Surgical strategies for patients with congenital heart disease and severe pulmonary hypertension in low/middle-income countries

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ABSTRACT

In this review, we discuss specific surgical strategies that are used in patients with congenital heart disease and severe pulmonary arterial hypertension. Our own experience, with the use of unidirectional valved patches in managing these patients, is also discussed in detail.

INTRODUCTION

It is estimated that globally about 600 000 babies are born with significant congenital heart disease (CHD) every year[1]—an incidence of about 8–10 per 1000 live births, which remains similar across countries and among races.[2 3] An estimated 4%–15% of patients with CHD eventually develop significant pulmonary arterial hypertension (PAH).[4] Early repair of these congenital heart defects usually prevents the development of pulmonary vascular disease. However, the availability and access to specialist healthcare facilities is skewed in favour of developed countries, and only about 2%–15% of patients with congenital cardiac lesions actually receive timely surgical intervention.[1] Therefore, it is not uncommon to encounter patients with CHD with severe PAH beyond infancy/childhood in many parts of the world.[5] Management of these so-called late-presenting patients with advanced pulmonary vascular disease poses significant challenge to the healthcare providers. In this review, we will discuss some basic problems and surgical solutions, including our own innovations with the use of unidirectional valved patches (UVPs) to address these issues.

PATHOPHYSIOLOGY OF PAH IN CHD

Development of PAH in CHD is multifactorial. Volume and pressure overload in the pulmonary circulation is a trigger for unfavourable vascular remodelling. Increased pressure in the pulmonary arteries (PAs) leads to abnormal shear stress, circumferential wall stretch and endothelial cell dysfunction. The resultant imbalance in expression of vasoactive mediators such as endothelin-1, prostacyclin, nitric oxide, transforming growth factor-β1, vascular endothelial growth factor and fibroblast growth factor-2 culminates in smooth muscle hypertrophy and proliferation, increased intracellular matrix deposition, vasoconstriction, inflammation, thrombosis, impaired apoptosis and fibrosis.[1 6–8] As a result, there is progressive increase in the pulmonary vascular resistance (PVR).

The prevalence of PAH in patients with CHD depends on size and location of the defect and the duration of illness.[9] Pretricuspid defects like atrial septal defects (ASDs) or unobstructed anomalous pulmonary venous return lead to pulmonary volume overload only, whereas post-tricuspid defects like ventricular septal defect (VSD), patent ductus arteriosus, atrioventricular septal defects, truncus arteriosus lead to volume as well as pressure overload on the pulmonary circulation. Thus, severe PAH and Eisenmenger's syndrome are predominantly seen in post-tricuspid shunts. In general, pulmonary vascular remodelling is reversible if the defect is closed/treated in infancy, that is, before the first birthday, though associated transposition or Down's syndrome may prevent favourable outcomes even in 6 months old infants. If the cardiac defect is corrected within a few months of age, the changes reverse completely, and the patient's PVR drops to normal gradually (surgical cure). If the surgery is delayed beyond 2 years of age, the changes are only partially reversible, and PVR decreases from the preoperative level, but may not normalise.[10] Correction of the defect in the presence of established irreversible PAH is detrimental and usually leads to accelerated disease progression and onset of right heart failure. If the defect is left uncorrected, it leads to progressive increase in PVR, resulting in Eisenmenger's syndrome defined as pulmonary hypertension at systemic level, due to high PVR index (PVRI; 10 Woods unit/m^2) with reversed (right to left) or bidirectional shunt through a septal defect.[11–14]

CONCEPT OF OPERABILITY IN PATIENTS WITH HIGH PVRI

It is a known fact that correction of a septal defect in patients with irreversible PAH is often associated with worse prognosis than leaving it uncorrected.[13] Lung biopsy was used in the past to assess operability based on the histopathological changes in the pulmonary vasculature.[15 16] However, the role of lung biopsy is limited in clinical practice because it is invasive, is difficult to perform, has its own complications and provides information about only one randomly selected area of the lung, and sometimes, there is poor correlation between the changes on histopathology and reversibility of PAH.[17]

Demonstration of enlarged left-sided chambers and increased PA flow on echocardiography is a useful criterion to identify operable lesions, but

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echocardiography is not accurate enough for assessing patients with borderline operability.

