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Hybrid Procedure in Neonatal Critical Aortic Stenosis and Borderline Left Heart: Buying Time for Left Heart Growth

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1. Introduction

Infants with “borderline” left ventricles due to congenital aortic valve stenosis may be critically ill soon after birth since the left ventricle may be unable to sustain the complete systemic circulation.

In these hearts, the stroke volume of the left ventricle is markedly decreased because of diminutive left ventricular internal dimensions. As the arterial duct closes in the first hours after birth, the left ventricle becomes responsible for generating the full systemic output. If the left heart is of borderline dimensions, this may lead to shock and even death.

However, postnatal loading and growth conditions of the left ventricle differ significantly from foetal life and in so doing influence the postnatal course. Left ventricular growth potential remains and is likely to be stimulated by these physiological changes. Some may occur rapidly following birth whilst others may take weeks to months.

Infants with borderline left ventricles present the physician with a unique dilemma: is this child able to tolerate a biventricular circulation or should one rather embark on a univentricular strategy? In recent years, experience with the hybrid procedure for hypoplastic left heart syndrome showed that indications may be extended to patients with critical aortic stenosis and borderline left heart. A hybrid procedure typically consists of surgical bilateral banding of the branch pulmonary arteries in combination with placement of a stent in the ductus arteriosus after balloon angioplasty of the stenosed aortic valve (Fig 1). As a result, a parallel circulation is created and the right heart can support the left heart to maintain adequate systemic output. This “buys time” to observe the growth potential of the left heart and allows the physician more room to make an informed decision regarding univentricular or biventricular long-term strategy.

In this chapter we describe the background, possibilities, technical aspects and results of this strategy in neonates with critical aortic stenosis and borderline left heart.

2. Background of borderline left ventricle

It is not entirely clear exactly what is referred to when using the term “borderline left heart”. Some refer to quantitative and others to qualitative characteristics. The fact remains that it is

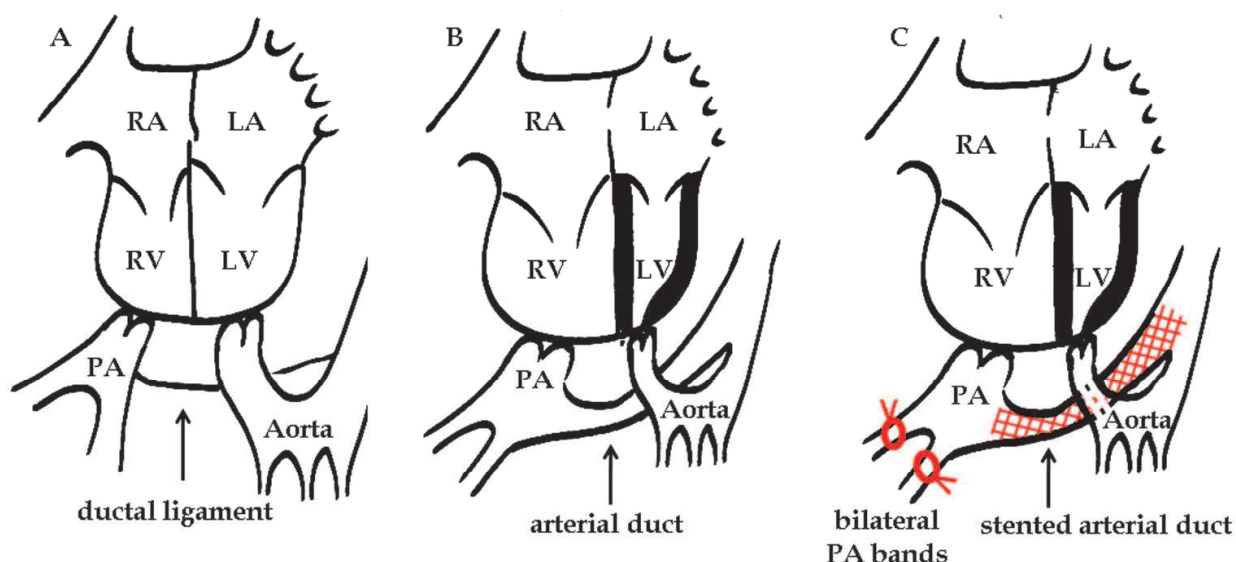


Fig. 1. Schematic drawing of a normal neonatal heart with constriction of the arterial duct (A), a borderline left heart with aortic stenosis, left ventricular hypertrophy, open arterial duct (B) and a borderline left heart after the hybrid procedure with bilateral banding of the pulmonary arteries (PA), stent in the arterial duct (C). RA: right atrium, RV: right ventricle, LA: left atrium, LV: left ventricle.

difficult to define and that a more comprehensive approach is required when facing a left ventricle that is smaller than normal.

In essence, a normal left ventricle should be able to provide satisfactory forward flow by means of generating and maintaining sufficient stroke volume and aortic pressure as well as being compliant enough to keep left atrial and pulmonary venous pressures low. A borderline left ventricle may thus be recognised by being smaller than normal (quantitative) combined with functional (qualitative) inability to provide either forward flow and/or leading to excessive pulmonary venous congestion.

