Archives of Cardiovascular Disease (2009) **102**, 549—557

Available online at
ScienceDirect
www.sciencedirect.com

Elsevier Masson France
EM|consulte
www.em-consulte.com

REVIEW

Palliative procedures for congenital heart defects

Chirurgies palliatives dans les cardiopathies congénitales

Shi-Min Yuan, Hua Jing*

Department of Cardiothoracic Surgery, Jinling Hospital, Nanjing, 210002 Jiangsu Province, People's Republic of China

Received 2 April 2009; received in revised form 23 April 2009; accepted 23 April 2009
Available online 28 July 2009

KEYWORDS
Cardiac surgery;
Congenital heart defects;
Palliative procedures

Summary Although total repair of some congenital heart defects is possible in young children, palliative procedures still play an important role in relieving patients' symptoms, particularly in emergent settings, when total correction or surgical repair is not available. However, the concepts and taxonomies of the palliative procedures for congenital heart diseases sometimes seem to be ambiguous and confusing for young cardiac surgeons and cardiologists. This article gives a full-scope overview of the concepts, categories, indications, historical developments and clinical outcomes of the palliative procedures for congenital heart defects that have been documented in the literature. In total, there are 21 palliative procedures for the surgical management of congenital heart defects, which can be classified into four categories: firstly, increasing pulmonary artery flow for pulmonary oligaemia (including shunt procedures); secondly, decreasing pulmonary artery flow for pulmonary overcirculation (pulmonary banding and Norwood procedure); thirdly, enhancing intracardiac blood-oxygen mixture for systemic hypoxaemia (atrial septostomy subjected to different techniques); and, finally, other procedures, including congenital mitral or aortic stenosis palliation, coarctation of aorta palliation and hybrid palliative procedures for hypoplastic left heart syndrome. Modified Blalock-Taussig's and Glenn's shunts and pulmonary artery banding represent the pre-eminent palliative procedures for congenital heart defects and have been proven to be satisfactory after long-term clinical application. It seems that there is a growing trend towards the use of interventional techniques with stent deployment as an alternative to the surgical approach.

Abbreviations: HLHS, Hypoplastic left heart syndrome.

* Corresponding author. Fax: +86 25 84824051.
E-mail address: dr.jing@163.com (H. Jing).

1875-2136/$ — see front matter © 2009 Published by Elsevier Masson SAS.
doi:10.1016/j.acvd.2009.04.011

MOTS CLÉS
Chirurgie cardiaque ;
Cardiopathie congénitale ;
Chirurgie palliative ;
Pédiatrie

Résumé Bien que la réparation de nombre de malformations cardiaques soit possible chez le jeune enfant, les procédures palliatives continuent de jouer un rôle important dans le traitement des cardiopathies non réparables. Cependant, les concepts et procédures palliatives de correction des cardiopathies congénitales sont parfois ambigus et sources de confusion pour les chirurgiens et les cardiologues prenant en charge ces jeunes patients. Cette revue de la littérature apporte un éclairage sur les concepts, le contexte, les indications, les développements et le suivi clinique des patients ayant bénéficié d'une procédure palliative pour cardiopathies congénitales complexes. Au total, 21 procédures palliatives chirurgicales ont été revues et classées en quatre catégories : premièrement, augmentation du flux artériel pulmonaire en cas de débit pulmonaire diminué (en incluant les corrections de shunt), deuxièmement, diminution du flux artériel pulmonaire en cas de débit pulmonaire augmenté (cerclage pulmonaire et en incluant la procédure de Norwood), troisièmement, enrichissement du sang artériel en cas d'hypoxémie systématique (septostomie atriale selon différentes techniques) et, finalement, autres procédures incluant les traitements palliatifs pour sténoses mitrale ou aortique, les corrections palliatives des coarctations de l'aorte et les procédures hybrides proposées en cas d'hypoplasie du coeur gauche. Les shunts modifiés de Blalock-Taussig et de Glenn, le cerclage de l'artère pulmonaire représentent les procédures palliatives les plus pratiquées dans la prise en charge des cardiopathies congénitales complexes ; elles ont été validées lors des applications et des évaluations à long terme. Les techniques hybrides utilisant de façon concommittante la chirurgie et le cathétérisme interventionnel semblent très prometteuses.

Introduction

Operations for congenital heart disease may be classified as palliative, reparative or corrective with respect to the goals of treatment (i.e., obtaining normal heart function or relieving the symptoms of the heart disease) [1]. A palliative operation does not correct but is required to improve an abnormal heart function, minimizing the disorder, usually in children too young for corrective surgery. The aim is to lessen cyanosis, control heart failure or prepare the circulation for later correction when the baby grows up to an age and body weight that are suitable for the available techniques [2]. Although early total correction is now possible in many congenital anomalies, there is still need for palliation when definitive repair is not an option at the time or when intracardiac correction may carry a significantly higher mortality risk than staged shunting and subsequent correction [3].

There are 21 palliative procedures in total, which have been categorized into four classes, according to their goals and indications:

- increasing pulmonary artery flow for pulmonary oligaemia (including shunt procedures);
- decreasing pulmonary artery flow for pulmonary overcirculation (pulmonary banding and Norwood procedure);
- enhancing intracardiac blood-oxygen mixture for systemic hypoxaemia (atrial septostomy subjected to different techniques);
- other procedures, including congenital mitral or aortic stenosis palliation, coarctation of aorta palliation and hybrid palliative procedures for hypoplastic left heart syndrome (HLHS).

Palliative procedures

Shunt procedures

A systemic-to-pulmonary artery shunt has been used for many years to establish unobstructed systemic blood flow, normalize pulmonary blood flow and pressure and relieve pulmonary venous obstruction [4,5]. An ideal shunt is expected to have the following attributes: technical simplicity, good functionality, good long-term patency, easy takedown before repair and no residual shunt after closure. The indications for a systemic-to-pulmonary artery shunt include tetralogy of Fallot, tricuspid atresia, pulmonary atresia with intact ventricular septum, pulmonary atresia and ventricular septal defect, Ebstein's anomaly, single ventricle situation with pulmonary or aortic atresia and hypoplastic left heart syndrome (HLHS). There are 11 shunt procedures in total.