Right heart catheterisation is the gold standard for measurement of haemodynamic parameters in patients with doubtful operability. However, there are few evidence-based guidelines for this subset of patients (box 1). Lopes and O'Leary[18] suggested that both PVR and the ratio of PVR to systemic vascular resistance (SVR) and their response to acute vasodilator challenge should be considered to decide on operability.

However, one should keep in mind that there is no consensus as to whether vasoreactivity testing is accurate enough to discriminate between patients who will or will not have a good surgical outcome. Giglia and Humpl[19] reported that precise values of haemodynamic measures of pulmonary vascular disease to determine the level of risk of death or persistent PVR following biventricular repair are unknown, and it is unclear, which preoperative pulmonary haemodynamic parameters correlate best with outcomes, and what is the influence of individual patient factors such as cardiac lesion type and genetic predisposition on these parameters. Further, recommendations of Lopes and O'Leary[18] do not apply to patients with single ventricle physiology who should ideally have near normal levels of PVR. In patients with transposition of great arteries (TGA), accurate calculation of PVR is difficult and unreliable, owing to high PA saturation, low pulmonary arteriovenous oxygen content difference, increased bronchial blood flow and limitations in using the standard cardiac catheterisation data.[20]

As per Grown-Up Congenital Heart guidelines,[21] patients with ASD with significant interatrial shunt (pulmonary blood flow/systemic blood flow (Qp/Qs) >1.5 or signs of right ventricular volume overload) and PVR <5 Wood units should undergo ASD closure (possibly percutaneously) regardless of symptoms (recommendation class I, level of evidence B). Patients with ASD, Qp/Qs >1.5 and PVR ≥5 Wood units, but less than two-thirds of SVR, or pulmonary artery pressure (PAP) less than two-thirds systemic pressure (baseline or when challenged with vasodilators, preferably nitric oxide, or after PAH-specific therapy) should undergo ASD closure (recommendation class IIb, level of evidence C). Similarly, patients with VSD are ideal candidates for closure if Qp/Qs is >1.5, and PVR is normal (<5 Wood units). They should be considered for closure (recommendation class IIb, level of evidence C) when there is still net left-to-right shunt (Qp/Qs >1.5) present, and PAP or PVR is less than two-thirds of systemic values (baseline or when challenged with vasodilators, preferably nitric oxide, or after PAH-specific therapy).

Circulating endothelial cells have been studied as a promising non-invasive marker for assessing the operability of patients with PAH. Circulating endothelial cell count has been shown to be significantly raised in patients with CHD with irreversible PAH postsurgery.[22] Further studies are required to validate their use in clinical practice.

In addition to the above criteria, in our practice, we subject a patient to closure of the septal defects if there is more than 20 mm Hg difference between aortic and PA diastolic/mean pressure on oxygen (in the absence of significant pulmonary regurgitation), along with Qp/Qs >1.5:1 and basal saturations not <95%. Also it is important to look at the total picture that includes clinical evaluation, chest X-ray and ECG.

Box 1 Cardiac catheterisation criteria of operability in patients with left-to-right shunts

A. Wood's criteria[12]
PVRI <10 Woods unit/m^2 with Qp/Qs ratio of at least 2:1[12]
B. Lopes and O'Leary[18]
Baseline PVRI <6 Wood units/m^2 associated+PVR:SVR ratio <0.3: a vasoreactivity test: not needed
Baseline PVRI 6–9 Wood units/m^2 associated+PVR:SVR ratio <0.3–0.5: a vasoreactivity test: needed
i. PVRI drops by 20%: operable
ii. PVR:SVR ratio drops by 20%: operable
iii. Final PVRI <6 Wood units: operable
iv. PVR:SVR ratio <0.3: operable
C. More than 20 mm Hg difference between aortic and PA diastolic/mean pressure on oxygen with Qp/Qs >1.5:1 and basal saturations not <95%.
PA, pulmonary artery; PVR, pulmonary vascular resistance; PVRI, pulmonary vascular resistance index; Qp, pulmonary blood flow; Qs, systemic blood flow; SVR, systemic vascular resistance.