In a comprehensive review Corno identified risk factors such as quantitative measurements of left ventricular enddiastolic dimensions, aortic and mitral valve orifices indexed for body surface area (Corno, 2005). Associated abnormalities such as younger age, endocardial fibroelastosis, non-compaction of the left ventricle, aortic coarctation, mitral valve abnormalities and pulmonary hypertension are also important risk factors to be considered (Hickey et al., 2007). In the end, both objective measurements and clinical judgment need to be taken into account when deciding what is considered a borderline left ventricle. This is important, especially since the decision whether to proceed to univentricular or biventricular repair, will be influenced by how borderline left heart is defined.

3. Pathophysiological and clinical correlates

3.1 Deleterious factors in borderline left ventricle with aortic stenosis

Prenatally, the foetal left ventricle receives 40% of the combined cardiac output: 10% of the cardiac output enters the left atrium via the pulmonary veins whilst 30% of the combined

cardiac output is received due to right-to-left shunting over the foramen ovale. Good ventricular filling is a prerequisite for adequate cardiac growth. In the normal heart, the neonatal transition from a foetal parallel circulation to a circulation in series requires the left ventricle to generate sufficient aortic pressure and output. Simultaneously, left atrial pressure has to remain low and pulmonary artery pressure has to decrease to normal levels within weeks. In infants with aortic stenosis and borderline left ventricle, symptoms occur during or after the transition from the parallel foetal circulation to the circulation in series when the arterial duct closes.

3.1.1 Foetus

In infants with severe left ventricular outflow tract obstruction such as critical aortic stenosis, left ventricular pressure overload results in significant left ventricular hypertrophy while pressure overload and subendocardial ischemia induce endocardial fibroelastosis. Myocardial hypertrophy and endocardial fibroelastosis result in decreased ventricular volumes and myocardial compliance. The limited blood flow results in reduction of growth of the left ventricular cavity. The foramen ovale may also be restrictive, thereby further limiting left ventricular filling and subsequently growth by up to 75%. Similarly, reduced mitral inflow as a result of mitral valve abnormalities will further compromise foetal left ventricular growth.

Antenatally, the circulation of foetuses with borderline left hearts does not lead to problems. Adequate systemic perfusion is maintained despite the small size of the left ventricle due to increased right-to-left shunting over the ductus arteriosus. Therefore, overall foetal development and somatic growth are usually not jeopardized by a borderline left heart.

3.1.2 After birth

In the delivery room, the newborn with borderline left heart (Fig 2) will be hemodynamically stable as long as the arterial duct remains open. Patients with a severely restrictive, hypoplastic left ventricle and absence of atrial left-to-right shunting, may present with symptoms appearing immediately after birth. However, the latter subset of patients is not the topic of this chapter.

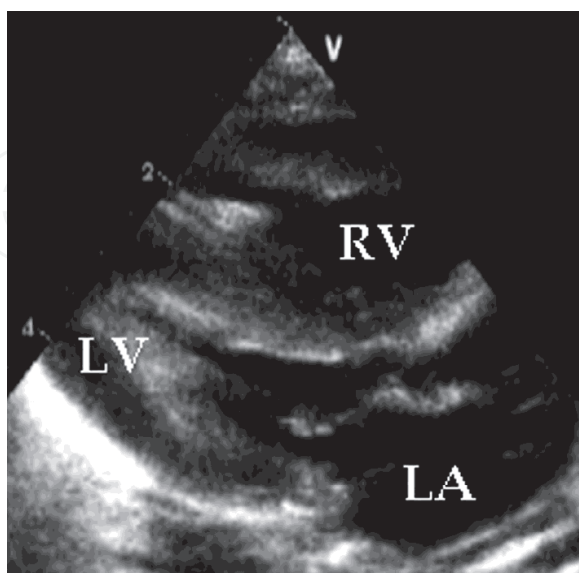


Fig. 2. Echocardiographic parasternal long axis view of borderline left heart in a neonate with aortic stenosis, left ventricular hypertrophy, and endocardial fibroelastosis.

3.1.3 Circulatory failure in the first postnatal days

Acute failure occurs when closure of the arterial duct results in low systemic output due to insufficient left heart size and function. As indicated in the definition of borderline left heart, failure may be due to an inability to generate proper flow and pressure (forward failure) with or without left atrial hypertension and pulmonary venous congestion (retrograde failure). The extent of systemic under perfusion is variable and includes cold extremities, pallor and decreased peripheral pulses. When systemic output is further compromised, hypoperfusion of internal organs results in progressive metabolic acidosis. In the most severe cases, closure of the arterial duct rapidly results in severe arterial hypotension, shock and death. Associated lesions, such as coarctation of the aorta, may even further compromise circulation.

3.1.4 Chronic circulatory failure

Cardiac failure can occur in weeks to months after birth due to pulmonary hypertension. After birth, pulmonary venous return to the left atrium is greater than before resulting in increased left ventricular preload. If the left ventricular cavity is too small or if left ventricular filling is restricted (especially in the presence of mitral valve stenosis or impaired left ventricular compliance), left-to-right shunting over the interatrial septum increases. When interatrial communication as well as left ventricular filling are restrictive, it will lead to left atrial congestion, retrograde pulmonary venous and arterial hypertension and/or pulmonary oedema.

Severe pulmonary hypertension can manifest several months after birth with low output cardiac failure. In these patients with severe pulmonary hypertension, compression of the small left ventricle may occur due to septal deviation as a result of the markedly enlarged hypertensive right ventricle (Smallhorn, 2009), thereby further compromising left ventricular function and growth.

