Primary correction of total anomalous pulmonary venous return with a modified sutureless technique

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INTRODUCTION

Total anomalous pulmonary venous return (TAPVR) is a rare congenital malformation occurring in 5.9–7.1 newborns per 100,000 live births [1], thus constituting nearly 0.8% of all congenital heart defects. It can occur in isolation or in conjunction with various malformations such as atrial isomerism and functional univentricular hearts. If left untreated, the majority of children succumb in the first few months of life. Thanks to improvements in the surgical technique and neonatal intensive care, in particular, the availability of nitric oxide and refinements in extracorporeal life support, the results of surgical repair have continued to improve with current early survival ranging from 82 to 98% [2–5].

However, there is an ongoing risk following a conventional repair with a late morbidity of 5–18%, most frequently attributable to pulmonary venous obstruction (PVO) and intractable pulmonary hypertension (PHT). Traditionally, 40–66% of patients with postoperative PVO die eventually, in spite of treatment [1, 2, 6–9].

Groups led by Najm [10] and Lacour-Gayet [11] have described a sutureless repair technique for post-repair PVO, in which there is no direct anastomosis between left atrial and pulmonary venous tissue. When used for primary repair of TAPVR, sutureless repair has demonstrated improved survival and reduction in the need for reintervention for recurrent PVO [6]. Potential advantages of this technique that could favourably impact the pathogenesis of recurrent PVO include the absence of mobilization and distortion of the collector, the absence of sutures on often small pulmonary veins, limited reactive intimal proliferation and the absence of an anastomotic fibrous scar. It is hence assumed that a sutureless repair, when impeccably performed, results in optimal characteristics for a given vein, at the orifice as well as all through its course up stream. This philosophy has been our rationale for the use of this technique for primary correction of all supra- as well as infracardiac TAPVR. We present our technique of performing this repair, under moderate hypothermia, while minimizing the duration of circulatory arrest.

MATERIALS AND METHODS

Patient demographics

Data on seven consecutive patients (2009–11) with supra- or infracardiac TAPVR undergoing primary sutureless repair at our...
institution were retrospectively analysed. The study protocol was reviewed and approved by the Institutional Committee on Clinical Investigation. This cohort included all patients who underwent repair of infracardiac (three) and supracardiac (four) TAPVR during this period.

All three patients with infracardiac TAPVR were obstructive, with the venous confluence being small in size, measuring between 3 and 3.1 mm, in two cases with a substantial downstream narrowing of the vertical vein of 1.6 mm.

Four patients had supracardiac TAPVR, none of them showing obstruction in the vertical vein. Despite being nonobstructive, the vertical veins had a median diameter of 3 mm in this group. Patient demographics were as shown in Table 1. One of the patients with supracardiac TAPVR had a single ventricle morphology with transposition of the great arteries and pulmonary atresia and underwent single ventricle palliation in preparation for a subsequent Fontan pathway.

**Surgical technique**

After median sternotomy, moderately hypothermic (median temperature 28°C) CPB is instituted using aorto-bicaval cannulation. The aorta is cross-clamped and cardiac arrest achieved using antegrade cold blood cardioplegia. The apex of the heart is luxated towards the right shoulder, using stay sutures placed at the base of the heart (Fig. 1). Without any posterior attachment of the left atrium to the pulmonary veins, the heart can be generously dislocated towards the right shoulder, exposing the posterior pericardial surface. The parietal pericardium overlying the pulmonary venous confluence is slit open and reflected away from the anterior aspect of the pulmonary venous collector, the pulmonary veins as well as the connecting vein. A pericardial well is thus created. The dissection encompasses the anterior two-thirds of the circumference of the collector. The posterior one-third is not disturbed, so as to avoid injuring the mediastinal pleura and entering the pleural space.

The right lateral aspect of the pericardial well is dissected along the interatrial groove and the course of the right upper and lower pulmonary veins draining into the collector is identified (Fig. 2).