ROLE OF MEDICAL MANAGEMENT OF PAH ASSOCIATED WITH CHD

Medical management has a definite role in the management of these patients as a useful adjunct to surgery. A detailed discussion of this is outside the scope of this review. Of importance is that several case reports have shown variable success of pretreatment with prostanoids, endothelin receptor antagonists and phosphodiesterase-5 inhibitors in preparing equivocal or inoperable patients for surgical correction.[23] Such a 'treat and close strategy' appears to be promising. However, the evidence and experience are limited to a few case reports, and only short-term follow-up is available. Further, choice of agent, the dose, duration of presurgical and postsurgical treatment is not well established. Current evidence does not favour the adoption of this strategy as routine.

SURGERY FOR PAH ASSOCIATED WITH CHD

Surgical closure of late-presenting septal defects in patients with severe PAH is fraught with early and late complications associated with long-standing PAH. The pulmonary vasoconstrictive effects of cardiopulmonary bypass and exacerbation of pulmonary vasoconstriction in early postoperative period are well known.[24 25] Institution of cardiopulmonary bypass, infusion of protamine and other factors such as the use of cardiotomy suckers during surgery may result in release of vasoconstrictive substances like thromboxane A2 and catecholamines, which results in acute pulmonary hypertensive crisis.[24–28] Pulmonary hypertensive crisis, acute congestive heart failure and acute respiratory failure are the principal causes of postoperative death in such patients.[29] In developed countries, advanced pharmacological support like inhaled nitric oxide or endothelin receptor antagonist and advanced mechanical support in the form of extracorporeal membrane oxygenation are used in the immediate postoperative period to tide over the phase of acute pulmonary hypertensive crisis, which are essentially episodic and slowly wane over time. Such facilities are not universally available or affordable in many parts of the world. Surgical options in such patients are limited (box 2). All these options have their own merits and demerits, and are briefly discussed here.

Small interatrial communication

Leaving behind a small communication at the atrial level serves the purpose of decompressing the right ventricle by acting as a

Box 2 Strategies for patients with congenital heart disease with pulmonary arterial hypertension and borderline operability

Inhaled nitric oxide
Extracorporeal membrane oxygenation support
Pulmonary artery banding and second-stage closure of septal defect
Leaving a communication at the atrial level
Partial closure of the septal defect
Unidirectional valved patch closure of the septal defect
Heart–lung transplantation
Pott's shunt

pop-off to allow a right-to-left shunt during episodes of elevated right-sided pressure. It effectively prevents acute right ventricular failure, although at the cost of slight systemic desaturation. Ease of creating this communication makes it an attractive option; however, the degree of shunting through this communication and the long-term patency are at best unpredictable. Should the right-sided pressures normalise over a period of time, this communication may rarely allow a left-to-right shunt, and may be a potential site for paradoxical embolism. Rarely, it may be required to close these communications by percutaneous intervention.

Partial closure of the septal defect

Pop-off for allowing a right-to-left shunt during episodes of acute pulmonary hypertensive crisis may similarly be provided at the level of the septal defect itself by using a fenestrated patch to close the defect incompletely. In the event of elevation of right-sided pressures, there is a right-to-left shunt through the fenestration. However, if the PA pressures fall at follow-up, a left-to-right shunt may ensue, necessitating a percutaneous intervention/surgical intervention to close the defect. Additionally, when a VSD is closed with a fenestrated patch, there is definite risk of endocarditis.[30] For these reasons, we do not use the fenestrated patch at our centre. The size and shape of fenestration varies with surgeon.

