible during surgery. Intervention is performed under fluoroscopy or TEE guidance. According to anatomical location, size and length of the shunt, the cardiologist will choose the most suitable type of device (coils or vascular occluder) to be used for the intervention. Once obstruction of the shunt is obtained, cardiopulmonary bypass is initiated and the surgical procedure performed.

In these cases, collaborative planning between surgical and cardiological teams is mandatory before surgery in order to obtain the best result for the patient. Again, the main goal of this approach is to reduce duration of surgery and cardiopulmonary bypass.

Issues, Debates and Conflicts in Hybrid Procedures

Increasing numbers of publications have addressed the problems raised by hybrid procedures. The debate is particularly exacerbated when considering HLHS or similar pathologies.[20,21] Based on mortality, length of stay in intensive care unit and hospital, and short- and medium-term survival, many publications have focused on whether hybrid therapy offers the same results as a standard surgical approach. In these articles, hybrid therapy is offered, most of the time, to patients facing high-risk surgery, and standard surgery is offered to those facing simpler surgeries.[22,23] Most of the published experience comes from leading centres where a high amount of surgery is performed. Their experience, however, may not reflect that of other centres worldwide. Indeed, depending on the surgical team and intensive care unit experience, differences in mortality or other short-term issue can vary dramatically. In our opinion the main issue is not whether hybrid therapy is performed as well as a standard surgical approach but whether changing our surgical strategy for a less aggressive strategy will change long-term neuro-developmental outcomes.

Conclusion

Congenital heart disease represents a large panel of different diseases, with different problematic clinical and follow-up outcomes. In this area, the development of hybrid procedures is a promising tool.

This new way of thinking allows new strategies of palliation or correction to be offered for simple and complex congenital heart diseases. For some rare cases it may be a unique way of allowing an acceptable result for the patient. Although many of these types of intervention are already standardised, many will never be. Some cases are so unusual that the applied strategy would never be comparable with any 'gold standard' surgery. The main issue of implementing a hybrid therapy programme is a trend in avoiding surgical insults to infants and children with congenital heart disease. This has been carried out by creating or inventing new procedure strategies. This will be acceptable only if these new strategies obtain results as good as the previous ones and demonstrate a beneficial effect on the long-term follow-up of patients. This question is still unanswered at this point.

Finally, in our opinion, hybrid therapy is an alternative approach for dealing with congenital heart disease and a great opportunity for surgical and medical teams to think and plan together to overcome the limitations of the usual surgical or interventional procedures. ■

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Associated Papers

New Developments in the Treatment of Hypoplastic Left Heart Syndrome

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In the current decade, the prognosis of newborns with hypoplastic left heart syndrome, previously considered a uniformly fatal condition, has dramatically improved through refinement of rapidly evolving treatment strategies. These strategies include various modifications of staged surgical reconstruction, orthotopic heart transplantation and hybrid palliation using ductal stenting and bilateral pulmonary artery banding. The variety of treatment approaches are based on different surgical philosophies, and each approach has its unique advantages and disadvantages. Nonetheless, multiple experienced centres have reported improved outcomes in each one of those modalities.

The purpose of this review is to outline recent developments in the array of currently available management strategies for neonates with hypoplastic left heart syndrome. Because the vast majority of deaths in this patient population occur within the first months of life, the focus of the review will be evaluation of the impact of these management strategies on survival in the neonatal and infant periods. ■

Improving the Outcome of High-risk Neonates with Hypoplastic Left Heart Syndrome: Hybrid Procedure or Conventional Surgical Palliation?

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Despite significant progress, surgical outcome for high-risk patients with hypoplastic left heart syndrome (HLHS) remains suboptimal. The hybrid palliation lessens the initial operative insult and is expected to improve overall survival; however, the outcome of this management sequence is unknown. In a retrospective review of all high-risk neonates undergoing initial palliation for HLHS either by hybrid or stage I Norwood procedure at a single institution between January 2001 and December 2006, the two strategies were compared using survival after stage II as the end-point for outcome. The cohort included 33 patients (14 hybrid and 19 Norwood). Patients undergoing hybrid palliation had a lower pre-operative pH, higher incidence of organ dysfunction and fewer associated cardiac anomalies. Hospital mortality and interstage mortality was 7/33 (21%) and 6/26 (23%) for the entire cohort, without significant differences between the hybrid and the conventional Norwood strategies. Although the hybrid approach reduces the initial surgical insult, important interstage mortality and ongoing morbidity result in no differences in survival compared with conventional surgical palliation. ■