Blalock-Taussig's shunt

The classic Blalock-Taussig's shunt is a direct anastomosis between the transected subclavian artery (or the innominate artery) and the pulmonary artery (Fig. 1). It does not require the use of prosthetic material and offers the theoretical possibility for growth but requires extensive surgical dissection and sacrifices the subclavian artery [2]. The major disadvantages of classic shunts were long operative dissection time, phrenic nerve injury, technical difficulties during takedown and possible arm ischaemia [6]. After a classic shunt, distortion is usually explained by the failure of the anastomosis to grow and the effects of the scar tissue around the suture material. When classic shunts are performed, it is

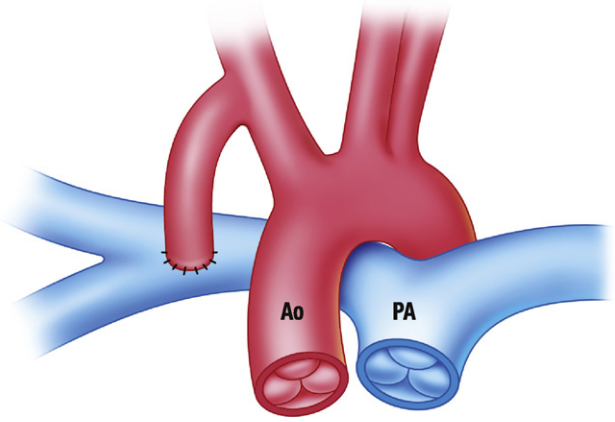

Figure 1. Classic Blalock-Taussig's shunt. AO: aorta; PA: pulmonary artery.

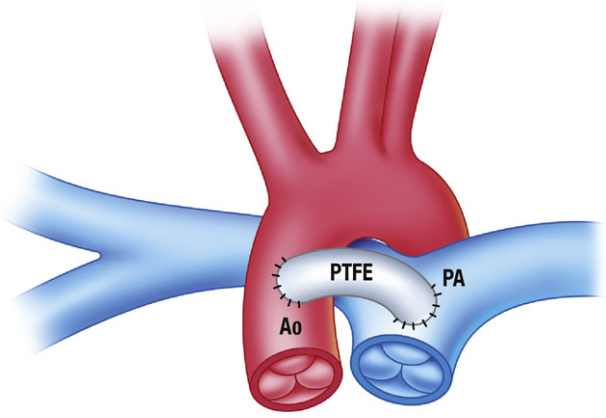

Figure 3. Central shunt. AO: aorta; PA: pulmonary artery; PTFE: polytetrafluoroethene.

usually reported that they should be constructed on the side opposite to the aortic arch to avoid pulmonary artery kinking [7]. Better understanding of the condition led to a revolution in the surgical technique for the Blalock-Taussig's shunt: an interposed graft (prosthetic or human vascular) was used between the subclavian and pulmonary arteries [8]; this is called a modified Blalock-Taussig's shunt (Fig. 2).

Central shunt

A central shunt is an anastomosis between the ascending aorta and the main pulmonary artery, made of prosthetic or other materials (Fig. 3) [9]; it is also known as the Mee's shunt [10]. The internal mammary artery is used to create a systemic-to-pulmonary artery shunt after failure of a previous Blalock-Taussig's shunt; this offers the advantage of growth and flow adaptation, eliminates the risk of prosthetic graft infection and does not affect blood flow to the arms [11]. The advantages of this technique are its applicability in small children with small peripheral vessels, prevention of distortion of pulmonary arteries, provision of equal pulmonary blood flow to both lungs, lower occlusion rate, avoidance of subclavian artery steal and ease of closure during corrective repair. The primary disadvantages are entry into the pericardium and inapplicability in patients without a patent ductus arteriosus or other source of pulmonary blood flow.

Glenn's shunt

A connection between the superior vena cava and the right pulmonary artery is known as a classic Glenn's shunt (Fig. 4). Since the late 1950s, the Glenn's shunt has been performed to improve pulmonary blood flow in patients with diverse cyanotic congenital heart disease. The Glenn's shunt does not create volume overload of the ventricle or increased work for the ventricle, as is the case with systemic-pulmonary artery shunts. It provides venous flow to the lung fields for oxygenation, rather than an arteriovenous mixture [12]. The bidirectional shunt is performed by connecting the superior vena cava to the right branch of the pulmonary artery using fine sutures and by dividing or tying up the pulmonary artery (Fig. 5). It decreases volume load on the single ventricle while improving oxygen saturation.

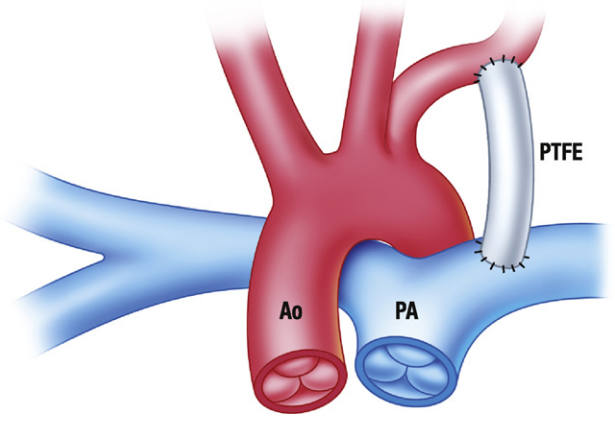

Figure 2. Modified Blalock-Taussig's shunt. AO: aorta; PA: pulmonary artery; PTFE: polytetrafluoroethene.

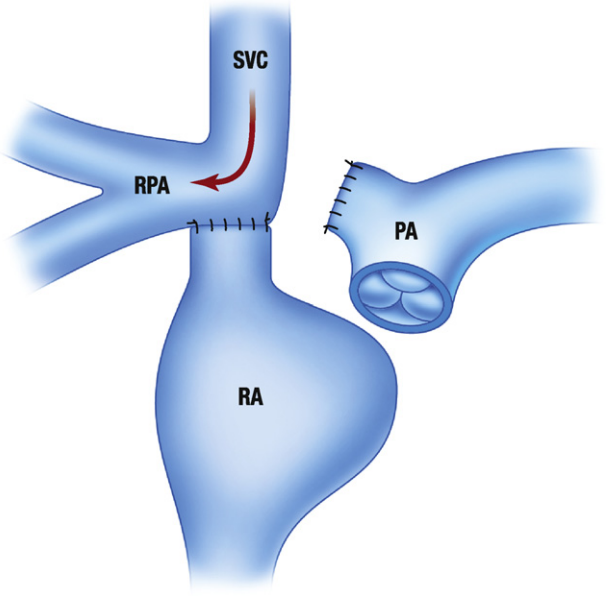

Figure 4. Classic Glenn's shunt. PA: pulmonary artery; RA: right atrium; RPA: right pulmonary artery; SVC: superior vena cava.