3.2 Favourable factors promoting postnatal left ventricular growth

Following birth, several circulatory factors change significantly compared to foetal conditions. These changes in preload, afterload and growth factors promote growth of the left ventricle.

3.2.1 Increased preload

After birth, pulmonary flow will significantly increase with augmented pulmonary venous return to the left heart. Preload to the left ventricle will therefore always increase after birth, giving rise to the phenomenon of “unfolding” of the left ventricle (Fig. 3). This is often observed where, within minutes after birth, the left ventricle enlarges in infants where foetal ultrasound demonstrated asymmetric four chamber views with small left ventricles. The overall increased flow also results in catch-up growth. Such catch-up growth, however, may take weeks or months to occur.

3.2.2 Reduced afterload

The neonate with critical aortic stenosis will typically be offered percutaneous balloon angioplasty of the aortic valve, thereby decreasing the pressure overload of the left ventricle. This will eventually result in reduction of hypertrophy, increased left ventricular volumes and better compliance with improved filling. These changes may take weeks or even

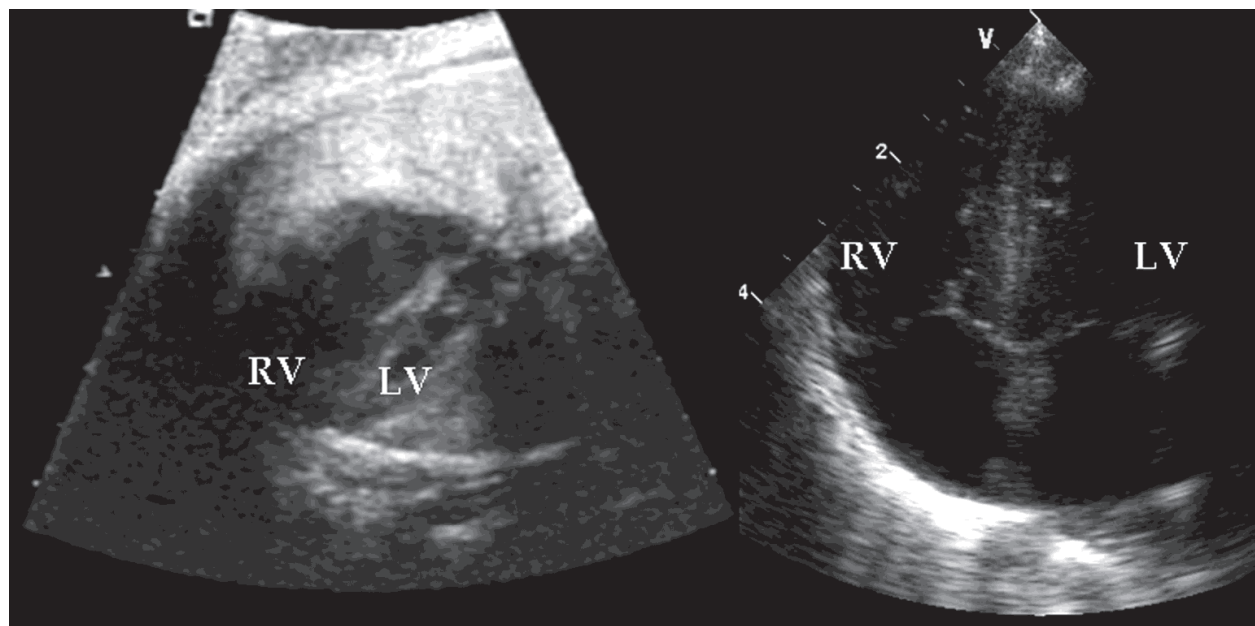


Fig. 3. Four chamber echocardiographic views demonstrating small left ventricle (due to restrictive atrial flow) in foetus (left) and unfolding of the left ventricle 4 hours after birth (right).

months before a significant clinical effect can be observed. In a series of 53 neonates undergoing aortic valve balloon angioplasty, significant growth of aortic valve annulus, aortic sinus, and left ventricular dimensions z-score have been demonstrated (Han et al., 2007).

3.2.3 Growth factors

Insulin and insulin-like growth factor I levels are increased in foetuses of mothers with pre-existing or gestational diabetes. Circulating insulin-like growth factor I in the foetus is a very potent cardiac growth factor, inducing cardiac hypertrophy and hyperplasia (Hayati et al., 2004). Furthermore, insulin-like growth factor I is known to mediate many of the anabolic effects of growth hormone on the heart (Hayati et al., 2004). After birth, insulin levels will drop to normal ranges, removing the pathological stimulus for cardiac hypertrophy. Similarly, increased maternal cortisol levels during late pregnancy induce foetal cardiac hypertrophy (Reini et al., 2008). Again, the effect of postnatal remodelling and the regression of hypertrophy can only be observed after some weeks to months.

4. Treatment options in patients with borderline left ventricle

4.1 Decision making in the neonatal phase

A treatment strategy has to be chosen in the neonatal phase, and previously implied an early choice for either a biventricular or univentricular strategy. On the extreme ends of the spectrum of decreased left heart size and aortic stenosis, the decision concerning the definitive strategy can readily be made in the neonatal phase (Fig. 4). On the one end, hypoplastic left heart syndrome will be palliated by a univentricular strategy. On the other end of the spectrum with a near-normal sized left ventricle, a biventricular repair is the rule. However, in true borderline cases, this early decision proves to be difficult due to lack of guidelines with clear -cut predictability.