UVP for closure of septal defects

Basis

The principles of UVP have been discussed in detail in our prior publication.[31] UVPs are aimed at creating a one-way mechanism at the level of the ventricular septum that permits the blood to flow from the right ventricle to the left ventricle when the pressure in the former exceeds the pressure in the latter. This prevents acute right ventricular failure and maintains cardiac output. Subsequently, when right-sided pressures gradually fall in the postoperative period, the gradient across the valve will fall, and it would simply close, preventing any left-to-right shunt. Multiple techniques have been used to create this valve mechanism by various researchers with similar results.[29–34]

Techniques and variations

Zhou *et al*[29] first described UVP for closure of VSD in patients with severe PAH. They fashioned the UVP from a patch with an eccentric fenestration. The fenestration was covered with a piece of pericardium sutured on the left ventricular side of the Dacron patch on three edges leaving one edge unattached. This unattached edge provided a small opening through which a right-to-left shunt could take place, if required (figure 1). Novick *et al*[32] were the first to modify this technique. Instead of pericardium, they used two patches of Dacron. After sizing the VSD patch, they created a fenestration in its centre. A 4–5 mm fenestration was created for patients below 20 kg and above this, a 6 mm fenestration was deemed suitable. Another patch larger than the size of the fenestration was sutured around the latter. They further modified the technique in 2005, where the aortic annulus diameter was used as a guide to the size of the fenestration.[33] The VSD patch was oriented to ensure that the valve opened towards the left ventricular apex to avoid subaortic obstruction. Novick *et al*[32 33] recommended the use of this

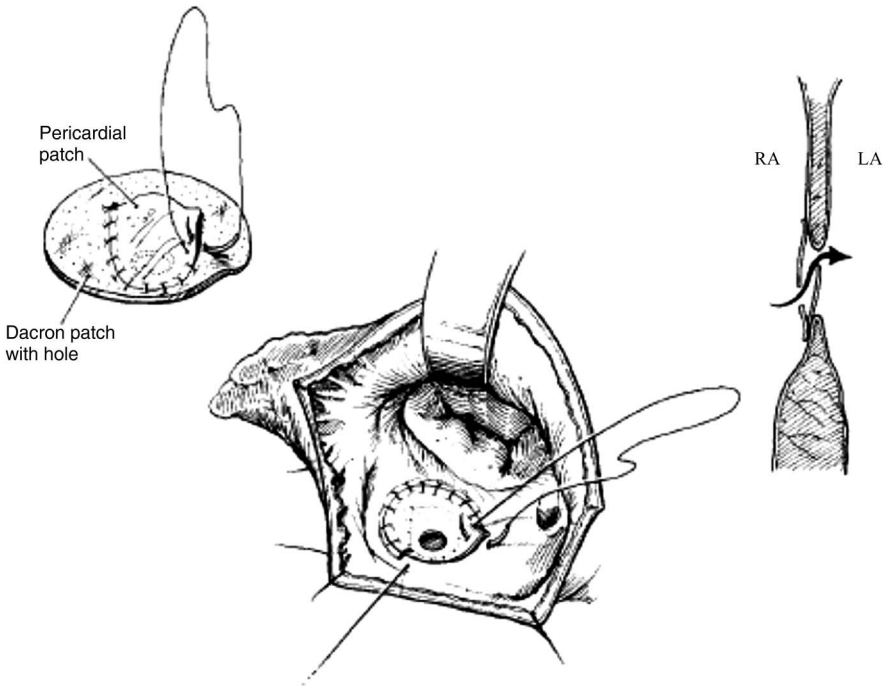

Figure 1 Zhou's technique. Three sides of the pericardial patch are attached and one side is open to function as a valve. The pericardial valve flap is placed on the left side of the defect. This allows the blood to flow from the right atrium (RA) to the left atrium (LA) or across the ventricular septum when used to close the ventricular septal defect. (Reproduced with permission from Zhou *et al*,[29] Copyright Society of Thoracic Surgeons.)

technique to improve the quality of life even for patients with Eisenmenger's syndrome as an alternative to heart–lung transplantation.

In 2007, Zhang *et al* used an aortic homograft with its attached mitral leaflet as the UVP.[34] The aortic valve homograft was incised, and two of its cusps were removed, preserving the mitral leaflet and the third aortic cusp. The mitral leaflet formed the UVP.