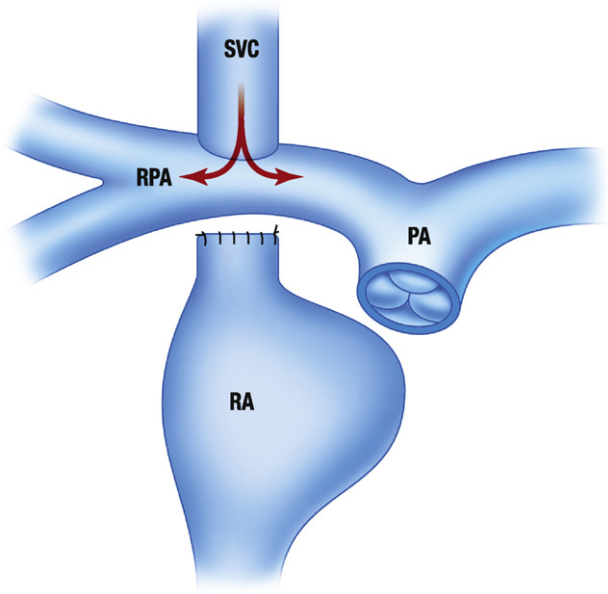

Figure 5. Bidirectional Glenn's shunt. RA: right atrium; RPA: right pulmonary artery; SVC: superior vena cava.

The bidirectional Glenn's shunt is preferred in very small babies, especially those aged less than 2 years, in whom lung vessel resistance is still quite high, and in borderline cases with abnormal pulmonary arteries. While avoiding the risk of failure of a complete Fontan operation, it also partly relieves symptoms [13]. The bidirectional Glenn's shunt is equivalent physiologically to half a Fontan's shunt, hence it is also referred to as a hemi-Fontan procedure. Progressive cyanosis, which may be due to the development of systemic venous collaterals that decompress the superior caval system into the inferior caval system, and the formation of a diffuse arteriovenous shunt not amenable to coil embolization are the two problems that occur most frequently after a bidirectional Glenn operation [14].

Sano's shunt

The most recent and intriguing modification to the Norwood procedure was the use of the right ventricle to pulmonary artery conduit, referred to as the Sano modification of the Norwood procedure, which allows blood to be pumped directly to the lungs. Sano contributed to the first application and description of this modification to the Norwood procedure. The technique was adapted after its initial description and greater haemodynamic stability was noted with this Sano modification than with the modified Blalock-Taussig's shunt [15].

''Wanna-be'' Blalock-Taussig's shunt

Ductus stenting in neonates and infants with duct-dependent cyanotic congenital heart disease has eliminated the need for palliative surgery. The procedure carries no risk of serious complications or pulmonary artery distortion and stenosis and gains time for the child and the pulmonary arteries to grow, while leaving the operative field for definitive surgery untouched [16]. Ruiz and Bailey [17] named ductus arteriosus stenting a ''wanna-be'' Blalock-Taussig's shunt.

Other shunts

The Potts's (Potts-Smith-Gibon) shunt is a connection between the descending aorta and the left pulmonary artery which was proposed initially as an alternative to the classic Blalock-Taussig's shunt in neonates. This shunt was abandoned because of the high incidence of subsequent pulmonary hypertension, the preferential blood flow to one lung with kinking and distortion of the pulmonary artery and the technical difficulties with takedown.

The Waterston-Cooley's shunt is a side-to-side anastomosis between the ascending aorta and the right pulmonary artery (extrapericardial [Waterston] and intrapericardial [Cooley]), which is indicated for tetralogy of Fallot. Because of complications (including preferential distribution of most or all shunt flow to the right lung, narrowing or obstruction of the right pulmonary artery at the anastomotic site, increasing stenosis or atresia of the right ventricular outflow tract, hypoplasia of the left pulmonary artery and obstruction of the shunt itself with the Waterston's shunt [18]) and a similar mortality rate to that for the Blalock-Taussig's shunt, it has been largely abandoned and substituted by the latter, except in infants aged less than 2 weeks [19]. It has been recognized that the Waterston-Cooley's shunt is often complicated by pulmonary overcirculation, left ventricular failure and pulmonary microvascular hypertension [20]. It is technically difficult to close this shunt properly at the time of total correction. Most surgeons have abandoned the Waterston-Cooley's shunt.

The Shumacker-Mandelbaum's shunt was a shunt technique between the ascending aorta and either the right or left pulmonary artery or the main pulmonary artery. A graft composed of homologous aorta with a Dacron cuff at either end for end-to-side anastomoses was used [21].

The Redo-Ecker's shunt was named after Redo and Ecker [22], who used a woven teflon prosthesis anastomosed end-to-side between the aorta and the main pulmonary artery, just distal to the origins of these vessels within the pericardium. In this way, the blood is delivered directly into the proximal portion of the pulmonary artery and the artery is more suitable for enlargement, which benefits total correction in the future.

An aorta-to-right ventricle shunt was applied in only one of 19 patients requiring a systemic-to-pulmonary shunt, as reported by Nanton et al. [23].

Pulmonary artery banding and the Norwood procedure

Pulmonary artery banding

Pulmonary artery banding was first described by Muller and Dammann in 1951 [24], as the ''creation of pulmonary stenosis'' in a 5-month-old infant with a large ventricular septal defect and pulmonary overcirculation. Nowadays, pulmonary artery banding remains the preferred method of palliation in children born with cardiac defects characterized by left-to-right shunting and pulmonary overcirculation. This technique has been broadened to treat congestive heart failure caused by large ventricular septal defects, atrioventricular canal defects and tricuspid atresia and is indicated for patients with transposition of the great arteries for rapid ventricular retraining and for patients with

HLHS. Pulmonary artery banding is contraindicated in the presence of significant subaortic obstruction, such as single ventricle defects in which the aorta arises from an outflow chamber, and in patients who are at high risk of such an obstruction; in addition, it is not used in patients with truncus arteriosus.