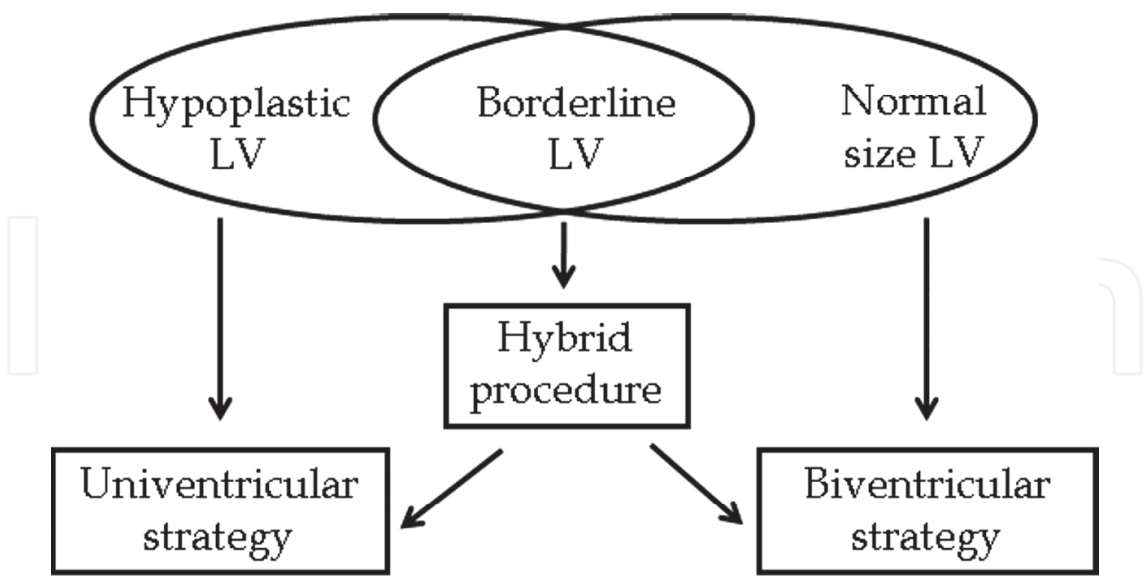


Fig. 4. Options in decision-making in the neonatal phase.

Several scoring systems such as the Rhodes score (Rhodes et al., 1991), Colan score (Colan et al., 2006) and the Univentricular Repair Survival Advantage Tool (Hickey et al., 2007) exist to predict the optimal type of repair. These scoring systems use several left ventricular parameters in an equation, the result of which above or below a given cut-off value generates the predicted most favourable treatment strategy. The Rhodes score involves the variables of body surface area, indexed aortic root size, left ventricle to heart long axis ratio and indexed mitral valve area (Rhodes et al., 1991). In the Colan score, the equation is based on body surface area, aortic annulus z-score, left ventricle to heart long axis ratio and degree of endocardial fibroelastosis (Colan et al., 2006). The Univentricular Repair Survival Advantage Tool is based on morphologic, functional and pathologic information; the exact equation is not published but the score can be calculated at the freely accessible Congenital Heart Surgeons’ Society website www.chss.org (Hickey et al., 2007). A recent small series, however, demonstrated that the results of the aforementioned scoring systems never provided unanimous recommendations regarding the optimal type of repair for the individual patient with borderline left heart (Davis et al., 2011). This important observation emphasizes the true “borderline” characteristics of this disease entity. Another issue to consider is the fact that body surface area, aortic and mitral valve diameters may change very little in the first 2 – 3 weeks, whilst left ventricular enddiastolic dimensions and compliance may change markedly; these are not taken into account in scoring systems. The main issue still remains – what is best: living with a restrictive biventricular circulation or a good univentricular circulation?

4.2 Univentricular strategy

The univentricular strategy typically implies three consecutive surgical procedures: reconstruction of the aortic arch and main pulmonary artery (Damus-Kaye-Stansel & Norwood I) with atrial septectomy and creation of a shunt to the pulmonary artery in the neonatal phase, followed approximately four to six months later by creation of a partial

cavo-pulmonary connection (Glenn shunt) and finally completion of the total cavopulmonary connection at the age of two to four years (Fontan circuit). When a univentricular strategy is followed from the neonatal phase, conversion to biventricular physiology may no longer be an option or may be extremely difficult.

Risk factors for increased mortality after univentricular repair consist of tricuspid valve regurgitation, presence of a large ventricular septal defect and smaller indexed dominant ventricular length (Hickey et al., 2007). Despite major improvement in survival rates of the Norwood stage I procedure, infants undergoing this procedure still face a mortality risk of 10-30%, depending on risk factors and the experience level of the team (Bacha, 2006, Stasik et al., 2006). In a recent study, survival rate of the Norwood stage I procedure exceeded 90% in an experienced unit after the initial learning curve (Rychik et al., 2010).

4.3 Biventricular strategy

The biventricular approach consists of treatment of left ventricular outflow obstruction, typically by balloon angioplasty of the aortic valve in a neonate. Surgical aortic valvotomy or the Ross-Konno procedure are alternative options usually performed at a later stage. In case of concomitant coarctation of the aorta, coarctectomy should be performed.