At the All India Institute of Medical Sciences (AIIMS), we have developed a simple technique (figure 2) that was first reported in 2007.[35] After inspecting the VSD/ASD, a patch of Dacron that is 1.5 times longer, but of same width, is fenestrated using a 4 mm punch. The patch is then folded on itself and sutured to the edges of the defect, placing the flap on the systemic ventricular/atrial side. The UVP is oriented downwards to prevent left ventricular outflow obstruction during systole. The advantages of the 'AIIMS technique', as detailed in our prior publications, are that it is simple, inexpensive and easily reproducible, does not require two patches or pericardium or homograft to prepare and is less time-consuming to prepare, thus reduces ischemic arrest time.[35–37]

Management during and after surgery

We pursue aggressive systemic and pulmonary vasodilatation using pharmacological manipulation. Prior to CPB, intravenous phenoxybenzamine 0.5–1 mg/kg is started. Cardiopulmonary bypass (CPB) is discontinued on elective support of dobutamine 5–10 μg/kg/min, nitroglycerin or nitroprusside 0.5–2 μg/kg/min or the phosphodiesterase inhibitor milrinone. Intraoperative transoesophageal echocardiography and postoperative transthoracic echocardiography is always performed to assess shunting across the UVP. In the operating room and in the intensive care unit, moderate hypocarbia is employed for pulmonary vasodilatation. We always aim to wean these patients off the mechanical respiratory support as soon as possible. Once oral intake is started, oral ACE inhibitor or oral phenoxybenzamine is administered, and the inotropes are weaned off. Sildenafil may be administered depending on the degree of PAH. As of now, we do not administer bosentan. Anticoagulants/aspirin are not administered.

After discharge from the hospital, follow-up consists of clinical examination, measurement of saturation using pulse oximetry and transthoracic echocardiography at regular intervals to estimate the PA pressure, assess the right ventricular function and demonstrate the degree of right-to-left shunting and cardiac catheterisation.[35]

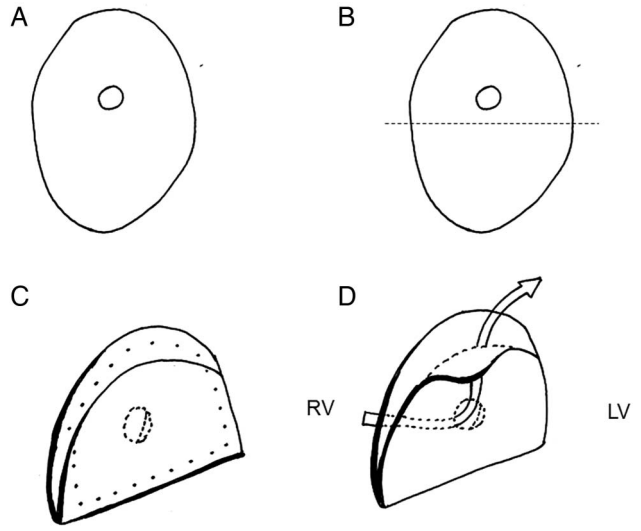

Figure 2 AIIMS technique of unidirectional valved patch. (A) Appropriate-size Dacron patch is selected and a fenestrated (B) patch is folded upon itself at the dotted line; (C) the patch is sutured (outer dots) to close the defect; (D) mechanism of right-to-left shunting through the patch—in-situ (arrow) is shown. Suture line has been removed for clarity. AIIMS, All India Institute of Medical Sciences; LV, left ventricular; RV, right ventricular. (Reproduced with permission from Talwar *et al*.[31] Redrawn with permission from Choudhary *et al*.[35] Copyright American Association for Thoracic Surgery and Elsevier.)

Results of UVP closure of septal defects

We have successfully used the 'AIIMS technique' for correcting a wide range of septal defects with borderline operability, including isolated VSD,[36 37] in patients with truncus arteriosus,[38] in patients with TGA with VSD,[39] in patients with aortopulmonary window with VSD and for closure of ASD in two patients with total anomalous pulmonary venous drainage.[40] We have successfully demonstrated the favourable effect of UVP on the immediate,[35] early[36 38 39] and mid-term clinical outcomes[37] and haemodynamic parameters[36] in patients with borderline operability.