Norwood procedure

This is a complex procedure, which includes neo-aorta reconstruction, aortic arch augmentation and a modified Blalock-Taussig's or Sano's shunt placement (Figs. 6 and 7) and was designed to treat HLHS, as well as aortic atresia or aortic stenosis with inadequate left ventricle, mitral atresia or stenosis, interrupted aortic arch and even double-outlet right ventricle with mitral atresia or complete transposition with a hypoplastic right ventricle [25].

Atrial septostomy subjected to different techniques

Blalock-Hanlon operation (atrial septectomy without cardiopulmonary bypass)

In 1948, Blalock and Hanlon [26] described an atrial septectomy in which an atrial septal defect was created to improve intracardiac mixing at the atrial level, which then became the standard treatment for transposition of the great arteries where the atrial septal defect was too small to allow complete mixing. In transposition of the great arteries with ventricular septal defect, the Blalock-Hanlon operation was added when pulmonary artery banding was carried out. In cases of transposition of the great arteries and subpulmonary stenosis, the Blalock-Hanlon operation could be performed simultaneously or before a systemic-to-pulmonary shunt [27]. The Blalock-Hanlon operation was also indicated for palliation of mitral atresia [28].

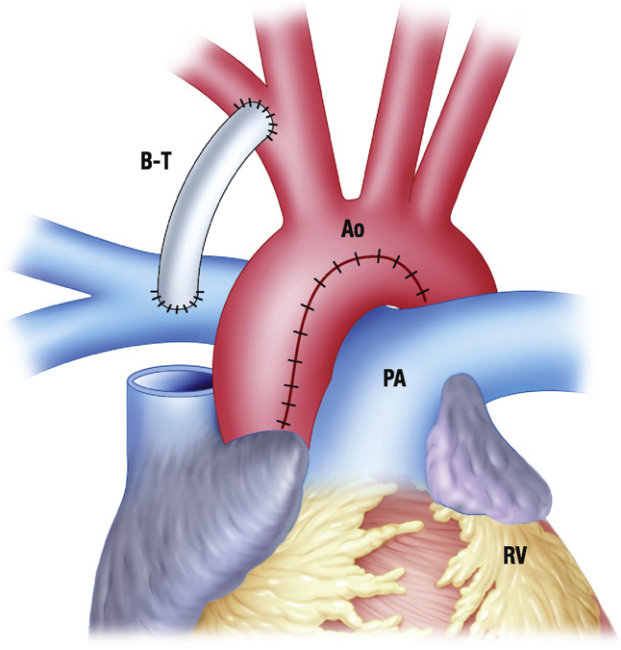

Figure 6. Norwood procedure with a modified Blalock-Taussig's shunt. AO: aorta; B-T: modified Blalock-Taussig's shunt; PA: pulmonary artery; RV: right ventricle.

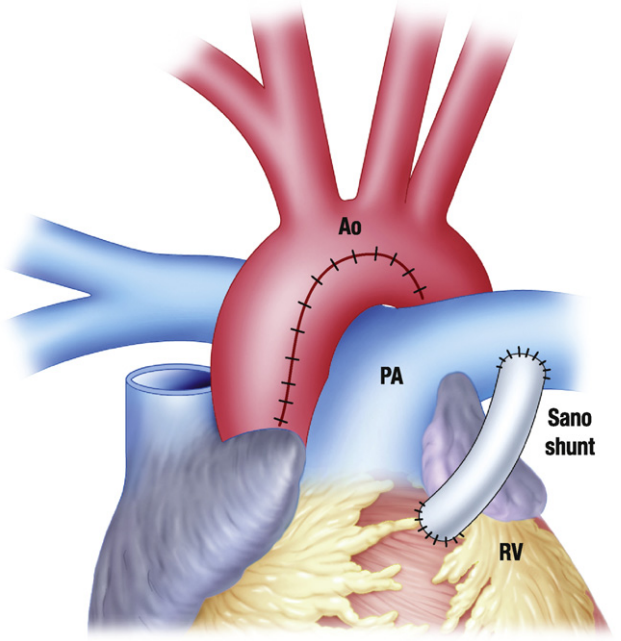

Figure 7. Norwood procedure with a Sano's shunt. AO: aorta; PA: pulmonary artery; RV: right ventricle.

Park procedure (balloon or blade atrial septostomy)

The main indication for this procedure is transposition of the great arteries, aiming at enlarging the foramen ovale, and thereby allowing some oxygenated blood to pass from the left ventricle into the aorta.

Rashkind procedure (balloon catheter atrial septostomy)

The balloon catheter technique for atrial septostomy was introduced by Rashkind and Miller [29] in 1966; it was applied broadly and displaced the surgical atrial septal defect creation technique by offering acceptable immediate palliation in small infants with complete transposition of the great arteries and those unlikely to achieve successful correction [30].

Atrial switch operation

There are two types of atrial switch operations: the Mustard's operation and the Senning's operation. Both are similar in principle but differ in technique. The atrial switch operation is an open-heart procedure under cardiopulmonary bypass, during which the right atrium is opened and the wall between the atria is fully removed. By using the pericardium (Mustard) or flaps made from the atrial septum and wall (Senning), a ''baffle'' is constructed, directing blood from the veins in the right atrium towards the left ventricle [31]. Palliative atrial switch procedures without closure of the ventricular septal defect have been used in the treatment of deeply cyanotic patients with severe pulmonary vascular obstructive disease [32].

Other palliative procedures

Mitral valve palliation

Congenital mitral stenosis requires surgical palliation in small infants in whom currently available prosthetic valves cannot be inserted into the mitral annulus. A left atrial-left ventricular conduit bypass is an alternative for the relief of congenital mitral stenosis and can obviate the need for valve replacement in infancy [33].

Aortic valvotomy

Aortic valvotomy for the relief of congenital aortic stenosis is a palliative operation [34]. Current therapy for critical congenital aortic stenosis includes either balloon or surgical valvotomy. Both procedures leave residual lesions that often require further treatment [35].

Palliation of coarctation of the aorta

Intravascular stent placement for native and recurrent coarctation of the aorta has been used successfully in various locations [36].

Hybrid palliation

Hybrid cardiac surgery was defined as combined catheter-based ductal stenting and surgical interventions such as bilateral pulmonary artery banding either in one setting or in a sequential fashion within 24 h [37]. Hybrid palliation, which was developed originally for HLHS, is now applied to patients with a functional single ventricle and restrictive systemic outflow tract, such as tricuspid atresia with transposition of the great arteries [38]. The primary palliation procedures include stenting of the ductus arteriosus with a self-expanding nitinol stent to secure adequate systemic blood flow, placement of an internal pulmonary arterial band to protect the pulmonary vascular bed and prevent pulmonary overcirculation and widening of the interatrial communication by blade and balloon septostomy or static balloon dilation to decompress the left atrium [39].