When a biventricular strategy is chosen early but eventually fails, usually due to low cardiac output or pulmonary hypertension, this can lead to death of the patient; otherwise conversion to a univentricular circulation is associated with markedly increased risk (Davis et al., 2011, Pizarro et al., 2009). Decreased survival rates after biventricular repair are found in patients with small left ventricular outflow tract size, small aortic arch size, presence of endocardial fibroelastosis or left ventricular dysfunction (Hickey et al., 2007). Inappropriate pursuit of biventricular repair occurs more often and entails poorer survival rates as compared to inappropriate univentricular repair (Hickey et al., 2007).

4.4 Strategies in the presence of indecision

The immediate decision can be delayed by prolonged infusion of prostaglandins. This will give the clinician one to two weeks to decide by allowing time for the left ventricle to unfold and left ventricular volumes to exhibit a “creep” phenomenon. Serial echocardiography to assess left ventricular dimensions and forward flow is imperative.

In patients with borderline left heart, a hybrid procedure may be considered since it “buys time” while the options for an eventual biventricular or univentricular strategy are kept open. Meanwhile, growth of the left heart is allowed and can be closely monitored. The postponement of immediate decision making offers a chance to fully explore the growth potential of the left heart and avoids the risks of early pursuit of an inappropriate strategy.

A more difficult group comprises infants where biventricular repair is considered and significant pulmonary hypertension develops after ductal closure. In these, usually with systemic or suprasystemic right heart pressures, options are limited: one can create a “reversed” central shunt between pulmonary artery and aorta and re-band the pulmonary arteries or consider a Damus-Kaye type procedure with a large central shunt, but run the risk of shunt blockage because of the underlying pulmonary hypertension. However, these late “change of mind” -procedures are associated with considerable morbidity and mortality.

5. Advantages of the hybrid procedure in borderline left ventricles

The hybrid approach was initially developed as an alternative to the Norwood I procedure for hypoplastic left heart syndrome. Factors driving the concept of the hybrid route were the relatively high mortality rates of surgical stage I (5-30%) and a relatively bloodless intervention avoiding cardiopulmonary bypass.

In the setting of a borderline left ventricle this approach has distinct advantages:

- the hybrid procedure allows the physician to buy time for decision making in neonates with borderline left heart, as previously mentioned.
- in patients with increased operative risk for the first stage in a univentricular strategy (birth weight < 2.5 kg, prematurity < 34 weeks gestational age, genetic malformations, additional cardiac anomalies), the hybrid procedure offers an alternative, lower risk approach (Bacha, 2006).
- initial cardiopulmonary bypass surgery is avoided. In case a univentricular strategy is eventually selected, the hybrid approach allows for postponement of the Norwood I procedure to a later moment when it can be combined with the creation of the Glenn shunt. However, only two instead of three cardiopulmonary bypass procedures are thus necessary to reach the Fontan circulation, since the hybrid procedure is performed off-pump (Akintuerk et al., 2002, Venugopal et al., 2010). In addition, when the child is older and larger cardiopulmonary bypass surgery and circulatory arrest are better tolerated (Bacha, 2006, Corsini et al., 2011).

6. Technical considerations

The goal is to create a balanced pulmonary and systemic circulation in parallel, with sufficient oxygenation of blood in addition to adequate systemic perfusion simultaneously maintaining low pulmonary vascular resistances. Application of the hybrid procedure in the setting of a borderline left heart aims to buy time allowing the left heart to grow without being exclusively responsible for systemic output.

In order to achieve this, left ventricular outflow tract obstruction is treated, the arterial duct is stented to provide adequate systemic perfusion, whilst the branch pulmonary arteries are banded to protect the pulmonary vasculature from excessive flow and pressure (Fig. 1). If the interatrial communication is significantly restrictive, atrial left-to-right shunting may be improved by percutaneous intervention.

Ideally, the procedure is performed in a “hybrid” theatre, where both cardiopulmonary bypass and catheterization equipment is present (Bacha, 2006). In many centres, the surgical procedure and catheterisation procedure are performed separately, albeit with only a short time interval between the events.

6.1 Prostaglandin infusion

Prostaglandin E₁/Alprostadil is administered for temporary maintenance of arterial duct patency. Prostaglandin E₁ causes vasodilatation by its effect on vascular and ductus arteriosus smooth muscle (Roth, 2008). Given the short half-life of 5-10 minutes of this drug, it is administered by continuous intravenous infusion. The onset of action is usually within 30 minutes after administration is started and the maximum effect is observed after 1.5-3 hours (Roth, 2008). More than 70% is metabolized in the lungs, and 90% is excreted as metabolites in the urine within 24 hours (Roth, 2008). Concomitant use of antihypertensive

drugs can increase the risk of hypotension. The major adverse effect seen in 10% of newborns is apnoea requiring endotracheal intubation and artificial ventilation, with children < 2 kg being at increased risk (Roth, 2008). Other side effects such as flushing, bradycardia, tachycardia, hypotension, gastro-intestinal disturbances, oedema, seizures, electrolyte abnormalities, hypoglycemia, disturbed platelet aggregation and infection are seen less frequently.

6.2 Balloon dilation of the aortic valve

Standard techniques of balloon valvuloplasty in infants have been previously described (Kasten-Sportes et al., 1989). In order to reduce the risk of aortic regurgitation, balloon size should not be too large and an overall ratio of not more than 0.9 is recommended. In one large study on balloon dilation of the aortic valve in neonates, aortic regurgitation occurred in 15% (McElhinney et al., 2005). Rapid right ventricular pacing to improve balloon stability may be used, but is hardly ever required in these infants. Many interventionalists prefer a retrograde approach, but the procedure can be performed antegrade as well. Alternatively, a carotid artery approach provides easy, rapid access but concerns exist regarding stenosis of the carotids on long-term follow-up.