Recently, we have reported a 12-year-old patient with truncus arteriosus who underwent successful surgical repair using UVP to close the VSD to act as a safeguard in the event of postoperative pulmonary hypertension and right ventricular decompensation.[38] This was the first instance of a patient with truncus arteriosus, in which, UVP was used beyond first decade of life.

We have also used, with acceptable early results, UVP in management of patients with dextro-TGA with VSD and PAH with acceptable early results, for which, other option would have been palliative arterial switch operation (ASO) leaving the VSD open.[39] Between July 2009 and February 2011, six patients with TGAs, VSD and severe PAH (mean age 39.8 +47.4 months, range 8–132 months), weighing 10.7+9.2 kg (median 8.6, range 4.3–29 kg), underwent ASO with VSD closure using our technique of UVP. Mean PA systolic pressure before the operation was 106+12.7 mm Hg (range 95–126 mm Hg) and PVR was 9.5+4.22 units (range 6.6–17.1 Wood units). There were no deaths. All patients had a postoperative systemic arterial saturation of more than 95%, although there were frequent episodes of systemic desaturation due to right-to-left shunt across the valved VSD patch (as seen on transoesophageal and transthoracic echocardiograms). Mean follow-up was 10+7.6 months (range 1–22 months). At most recent follow-up, all patients had systemic arterial saturation of more than 95% and no right-to-left shunt through the VSD patch. In one patient, the follow-up cardiac catheterisation showed a fall in PA systolic pressure to 49 mm Hg.

Mid-term clinical results of our UVP technique for closure of VSD have been gratifying.[37] Between January 2006 and December 2010, 17 patients (age 2–23 years, median 9 years) with a large VSD and severe PAH underwent VSD closure with UVP. Preoperative mean PVRI was 10.9±2.2 Wood units, and mean preoperative systemic saturation was 93.4±2.6%. Shunt was bidirectional in 15 patients and predominantly right to left in 2. After VSD closure, intraoperative transoesophageal echocardiography (figure 3) revealed a right-to-left shunt across the patch in three patients 2, 7 and 9 years of age who had preoperative PVRI of 9.5, 9.8 and 11.1 Wood units, respectively. There were no inhospital deaths, and all patients had uneventful recovery. Mean follow-up was 30±14.7 months, and all patients

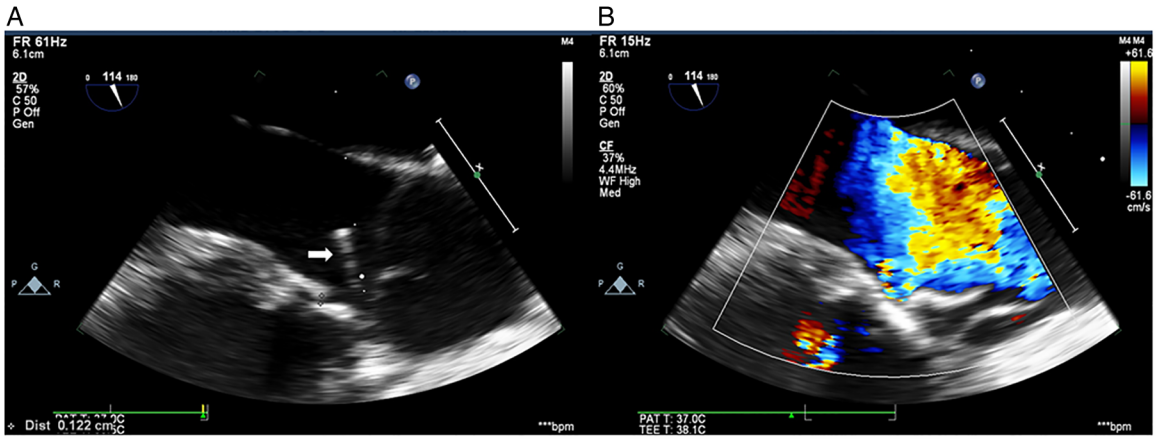

Figure 3 Intraoperative transoesophageal echocardiography showing right-to-left shunt across the valved patch (arrow) in a 9-year-old patient who underwent closure of a ventricular septal defect.

were well without cyanosis. Echocardiography showed no shunt across the patch, and all patients had systemic saturation >95%.