Discussion

Although the indications for palliative procedures for congenital heart defects are dwindling due to the growth of primary total correction in neonates and children, palliative procedures still play an important role in high-risk patients with complex cardiac lesions. Advances in congenital heart surgery have given greater insight into the haemodynamic features of the defects and those subjected to the underlying operations, and have resulted in continual innovation in palliative procedural techniques.

Of the shunt procedures, the modified Blalock-Taussig has remained the preferred procedure for relieving pulmonary overcirculation since its advent, despite some operative complications (such as chylothorax, chylopericardium, phrenic nerve paralyses, serous fluid leak, steal syndrome, pulmonary arterial disease, etc). Central shunts have fallen largely into disfavour because of the high incidence of complications, including shunt thrombosis, congestive heart failure and pulmonary artery distortion [40]. But according to a recent study, the use of the Shelhigh internal mammary artery graft instead of synthetic tubular grafts in the modified Blalock-Taussig and central shunts for congenital heart diseases with decreased pulmonary blood flow did not show any evidence of calcification. Echocardiography evaluations revealed no shunt obstruction for the early (first postoperative week) or middle (postoperative week 24) period [41]. The bidirectional cavopulmonary shunt has been widely accepted and used in patients with diverse cyanotic congenital heart disease to improve systemic arterial oxygen saturation without increasing ventricular workload or pulmonary vascular resistance [42]. A recent report has revealed successful transcatheter Glenn's or central shunts in swine, representing a modern technical orientation for shunt procedures [43]. The ''wanna-be'' Blalock-Taussig's shunt produced an overall survival rate of 86% at 6 year postoperative follow-up [44]. Other shunts, like Potts, Waterston-Cooley, Shumacker-Mandelbaum, Redo-Ecker and aorta-right ventricle shunts were substituted due to their unacceptable complications or merely had an occasional application with transient instrumentality.

Pulmonary artery banding has become the procedure that is used most frequently to relieve pulmonary overcirculation. The goal of pulmonary artery banding is to produce a distal pulmonary artery pressure of 30 to 50% of systemic pressure. A variety of banding material is available but umbilical tape is broad enough to minimize the risk of eroding through the pulmonary artery wall. An adjustable pulmonary artery banding has been applied since 1972 [45]. A percutaneously-adjustable band with a fluid-filled reservoir that allows variable constriction was developed in 1986, followed by an implantable, telemetrically-controlled, battery-free device (FloWatch) in recent years. All have been proven to be effective in either experimental animals or humans [46,47].

An important modification of the Norwood procedure was a Sano's shunt insertion instead of the modified Blalock-Taussig's shunt [25]. This Sano modification of the Norwood procedure resulted in a more favourable distribution of systemic, pulmonary and coronary blood flow [48]. Apart from a higher diastolic blood pressure in the right ventricle-pulmonary artery conduit group, no difference was found in early haemodynamic profile or postoperative mortality rate between patients undergoing a Sano or a Blalock-Taussig's shunt [49—51]. Some authors suggested that the pulmonary artery growth was similar with the Blalock-Taussig's and the Sano's shunts [52]; others reported that the pulmonary artery index was greater in Sano's shunt patients [53], with preferential growth to the left branch [54]. Hospital mortality rate after the Norwood procedure was 8% and overall survival was 76%. The estimated 1 year and 5 years survival rates were 80 and 73%, respectively. Using Cox regression analysis, body weight is less than 2.5 kg and tricuspid regurgitation is grater or equal to grade 2+ were two independent factors associated with midterm survival. Low body weight and tricuspid valve regurgitation were associated with worse outcome [55]. Based on a large patient population, including 199 patients initially with Norwood stage 1, univariate analysis demonstrated the following significant predictors of mortality: right ventricular dominance ($p = 0.0023$), mechanical circulatory support before stage 1 ($p = 0.0192$) and significant non-cardiac abnormality

or syndrome, including Down's syndrome, Turner's syndrome, heterotaxy, asplenia, polysplenia, biliary atresia or other chromosomal abnormality ($p < 0.0001$). Multivariable logistic regression analysis revealed the presence of a significant non-cardiac abnormality or syndrome, or prematurity less than 35 weeks, or mechanical circulatory support before stage 1 to be significant predictors of mortality ($p < 0.0001$) [56]. A longer cardiopulmonary bypass time was another significant risk factor for in-hospital mortality [51].

The Blalock-Hanlon, Park and Rashkind's procedures are classic surgical methods indicated for the palliation of transposition of the great arteries. From 1948 to 1964, the Blalock-Hanlon atrial septectomy was associated with a 60% operative mortality rate. Since then, the mortality rate has declined steadily to 10.7 to 21% [57]. A modified version of atrial septectomy using a Ferris-Smith-Kerrison bone punch under transoesophageal echocardiography monitoring obtained better results, with arterial saturation increased from 62 to 80% [58]. The procedure performed in neonates and children with transposition of the great arteries, mitral atresia, tricuspid atresia or miscellaneous anomalies showed an improvement rate of 79% [59]. After blade atrial septostomy, the interatrial pressure gradient diminished remarkably with prompt clinical improvement [60]. Atrioseptostomy with a balloon catheter is a safe and effective procedure in neonates with transposition of the great arteries. It is usually performed on an emergent basis monitored by fluoroscopy or bedside transthoracic echocardiography. The access routes used most frequently for this procedure are the femoral and umbilical veins. The greatest advantage of umbilical access in neonates is that it vacates other vessels for future use. Modified techniques, such as a French 6 catheter enclosing a tiny surgical blade [61], a ''butterfly'' stent atrial septostomy guided by intracardiac ultrasound [62] or concentrical deployment of two stents [63], could obtain the precise diameter of the atrial defect or pose fewer major complications.

An atrial switch operation is indicated for transposition of the great arteries, ventricular septal defect and severe pulmonary vascular obstructive disease. In such patients, the ventricular septal defect was not closed, because closure would be associated with a prohibitive early and late mortality risk [64]. The results of palliative atrial switch operations have shown improved early mortality rates and considerable relief of symptoms in deeply cyanotic patients with severe pulmonary vascular obstructive disease [65]. Observations showed that patients have benefited from a palliative Mustard's or Senning's repair, in terms of a marked improvement in symptoms with a decrease in haemoglobin and an increase in arterial oxygen saturation [32].