6.3 Stenting of the arterial duct

Meticulous attention should be paid to the technique of ductal stenting. Stents can be introduced directly into the pulmonary artery after bilateral branch banding during the initial procedure or, alternatively, percutaneously a few days after surgery.

Prostaglandin infusion may be stopped 4 – 12 hours prior to ductal stenting, but in our experience, this is usually not needed in these cases. Stent selection is based upon ductal anatomy. In general, the diameter should be at least the size of the thoracic aorta and it is important that the whole length of the arterial duct should be covered by the stent. Positioning of the stent at the aortic junction of the ductus is of utmost importance: it should not protrude into the lumen of the aorta, since this will preclude percutaneous management in the future. At the pulmonary end, the stent should extend beyond the ductus-pulmonary artery junction. Pre-mounted self expandable stents are preferred since it averts the need for long sheaths and the ductus in these cases is usually wide and curved e.g. OptiMed® sinus stent range (Optimed Medizinische Instrumente GmbH, Ettlingen, Germany) and Andramed® U-Flex (Andramed, Reutlingen, Germany) both 5 F systems. Self expandable stents are available in open- as well as closed-cell designs. Closed-cell stents can easily be repositioned before full expansion but have less grip. Open-cell designs on the other hand, have the advantage that it anchors itself to the wall of the ductus and can therefore not be repositioned. In the presence of a stenotic ductus, a balloon expandable stent could be selected e.g. Palmaz® Blue™ stents (Cordis Corporation, a Johnson & Johnson company; Warren, NJ) – these are mostly closed-cell designs. Generally ductal stents would be in the range of 5 -7 mm in diameter with lengths varying between 10 and 20 mm.

Delivery of the ductal stent can be performed in the hybrid suite. After direct puncture of the right ventricular outflow tract or main pulmonary artery, a short sheath is placed by the surgeon 2-3 mm inside the vessel and secured by means of a purse-string suture (Bacha, 2006). Heparin should be administered at this stage. A guidewire (0.014" or 0.018") is passed through the ductus into the descending aorta. Angiography is performed to assess ductal anatomy and position of the pulmonary artery bands. The balloon-deployable or self-

expandable stent is then advanced over the guide wire and positioned to cover the entire ductal length. A second stent can be used if necessary. Control angiography is performed after stent placement to confirm correct positioning (Fig 5).

Percutaneous stents can be delivered either antegradely or retrogradely. Premounted stents are delivered using 5 F or 6F short sheaths. Overall, we prefer the antegrade placement of stents 1 – 3 days after pulmonary artery banding during which a balloon septostomy can be performed if necessary. Technique is similar, but sometimes a long delivery sheath to navigate the curves of the right ventricle may be required although it is rarely necessary.

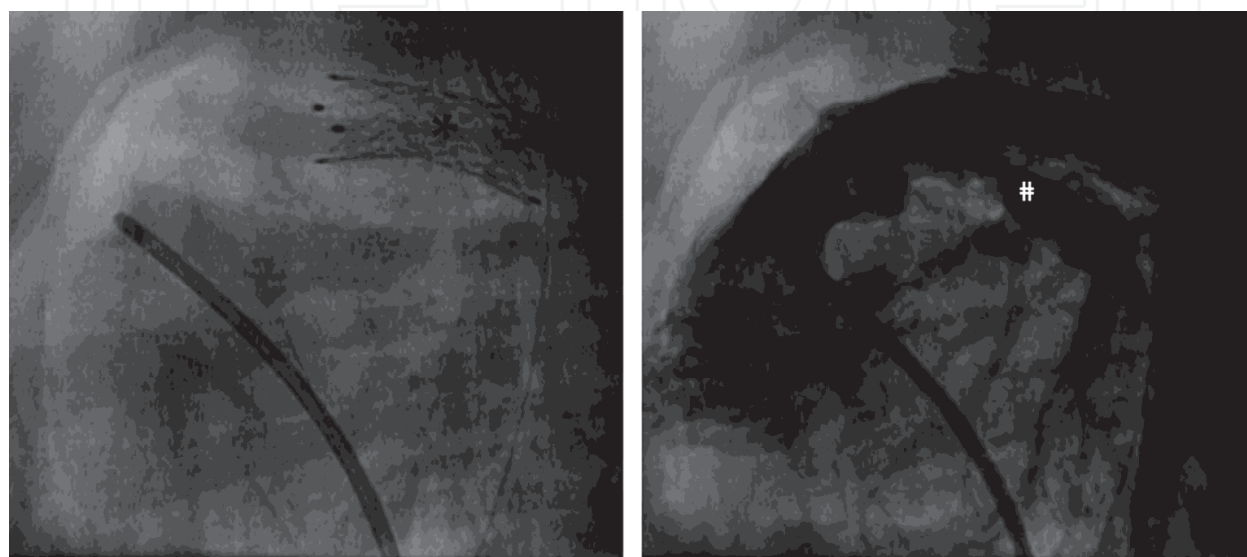


Fig. 5. Angiographic views of stent in the arterial duct. Pre and post contrast views showing patent stent in ductus (*) and pulmonary artery branch band (#) during contrast injection.