We have also assessed, and found acceptable, the haemodynamic outcomes of UVP closure of VSD in patients with VSD and PAH.[36] From January 2006 to January 2012, 20 patients with VSD with PAH and a PVRI >8 Wood units underwent VSD closure with a UVP. Although our clinical follow-up was 100% complete, only 13 patients agreed to follow-up cardiac catheterisation, and were studied at a mean follow-up of 34.7 ±18.6 months (range 2–56). The mean age of these 13 patients was 8.5±4.4 years (range 2–19), and the mean preoperative systemic saturation was 94.1%±3.4% (range 87–99). The mean preoperative PA systolic pressure was 96.2±13.6 mm Hg (range 75–115), and the mean preoperative PVRI was 10.0±2.1 Wood units (range 8.0–15.1, median 9.3). At follow-up cardiac catheterisation, the mean systemic saturation had increased to 98.92%. The PVRI had decreased significantly to 5.8±2.1 Wood units (p=0.02). A significant decrease was seen in the PA systolic, diastolic and mean pressures (p=0.000), and none of the patients had severe PAH. No patients died, and all patients were in New York Heart Association class I.

Other groups have also demonstrated favourable results with the use of UVP,[29 32–34] which is summarised in table 1.

Disadvantages of UVP

In the technique described by Novik *et al*,[32 33] the flap-valve flow is not in the direction of left ventricular flow, and it appears to interfere with flow when the left ventricular pressure starts to increase. Patients, in whom, the PVR continues to remain high or increases in postoperative period, presence of a one-way valve may be deleterious. In the early postoperative period, a UVP will allow only right-to-left shunt compared with bidirectional flow that occurs through a fenestration; this may lead to significant systemic desaturation and prolonged intubation. Over the long term, persistent pulmonary hypertension may lead to early development of cyanosis compared with non-operated case as described in one patient by Afrasiabi *et al*.[30]

The results of our haemodynamic study[36] show that PAPs decreased significantly, but they were not yet normalised. It is also a matter of speculation only that to what extent the pulmonary vascular changes at this level of PVRI are reversible. Hence, a simple assumption from these results that survival of these patients should not be different than those patients without significant PAH would be fallacious. For such an inference, a randomised study with preoperative and postoperative analyses and exercise testing would be required. Results of these studies cannot be extrapolated to patients with Eisenmenger's syndrome. Currently, we would offer a UVP to all patients with a high PVRI unless they have evidence of established Eisenmenger's syndrome.

PA banding and second-stage closure of VSD

PA banding is described as a means to reducing large left-to-right shunts and improving survival.[41] PA banding is associated with considerable mortality in immediate post-operative period, particularly in smaller infants. It can also lead to significant residual PA deformity.

Batista *et al* have reported complete regression of pulmonary vascular changes 1 year following PA banding in a 19-year-old patient with VSD and grade IV pulmonary vascular changes.[42 43] As a second definitive procedure, the septal defects were closed at a second surgery. They performed first stage of similar procedure in six more patients with similar results. They postulated that PA banding results in increase in right-to-left shunt and decreases aortic saturation with resultant decrease in PA saturation. Lower PA saturation was responsible for dilatation of pulmonary vascular bed and a decrease in PVR, causing regression of fixed pulmonary lesions. They also postulated that mutually opposite oxygen sensors are present in the PA vascular tree and the alveolar tree. Oxygen delivered through alveoli decreases PA vascular resistance and oxygen delivered through PA increases PA vascular resistance.