Left atrial-left ventricular valved bypass conduits have favoured adults, children and infants with stenotic mitral valves [66,67]. After insertion of the conduit, pulmonary artery pressure became normal, and persistent pulmonary oedema and ascites disappeared. Associated coarctation of the aorta, patent ductus arteriosus and ventricular septal defect were corrected simultaneously. In order to allow normal growth of all cardiac chambers, and to maintain the possibility of future total repair, a prosthetic patch-free method was adopted [68] and a policy of increasing the surface valve area rather than reconstructing an anatomic mitral valve was recommended [69].

The operative survival rate of aortic valvotomy has been 100% over the past 20 years for children and is currently greater than 80% in newborns in the absence of a hypoplastic left ventricle and/or endocardial fibroelastosis [34]. Open valvotomy for critical aortic stenosis in neonates carries a low operative risk and provides lengthy freedom from recurrent stenosis or regurgitation. Reoperations are inevitable but aortic valve replacement can be postponed until insertion of an adult-sized prosthesis is possible [70]. In neonates and infants with isolated aortic stenosis, an operative mortality rate of 6% was achieved with both open surgical and closed techniques [71,72], while the mortality rate was 36% in patients with complex aortic stenosis using the closed technique [71]. Transcatheter balloon dilatation is an accepted technique for palliation of congenital aortic valve stenosis. Efficacy has also been confirmed in patients who have undergone a previous surgical valvotomy [73].

Aortic coarctation palliation can be an alternative to surgery or balloon angioplasty, and has produced excellent results in the short term and intermediate term. Stents were implanted in 33 of 34 patients and outcome was successful in 32 of 33 patients. The peak systolic pressure gradient decreased from 32 ± 12 to 4 ± 11 mmHg. The coarctation site to descending aorta diameter ratio increased from 0.46 ± 0.16 to 0.92 ± 0.16 [36].

The hybrid procedure is important in the current era for high-risk HLHS neonates, yielding a hospital survival rate of 78.5% [74]. A hybrid procedure may combine variant manoeuvres such as bilateral pulmonary artery banding, ductus arteriosus stenting, balloon atrial septostomy and even main pulmonary artery-to-innominate artery shunt [75]. There are no significant differences in hospital and interstage mortality rates between the hybrid and the conventional Norwood strategies [76]. Using hybrid palliation, the Norwood stage I operation can be avoided in the neonatal period, so that the children can be scheduled for cardiac transplantation and be observed for left ventricular growth suitable for biventricular repair [77].

Conclusion

Modified Blalock-Taussig and Glenn's shunts and pulmonary artery banding represent the pre-eminent palliative procedures for congenital heart defects and have been proven to be satisfactory after long-term clinical application. It seems that there is a growing trend towards the use of interventional techniques with stent deployment as an alternative to the surgical approach. Meanwhile, greater interest has been generated in the hybrid procedure, which incorporates various interventional and/or surgical methods for HLHS neonates.

References

[1] Joffs C, Sade RM. Congenital Heart Surgery Nomenclature and Database Project: palliation, correction or repair? Ann Thorac Surg 2000;69:S369—72.

[2] Moulton AL, Brenner JI, Ringel R, et al. Classic versus modified Blalock-Taussig's shunts in neonates and infants. Circulation 1985;72:II35—44.