The left atrium is opened using two reference points: the base of the left atrial appendage and the inferior vena cava (IVC). The incision along these two points is thus like a diagonal across the posterior wall of the left atrium. A narrow strip of the left atrial wall is excised to ensure a round and wide opening.

The superior margin of the atroventricular anastomosis is performed first, starting from the far end, at the base of the left atrial appendage. The suture is run towards the right, up to under the superior vena cava (SVC). The thaws of suture run inside out on the pericardium and outside in on the left atrium (Fig. 3). An initial mattress suture on the left atrial wall ensures an eversion of the anastomosis.

The inferior suture line is then constructed, interrupting and running backwards a couple of times to evenly correct the discrepancy in the circumference of the left atrium as well as the pericardial well (Fig. 4).

**Table 1:** Patient demographics

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<tr>
<td>N</td>
<td>7</td>
</tr>
<tr>
<td>Age* (days)</td>
<td>6.4 (1–14)</td>
</tr>
<tr>
<td>Weight* (kg)</td>
<td>2.9 (2.3–3.2)</td>
</tr>
<tr>
<td>Male/female</td>
<td>2/5</td>
</tr>
<tr>
<td>Supracardiac (unobstructed) TAPVR</td>
<td>4</td>
</tr>
<tr>
<td>Infracardiac (obstructive) TAPVR</td>
<td>3</td>
</tr>
<tr>
<td>Univentricular/biventricular hearts</td>
<td>1/6</td>
</tr>
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*Values are in median and range.
The heart is repositioned into the pericardial cavity and the remaining anastomosis performed at the interatrial groove. Before performing the right lateral anastomosis, the pulmonary venous confluence is incised along its long axis during a short (3–5 min) phase of circulatory arrest. A small strip of the anterior wall of the collector is excised to prevent the recoil of the lateral wall. The incision is extended across the individual pulmonary venous ostia, particularly if it appears that a discrete stenosis could exist. The vertical vein is interrupted using a ligaclip or a ligature (Fig. 5).

The atrial septal defect is closed either directly when possible without creating any tension, or using an untreated autologous pericardium through a small right atriotomy. A 2-mm fenestration is often created in the atrial septum.

In the present cohort, the atrial septal defect was closed directly in four patients and with an untreated autologous pericardium in three patients. A 2-mm fenestration was performed in four patients (Fig. 6).

Postoperative protocol

All patients were anticoagulated therapeutically in the early postoperative period. They were switched to acetylsalicylic acid at the time of discharge, to be continued for 6 weeks.
repair for the treatment of post-repair discrete PVO is an elegant solution that has been the focus of efforts aimed at improvement. Sutureless repair techniques, such as those described by Najm [10] and Lacour-Gayet [11], have reported an incidence of postoperative PVO ranging from 1 to 18%, with a larger series showing an incidence of 6–9% [2, 6]. Various hypotheses about the etiopathogenesis of postoperative PVO and PHT revolve around inherent hypoplasia of the pulmonary venous architecture, discrete pulmonary vein stenosis or atresia, absence of confluence, discrete PVO as a consequence of direct suturing near a very small pulmonary venous orifice, or reactive/inflammatory thickening of pulmonary venous vasculature due to an obstructive/non-compliant connection between the collector and the left atrium. Occurrence of postoperative PVO has been associated with young age at initial surgery, infracardiac TAPVR and pre-existing PVO. Preoperative PVO, which is manifest as a small or absent venous confluence, and diffuse pulmonary venous narrowing, are important risk factors for the development of postoperative PVO and eventual death [2, 12–14].

Many groups speculate that PVO is a progressive disease. Seale et al. [1] have shown that discrete obstruction of pulmonary veins leads to pulmonary venous remodelling, smaller pulmonary veins and even the development of pulmonary vein atresia. If this occurs in gestation, it may result in generalized hypoplasia of the pulmonary vascular bed, with arterialization of the pulmonary veins and pulmonary lymphangiectasia. Pathological studies of obstructive infracardiac TAPVR have shown that in extreme forms, the entire pulmonary venous system may be small at birth and may be associated with lymphangiectasia. These are the patients who present early in a clinically poor shape. During gestation, since the lymphatic system often regresses after approximately 20 weeks, it has been speculated that in those cases where pulmonary lymphangiectasia is seen, foetal PVO could have existed as early as before 20 weeks of gestation [15].