Heart–lung transplantation

Heart and lung transplantation is a potential treatment option for patients with severe PAH and Eisenmenger's syndrome due to CHD, but indications are not clear because there is a very slow progression of disease in patients with PAH due to CHD, and natural history of patients with PAH due to CHD is far better than that of patients with idiopathic and other forms of PAH. Donor availability is a problem; centres performing this operation are extremely limited in the low/middle-income countries, and above all, heart transplant is not a benign procedure, and entails lifelong monitoring and immunosuppression with its attendant complications. Indications for transplant in patients with PAH due to CHD may be summarised as highly symptomatic patients with short life expectancy, refractory right heart failure, severe hypoxaemia and established Eisenmenger's syndrome.

Table 1 Summary of results obtained with the use of unidirectional valved patch

Author	Year	No	Age (years)	Preoperative SpO_2 (%)	Preoperative PASP (mm Hg)	PVRI (Wood units)	Qp:Qs	PASP/AOSP	Early deaths	Postoperative SpO_2 (%)	Early R–L shunt	Postoperative PASP (mm Hg)	Follow-up in years (mean)	Late deaths	No: with R–L shunt at last follow-up
Zhou *et al*[29]	1995	24	5–28 (15.8)	82–97 (87±4)	80±12	8–32 (16±7)	0.5–2 (1.2±0.5)	1–1.3 (1.1±0.1)	2 (8.3%)	96±1	12	56±18	0.3–3 (1.1)	Nil	Nil
Novick *et al*[32]	1998	18	1.5–15 (5.7±3.9)	89±5	105±16	11.4±4.1	1.4±0.41:1	NA	Nil	96±2	*	42±14	Details NA	1	Details NA
Novick *et al*[33]	2005	91	0.5–17 (4±3.1)	90±4	NA	10.5±4.9	1.6±0.8	0.96±0.12	7 (7.7%)	±	8	NA	0.1–7.6	7/78# (8.9%)	4
Zhang *et al*[34]	2007	27	6–31 (15±5.6)	89±1	81±12	(2.4±2.5)	0.5–2 (1.2±0.5)	1.05±0.1	2 (7.4%)	95±2	10	68±15	0.5–10 (4.1±1.3)	Nil	2
Talwar et al[36]	2014	13/20†	2–19 (8.5±4.4)	94±3.4	96±13	10.0±2.1*	1.5±0.6	NA	Nil	96±2	3	39±11	0.1–4.6 (2.9±1.5)	Nil	Nil

*Fluctuations were observed.
#Follow-up data available only for 78 patients.
†Postoperative catheterisation data available for 13 out of 20 patients.
AIIMS, All India Institute of Medical Sciences; AOSP, aortic systolic pressure; NA, data not available; PASP, pulmonary artery systolic pressure; PVRI, pulmonary vascular resistance index; Qp, pulmonary blood flow; Qs, systemic blood flow; R–L, right to left; SpO_2, oxygen saturation.

Pott's shunt

A direct anastomosis between the descending aorta and the left PA (Pott's shunt) has been used as a temporary measure to alleviate the right ventricular failure resulting from progression of PAH late after closure of septal defects.[44] The aim is to provide a right-to-left shunt and reduce afterload to the right ventricle. At best, this strategy is used to buy time for an eventual lung or a heart–lung transplant.

CONCLUSIONS

Despite our understanding of the mechanisms of PAH, predicting operability in CHD with PAH is an 'in-exact science'. More work is needed in this field. In the low/middle-income countries, we continue to see patients with CHD and PAH, and we must continue to develop strategies to address these patients. The UVP appears to be an attractive option. However, we need long-term data before the widespread application of this technique. In patients with established Eisenmenger's syndrome, its role remains questionable.

Contributors ST, SKC and BA are members of the surgical team who have developed this technique. ST: performed surgeries, detailed review of the literature, drafted the manuscript and finally submitted the manuscript. VKK: review of literature and drafting of manuscript. SKC and BA: performed surgeries, critically reviewed the manuscript. SKG, SR, AS, RJ and SSK: evaluation of patients, critical review of manuscript.

Competing interests None declared.

Provenance and peer review Commissioned; externally peer reviewed.

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