[3] Surgical procedures for congenital heart disease. Available from: http://www.echoincontext.com/advanced/chd_22.asp [accessed on 23 April 2009].
[4] Freedom RM, Yoo SJ, Russell J, Perrin D, Williams WG. Designing therapeutic strategies for patients with a dominant left ventricle, discordant ventriculo-arterial connections and unobstructed flow of blood to the lungs. Cardiol Young 2004;14:630—53.
[5] Lang P, Norwood WI. Hemodynamic assessment after palliative surgery for hypoplastic left heart syndrome. Circulation 1983;68:104—8.
[6] Karpawich PP, Bush CP, Antillon JR, Amato JJ, Marbey ML, Agarwal KC. Modified Blalock-Taussig's shunt in infants and young children. Clinical and catheterization assessment. J Thorac Cardiovasc Surg 1985;89:275—9.
[7] Godart F, Qureshi SA, Simha A, et al. Effects of modified and classic Blalock-Taussig's shunts on the pulmonary arterial tree. Ann Thorac Surg 1998;66:512—7, discussion 518.
[8] Lonyai T, Zaborszky B, Karpati P. Synthetic vascular prosthesis for Blalock-Taussig anatomosis. Acta Chir Acad Sci Hung 1966;7:361—9.
[9] Alboliras ET, Chin AJ, Barber G, Helton JG, Pigott JD, Norwood WI. Pulmonary artery configuration after palliative operations for hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 1989;97:878—85.
[10] Amark KM, Karamlou T, O'Carroll A, et al. Independent factors associated with mortality, reintervention and achievement of complete repair in children with pulmonary atresia with ventricular septal defect. J Am Coll Cardiol 2006;47:1448—56.
[11] Longaker MT, Merrick S, Crombleholme TM, Langer JC, Verrier ED, Turley K. Systemic-to-pulmonary artery shunt using the internal mammary artery. Ann Thorac Surg 1989;47:464—5.
[12] http://info.med.yale.edu/intmed/cardio/chd/resources/pdf/glenn.pdf [accessed on 23 April 2009].
[13] http://www.fontanoperation.com/fontan5.htm [accessed on 23 April 2009].
[14] Jonas RA. Indications and timing for the bidirectional Glenn's shunt versus the fenestrated Fontan circulation. J Thorac Cardiovasc Surg 1994;108:522—4.
[15] Reemtsen BL, Pike NA, Starnes VA. Stage I palliation for hypoplastic left heart syndrome: Norwood versus Sano modification. Curr Opin Cardiol 2007;22:60—5.
[16] Schneider M, Zartner P, Sidiropoulos A, Konertz W, Hausdorf G. Stent implantation of the arterial duct in newborns with duct-dependent circulation. Eur Heart J 1998;19:1401—9.
[17] Ruiz CE, Bailey LL. Stenting the ductus arteriosus: a ''wannabe'' Blalock-Taussig. Circulation 1999;99:2608—9.
[18] Levin DC, Fellows KE, Sos TA. Angiographic demonstration of complications resulting from the Waterston procedure. AJR Am J Roentgenol 1978;131:431—7.
[19] Laks H, Marco JD, Willman VL. The Blalock-Taussig's shunt in the first six months of life. J Thorac Cardiovasc Surg 1975;70:687—91.
[20] Reitman MJ, Galioto Jr FM, el-Said GM, Cooley DA, Hallman GL, McNamara DG. Ascending aorta to right pulmonary artery anastomosis. Immediate results in 123 patients and one month to six year follow-up in 74 patients. Circulation 1974;49:952—7.
[21] Shumacker Jr HB, Mandelbaum I. Ascending aorticpulmonary artery shunts in cyanotic heart disease. Surgery 1962;52:675—8.
[22] Redo SF, Ecker RR. Intrapericardial aorticopulmonary artery shunt. Circulation 1963;28:520—4.
[23] Nanton MA, Roy DL, Murphy DM, et al. Polytetrafluoroethylene shunts in congenital heart disease. Can J Surg 1982;25:134—8.
[24] Muller Jr WH, Danimann Jr JF. The treatment of certain congenital malformations of the heart by the creation of pulmonic stenosis to reduce pulmonary hypertension and excessive pulmonary blood flow; a preliminary report. Surg Gynecol Obstet 1952;95:213—9.
[25] Caspi J, Pettitt TW, Mulder T, Stopa A. Development of the pulmonary arteries after the Norwood procedure: comparison between Blalock-Taussig's shunt and right ventricular-pulmonary artery conduit. Ann Thorac Surg 2008;86:1299—304.
[26] Blalock A, Hanlon CR. The surgical treatment of complete transposition of the aorta and the pulmonary artery. Surg Gynecol Obstet 1950;90:1—15, illust.
[27] Zamora R, Moller JH, Lucas Jr RV, Castaneda AR. Complete transposition of the great vessels: surgical results of emergency Blalock-Hanlon operation in infants. Surgery 1970;67:706—10.
[28] Redo SF, Engle MA, Ehlers KH, Farnsworth PB. Palliative surgery for mitral atresia. Arch Surg 1967;95:717—23.
[29] Rashkind WJ, Miller WW. Creation of an atrial septal defect without thoracotomy. A palliative approach to complete transposition of the great arteries. JAMA 1966;196:991—2.
[30] Venables AW. Balloon atrial septostomy in complete transposition of great arteries in infancy. Br Heart J 1970;32:61—5.
[31] Transposition of great arteries. Available from: http://www.chdinfo.com/chdarticles/tga6.htm [accessed on 23 April 2009].
[32] Dunn JM, Donner R, Black I, Balsara RK. Palliative repair of transposition of the great arteries with criss-cross heart: ventricular septal defect and hypoplastic right (systemic) ventricle. J Thorac Cardiovasc Surg 1982;83:755—60.
[33] Midgley FM, Perry LW, Potter BM. Conduit bypass of mitral valve: a palliative approach to congenital mitral stenosis. Am J Cardiol 1985;56:493—4.
[34] Tveter KJ, Foker JE, Moller JH, Ring WS, Lillehei CW, Varco RL. Long-term evaluation of aortic valvotomy for congenital aortic stenosis. Ann Surg 1987;206:496—503.
[35] van Son JA, Reddy VM, Black MD, Rajasinghe H, Haas GS, Hanley FL. Morphologic determinants favoring surgical aortic valvuloplasty versus pulmonary autograft aortic valve replacement in children. J Thorac Cardiovasc Surg 1996;111:1149—56, discussion 1156-7.
[36] Hamdan MA, Maheshwari S, Fahey JT, Hellenbrand WE. Endovascular stents for coarctation of the aorta: initial results and intermediate-term follow-up. J Am Coll Cardiol 2001;38:1518—23.
[37] Bacha EA, Hijazi ZM, Cao QL, et al. Hybrid pediatric cardiac surgery. Pediatr Cardiol 2005;26:315—22.
[38] Yun TJ, Cho WC, Jung SH, Seo DM, Goo HW, Kim YH. Reverse Blalock-Taussig's shunt facilitates the growth of the ascending aorta after hybrid palliation. Ann Thorac Surg 2007;83:1886—8.
[39] Chan KC, Mashburn C, Boucek MM. Initial transcatheter palliation of hypoplastic left heart syndrome. Catheter Cardiovasc Interv 2006;68:719—26.
[40] Barragry TP, Ring WS, Blatchford JW, Foker JE. Central aorta-pulmonary artery shunts in neonates with complex cyanotic congenital heart disease. J Thorac Cardiovasc Surg 1987;93:767—74.
[41] Poyrazoglu HH, Avsar MK, Tor F, et al. Systemic pulmonary shunt performed with Shelhigh internal mammary artery: early results. Heart Surg Forum 2008;11:E50—3.
[42] Lamberti JJ, Spicer RL, Waldman JD, et al. The bidirectional cavopulmonary shunt. J Thorac Cardiovasc Surg 1990;100:22—9, discussion 29-30.
[43] Levi DS, Danon S, Gordon B, et al. Creation of transcatheter aortopulmonary and cavopulmonary shunts using magnetic catheters: feasibility study in Swine. Pediatr Cardiol 2009.
[44] Michel-Behnke I, Akintuerk H, Thul J, Bauer J, Hagel KJ, Schranz D. Stent implantation in the ductus arteriosus for pulmonary blood supply in congenital heart disease. Catheter Cardiovasc Interv 2004;61:242—52.
[45] Nahas R, Mundth ED, Ross B, Austen WG. Adjustable instrument for pulmonary artery banding. Description of instrument and

