Palliative procedures for congenital heart defects

Chirurgies palliatives dans les cardiopathies congénitales

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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.

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Abbreviations: HLHS, Hypoplastic left heart syndrome.
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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 (pulmonary banding and Norwood procedure); twoièmement, diminution du flux artériel pulmonaire en cas de débit pulmonaire augmenté (cerclage pulmonaire et en incluant la procédure de Norwood), troisièmement, enrichissement de 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 concomitante la chirurgie et le cathétérisme interventionnel semblent très prometteuses.

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. 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 3. Central 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.
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
Palliative procedures

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].

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, polyplenia, 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 interstitial 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 centronuclear 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


Palliative procedures 557