During the procedure, careful attention should be paid to the aortic arch and any associated abnormalities amenable to catheter intervention, for example coarctation of the aorta, should be addressed.

In a study of 58 patients undergoing the hybrid procedure, mortality related to percutaneous ductal stenting was 1.7% (Akinturk et al., 2007). Complications of ductal stenting include vascular injury, constriction of the ductus due to passage of the guide wire, insufficient coverage of the ductus necessitating re-intervention, ductal stent migration and thrombosis of the stent (Holzer et al., 2010).

6.4 Bilateral banding of the pulmonary arteries

Banding of the branch pulmonary arteries is performed to decrease pulmonary overflow and improve the balance of the systemic and pulmonary flows. Furthermore, banding provides protection of the pulmonary vasculature in order to be able to pursue a univentricular circulation or cardiac transplantation later in life (Pizarro et al., 2009). In a mathematical model of the circulation after the hybrid procedure in hypoplastic left heart syndrome, computational results indicate that the balance between systemic and pulmonary blood flow is sensitive to the degree of pulmonary artery banding rather than to the size of the ductal stent (Corsini et al., 2011).

By means of median sternotomy, bilateral banding of the branch pulmonary arteries is performed. The chest retractor has to be placed in such a way as to be removable during

fluoroscopy in the hybrid theatre. At our institution, Gore-Tex rings with diameters as suggested by Galantowicz are used: 3.0mm for infants < 2.5kg and 3.5mm for infants over 2.5kg (Galantowicz et al., 2008). The ring is cut through, placed around the artery and closed again using a 7/0 polypropylene stitch. The Gore-Tex rings are then fixed to the branch pulmonary arteries with a 7/0 polypropylene stitch. Rings should be placed close to the bifurcation of the pulmonary artery. This configuration creates breakable bands and is thus amenable to future percutaneous interventions. We have demonstrated that percutaneous saturations could be markedly improved after careful balloon angioplasty of such bands (Brown et al., 2010). The advantage of this concept is that, in a borderline left ventricle demonstrating growth, nonsurgical re-establishment of flows is possible (Brown et al., 2010).

Mortality for pulmonary arterial banding is as low as 1.7% as demonstrated by the Giessen experience which included 58 patients undergoing the hybrid procedure (Akinturk et al., 2007). Complications consist of band migration and pulmonary artery distortion which may require further intervention. Especially distal migration of bands to the hilus is an important cause of morbidity and need for surgical re-intervention.

6.5 Creating or enlarging the interatrial communication

If the interatrial septum is markedly restrictive, it should be addressed. In the vast majority of patients balloon septostomy will be adequate. Complication rates are low if the interatrial septum has normal anatomy, but much higher in the presence of abnormal anatomy (Holzer et al., 2008).

However, in the infant with a borderline left ventricle, mild restriction in order to force flow through the mitral valve is preferable and septostomy is rarely required. In our experience, infants with borderline left ventricles requiring septostomy often have less favourable long-term outcome and rarely proceed to biventricular repair.

7. Intermediate follow-up of hybrid procedures

Most of the data regarding the outcome of hybrid procedures have been obtained in infants with hypoplastic left heart syndrome. Hospital survival after the hybrid stage I procedure in infants with hypoplastic left heart has ranged from 70-90% (Bacha, 2006). In a series of 14 high-risk patients of whom 11 had hypoplastic left heart syndrome, in-hospital survival of the hybrid strategy was 11/14 (79%), death being caused by ductal stent embolization in one infant, progressive cardiac dysfunction in another, sepsis after cardiac arrest, extracorporeal membrane oxygenation and cardiac transplantation in one patient (Bacha et al., 2006). Reports have demonstrated that there is an initial learning curve and, in an expert center, in-hospital survival of hybrid stage I palliation in 40 infants with hypoplastic left heart syndrome was as high as 98% (Galantowicz et al., 2008). Between stage I and II palliation, 2 deaths occurred due to infection, 3 deaths were related to the stage II procedure and 1 death took place in-between stage II and III palliation (Galantowicz et al., 2008). This results in an overall survival of 83%, with interstage mortality and reintervention rate being similar as reported with consecutive Norwood procedures (Galantowicz et al., 2008). Other units have reported similar results (Honjo et al., 2009).

If growth of the left ventricle occurs and biventricular repair is considered, by using a hybrid procedure, it can be accomplished by either percutaneous or surgical means.

Percutaneous biventricular repair would consist of balloon angioplasty to remove the breakable bands and closure of the ductus arteriosus with a device (Brown et al., 2009). Redo aortic valve angioplasty may also be required at the time of intervention. Surgical alternatives would include removal of the bands plus ductal clipping with or without removal of stent or variants of the Ross-Konno procedure. In order to facilitate biventricular repair, aggressive left heart rehabilitation by resection of endocardial fibroelastosis and mitral valve cleavage have also been employed (Emami et al., 2009).

On the other hand, those who qualify for univentricular repair should proceed to stage II options as mentioned.