technique of application. J Thorac Cardiovasc Surg 1972;63: 732—4.
[46] Corno AF. Pulmonary artery banding. Swiss Med Wkly 2005;135: 515—9.
[47] Solis E, Heck CF, Seward JB, Kaye MP. Percutaneously adjustable pulmonary artery band. Ann Thorac Surg 1986;41:65—9.
[48] Simsic JM, Cuadrado A, Kirshbom PM, et al. Novel management strategy for severe cyanosis after Sano modification of the Norwood procedure. J Thorac Cardiovasc Surg 2005;129:1450—1.
[49] Aramburo A. Stage I palliation for hypoplastic left heart syndrome: is a right ventricle to pulmonary artery conduit associated with improved outcomes? Critical appraisal of Cua et al: early postoperative outcomes in a series of infants with hypoplastic left heart syndrome undergoing stage I palliation operation with either modified Blalock-Taussig's shunt or right ventricle to pulmonary artery conduit. Pediatr Crit Care Med 2008;9:438—40 [Pediatr Crit Care Med 2006;7:238-244].
[50] Edwards L, Morris KP, Siddiqui A, Harrington D, Barron D, Brawn W. Norwood procedure for hypoplastic left heart syndrome: BT shunt or RV-PA conduit? Arch Dis Child Fetal Neonatal Ed 2007;92:F210—4.
[51] Garcia-Hernandez JA, Gonzalez-Rodriguez JD, Martinez-Lopez AI, et al. Experience with the Norwood operation for hypoplastic left heart syndrome. Rev Esp Cardiol 2007;60:732—8.
[52] Nakano T, Fukae K, Sonoda H, et al. Follow-up study of pulmonary artery configuration in hypoplastic left heart syndrome. Gen Thorac Cardiovasc Surg 2008;56:54—61.
[53] Pizarro C, Norwood WI. Right ventricle to pulmonary artery conduit has a favorable impact on postoperative physiology after stage I Norwood: preliminary results. Eur J Cardiothorac Surg 2003;23:991—5.
[54] Lim DS, Peeler BB, Matherne GP, Kramer CM. Cardiovascular magnetic resonance of pulmonary artery growth and ventricular function after Norwood procedure with Sano modification. J Cardiovasc Magn Reson 2008;10:34.
[55] Sano S, Huang SC, Kasahara S, Yoshizumi K, Kotani Y, Ishino K. Risk factors for mortality after the Norwood procedure using right ventricle to pulmonary artery shunt. Ann Thorac Surg 2009;87:178—85, discussion 185-6.
[56] Jacobs JP, O'Brien SM, Chai PJ, Morell VO, Lindberg HL, Quintessenza JA. Management of 239 patients with hypoplastic left heart syndrome and related malformations from 1993 to 2007. Ann Thorac Surg 2008;85:1691—6, discussion 1697.
[57] Cohen DJ, Chopra PS. The Blalock-Hanlon operation: an anachronism? Ann Thorac Surg 1987;44:407—10.
[58] Takahashi M, Kanazawa H, Yamazaki Y, Ueno M, Sakano T. A new, safe and easy technique of atrial septal defect creation. Ann Thorac Surg 2000;69:293—4.
[59] Park SC, Neches WH, Mullins CE, et al. Blade atrial septostomy: collaborative study. Circulation 1982;66:258—66.
[60] Park SC, Neches WH, Zuberbuhler JR, et al. Clinical use of blade atrial septostomy. Circulation 1978;58:600—6.
[61] Ozkutlu S, Ozme S, Saraclar M, Baysal K. Balloon atrial septostomy using echocardiographic monitoring. Jpn Heart J 1988;29:415—9.
[62] Prieto LR, Latson LA, Jennings C. Atrial septostomy using a butterfly stent in a patient with severe pulmonary arterial hypertension. Catheter Cardiovasc Interv 2006;68:642—7.
[63] Girona J, Gran F, Garcia B, Marti G. Percutaneus double stent atrial septostomy. Catheter Cardiovasc Interv 2007;69:227—30.
[64] Mair DD, Ritter DG, Danielson GK, Wallace RB, McGoon DC. The palliative Mustard's operation: rationale and results. Am J Cardiol 1976;37:762—8.
[65] Allan LD, Leanage R, Wainwright R, Joseph MC, Tynan M. Balloon atrial septostomy under two dimensional echocardiographic control. Br Heart J 1982;47:41—3.
[66] Laks H, Hellenbrand WE, Kleinman C, Talner NS. Left atrial—left ventricular conduit for relief of congenital mitral stenosis in infancy. J Thorac Cardiovasc Surg 1980;80:782—7.
[67] Lansing AM, Elbl F, Solinger RE, Rees AH. Left atrial-left ventricular bypass for congenital mitral stenosis. Ann Thorac Surg 1983;35:667—9.
[68] Westerman GR, VanDevanter SH, Norton Jr JB, Readinger RI. Congenital mitral valve stenosis in infancy: a different approach to a difficult problem. J Thorac Cardiovasc Surg 1987;94:305—7.
[69] Serraf A, Zoghbi J, Belli E, et al. Congenital mitral stenosis with or without associated defects: an evolving surgical strategy. Circulation 2000;102:III166—71.
[70] Alexiou C, Langley SM, Dalrymple-Hay MJ, et al. Open commissurotomy for critical isolated aortic stenosis in neonates. Ann Thorac Surg 2001;71:489—93.
[71] Brown JW, Ruzmetov M, Vijay P, Rodefeld MD, Turrentine MW. Closed transventricular aortic valvotomy for critical aortic stenosis in neonates: outcomes, risk factors and reoperations. Ann Thorac Surg 2006;81:236—42.
[72] Miyamoto T, Sinzobahamvya N, Wetter J, et al. Twenty years experience of surgical aortic valvotomy for critical aortic stenosis in early infancy. Eur J Cardiothorac Surg 2006;30: 35—40.
[73] Sreeram N, Kitchiner D, Williams D, Jackson M. Balloon dilatation of the aortic valve after previous surgical valvotomy: immediate and follow up results. Br Heart J 1994;71: 558—60.
[74] Bacha EA, Daves S, Hardin J, et al. Single-ventricle palliation for high-risk neonates: the emergence of an alternative hybrid stage I strategy. J Thorac Cardiovasc Surg 2006;131:163e2—71e2.
[75] Caldarone CA, Benson LN, Holtby H, Van Arsdell GS. Main pulmonary artery to innominate artery shunt during hybrid palliation of hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 2005;130:e1—2.
[76] Pizarro C, Derby CD, Baffa JM, Murdison KA, Radtke WA. Improving the outcome of high-risk neonates with hypoplastic left heart syndrome: hybrid procedure or conventional surgical palliation? Eur J Cardiothorac Surg 2008;33:613—8.
[77] Akinturk H, Michel-Behnke I, Valeske K, et al. Hybrid transcatheter-surgical palliation: basis for univentricular or biventricular repair: the Giessen experience. Pediatr Cardiol 2007;28:79—87.