Hancock Friesen et al. [16] suggested that since none of the current perioperative or operative strategies have had an impact on the incidence of post-repair PVO, such a postoperative course may reflect an underlying predisposition.

However, we believe that primary sutureless repair could be an answer for a subset of these patients for whom the pathogenesis of post-repair PVO is attributable to the handling of pulmonary venous tissue.

This belief has been substantiated by reports that demonstrate a faster rate of decline of right ventricular systolic pressure in the...
primary sutureless repair group when compared with conventional repairs [6]. However, the same group has also reported the development of unilateral pulmonary vein occlusion after sutureless repair with one death due to PHT. This clinical course points towards a progressive disease pathology, which is not likely to be addressed with the current surgical strategies.

Survival

While Yun et al. [17], in a mixed cohort involving conventional and sutureless repairs, have reported a 5-year freedom from reoperation or death of 49%, Karamlou et al. [5], in a large series from Toronto, have reported an operative 5-year survival of 97%. A population-based multicentric study from Europe showed a 3-year operative survival of 85% [1]. Yong et al. [3] from Melbourne reported a 20-year survival of 83%, with an early mortality remaining unchanged at around 11% over a 3-decade period. Notwithstanding our very small numbers, we have no early or late death, with all children clinically healthy and thriving.

Vertical vein: to ligate or not?

In two of our patients, the vertical vein was initially left open, thinking that it would be a helpful pop-off in either direction; decompressing the right heart during pulmonary hypertensive crisis and decompressing the left atrium in case of non-compliance of the neo-left atrium and a relatively small left ventricle. However, we had to close them on postoperative days 3 and 11 due to low cardiac output, leading to gradual stabilization of haemodynamics. The literature is amass with arguments for [5] or against [18] closure of the vertical vein. Though we have no hard evidence, we favour ligation or restriction of the vertical vein, especially if other causes of destabilization are ruled out.

Technical aspects of sutureless repair

Yanagawa et al. [6], while reporting their experience with primary sutureless repair, have alluded to the risks of sutureless repair in the form of thrombogenicity of the exposed pericardial surface, air embolism from high-pressure ventilation, soft-tissue rupture during dissection leading to bleeding into the pleura and the hazards of closed chest compression potentially disrupting the fragile neoatrium. Although we have not encountered them in our limited experience, these are important pitfalls that need to be kept in mind.

Primary sutureless repair has technical advantages in the sense that since it is performed on a virgin field, the landmarks are easy to identify and surgical planes easy to dissect. A bleeding in the pleural spaces can be, due diligence, avoided or easily taken care of. Our technique involves small modifications in terms of avoidance of deep hypothermia, minimization of the duration of circulatory arrest and ensuring a relatively dry surgical field during most part of the operation, despite continuous moderately hypothermic CPB, thanks to the tactical manoeuvre of opening the collector when 80% of the atrio-pericardial connection is complete. Surgical dissection and performance of the anastomosis without the collector being opened simplifies the technique without ever needing to transect the IVC or the aorta to improve visualization. Laminar flow across the neo-left atrium without evidence of any post-capillary obstruction with growth raises hopes of harmonious growth of the left atrium at longer follow-up. These and the course of pulmonary artery pressures, however, would need to be followed over a longer term, to draw any definite conclusions.

CONCLUSION

Primary sutureless repair of TAPVR can be safely performed in newborns, with low mortality. Early results in terms of creation of an unobtrusive neo-left atrium and consequent resolution of PHT in most of the patients are gratifying and portend well for the long-term. Our technical modification in creating the sutureless anastomosis helps avoid deep hypothermia and minimizes the duration of circulatory arrest. Mid- and long-term results in a large cohort of patients will show how far sutureless repair goes in mitigating the risks of post-repair PVO.

Conflict of interest: none declared.

REFERENCES