8. Left heart growth and decision making after the hybrid procedure

8.1 Growth of left heart

Current experience suggests that left heart structures are capable of growing under certain physiologic conditions and in several series of hypoplastic left heart syndrome, results show that some patients proceeded to biventricular repair. In an interesting chick model, it was demonstrated that restriction of flow gave rise to left ventricular hypoplasia and that restoration of flow resulted in physiological growth associated with myocyte hyperplasia (deAlmeida et al., 2007). Recently it was shown that when there is very low flow in the left heart leading to left heart hypoplasia during mid and late gestation, the left ventricle still retained the potential to grow to adequate size for support of a biventricular circulation (Vogel et al., 2010). There is only scant data available concerning growth of left heart structures after the hybrid procedure in infants with borderline left heart and consist mostly of isolated case reports. Ballard described a small series of patients with borderline left heart: all seven patients underwent a hybrid procedure, after which aortic discriminant score ($12.12 \times (\text{body surface area}) + 0.59 \times (\text{aortic annulus z-score}) + 5.73 \times (\text{left ventricle long axis/heart long axis}) - 7.02$) increased in all patients. Four of the seven patients had aortic valve stenosis and were considered inappropriate for biventricular repair in the neonatal phase. After the hybrid procedure, growth of left heart structures allowed for biventricular repair in three out of four patients (Ballard et al., 2010).

In a large study of infants with valvular aortic stenosis undergoing balloon angioplasty during the neonatal phase, the vast majority of patients with a z-score < -1 for aortic annulus size and left ventricular size prior to balloon valvuloplasty, demonstrated normalization of the z-score value within 1-2 years (McElhinney et al., 2005). In another series, aortic valve annulus, aortic sinus, and left ventricular dimension z-scores increased significantly over time, while mitral valve z-score remained below normal during follow-up (Han et al., 2007).

8.2 Decision making during follow-up after the hybrid procedure

The anatomic and physiologic variability makes it impractical to regard a single therapeutic approach optimal for all infants with borderline left hearts. Contemporary experience suggests a rethinking of current strategies and evaluation of new therapeutic options. To the best of our knowledge, there is presently no scoring system available to guide the decision on a biventricular versus univentricular strategy after the hybrid procedure. The available scoring systems in use are designed to guide the decision in the neonatal phase, but have not been validated in older, larger children. At present, this decision regarding univentricular versus biventricular strategy is therefore based on scoring systems combined with the experience and clinical assessment of the multidisciplinary cardiac team. Factors

that should be taken into account are mitral valve size and function, estimated risk of retrograde pulmonary hypertension after repair, left ventricular size and function, left ventricular outflow tract size and obstruction, aortic valve size and function, degree of endocardial fibroelastosis, degree of tricuspid valve regurgitation, and presence of other associated heart defects.

On a practical note, we have found echocardiographic demonstration of good antegrade flow in the distal aortic arch helpful when choosing between univentricular and biventricular repair. In doubtful cases, a cardiac catheterization with test occlusion of the stented ductus and atrial septal defect can be performed to assess suitability for a specific strategy provided that the pulmonary artery bands are not too tight.

Important questions still need to be addressed, for example: is a high risk biventricular repair always preferable to univentricular repair? In our institution, whenever possible, biventricular repair is favoured, but when findings are not convincingly in favour of biventricular repair, we tend to go the hybrid route. One should take into consideration that a direct univentricular strategy has a more favourable outcome compared to univentricular repair after failed biventricular attempt. It is important to recognize that embarking on an initial hybrid strategy allows time for left heart growth and also keeps the options of either single or biventricular repair open. With increasing number of hybrid procedures performed, more data providing tools for decision making will become available.

9. Prenatal intervention

Currently, many patients with borderline left heart are diagnosed antenatally when an asymmetric four chamber view is observed on foetal echocardiography. In some institutions, intra-uterine balloon dilation of the aortic valve is offered, although success rates are variable. Improvements in technique may further allow physicians to manipulate left heart flows and growth.

10. Conclusion

In the neonate with aortic stenosis and borderline left heart, the choice between a univentricular versus biventricular strategy can be very difficult. Application of the hybrid procedure not only buys time for left heart growth, but also does not preclude a patient from either single or biventricular repair whilst simultaneously keeping future therapeutic options open.

11. Acknowledgements

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12. References

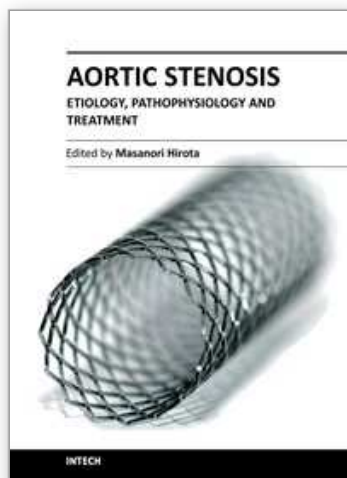
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Currently, aortic stenosis (AS) is the most prevalent valvular disease in developed countries. Pathological and molecular mechanisms of AS have been investigated in many aspects. And new therapeutic devices such as transcatheter aortic valve implantation have been developed as a less invasive treatment for high-risk patients. Due to advanced prevalent age of AS, further discovery and technology are required to treat elderly patients for longer life expectancy. This book is an effort to present an up-to-date account of existing knowledge, involving recent development in this field. Various opinion leaders described details of established knowledge or newly recognized advances associated with diagnosis, treatment and mechanism. Thus, this book will enable close intercommunication to another field and collaboration technology for new devices. We hope that it will be an important source, not only for clinicians, but also for general practitioners, contributing to development of better therapeutic adjuncts in the future.

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