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ORIGINAL ARTICLE

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

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Abstract

OBJECTIVES: The objective was to evaluate primary sutureless repair of total anomalous pulmonary venous return (TAPVR) in neonates using a modified technique that minimizes hypothermia and circulatory arrest times.

METHODS: From 2009 to 2011, seven consecutive patients underwent primary sutureless repair for the treatment of TAPVR, by which the prepared posterior pericardium was sutured to an opening in the left atrium. Three patients had the obstructed infracardiac type, and four patients had the unobstructed supracardiac type of TAPVR. Moderate hypothermia was used in all patients with a median temperature of 28°C (26–32). Circulatory arrest was not used except for the opening of the collector, which lasted between 3 and 5 min. The connecting vein was ligated in all seven patients (five during repair and two early postoperatively). The follow-up was 100% complete, with a median duration of 652 (range 370–1023) days.

RESULTS: There was no operative mortality and no late death. No patient required reoperation. Postoperative echocardiography showed unobstructed pulmonary venous flow in all patients. Recurrent pulmonary venous stenosis was not seen during the follow-up in any patient.

CONCLUSIONS: The sutureless technique is an effective technique with potential advantages even for the primary correction of TAPVR. With the described technique, the need for circulatory arrest is substantially reduced. Not handling the pulmonary venous collector by avoiding a direct anastomosis may contribute to better compliance, better growth and the absence of subsequent stenosis.

Keywords: Congenital cyanotic heart disease • Pulmonary venous obstruction • Total anomalous pulmonary venous return • Total anomalous pulmonary venous connection • Sutureless repair • Pulmonary hypertension

SURGICAL TECHNIQUE

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 flow 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

Table 1: Patient demographics

N	7
Age[a] (days)	6.4 (1–14)
Weight[a] (kg)	2.9 (2.3–3.2)
Male/female	2/5
Supracardiac (unobstructed) TAPVR	4
Infracardiac (obstructive) TAPVR	3
Univentricular/biventricular hearts	1/6

[a]Values are in median and range.

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 atriopericardial 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

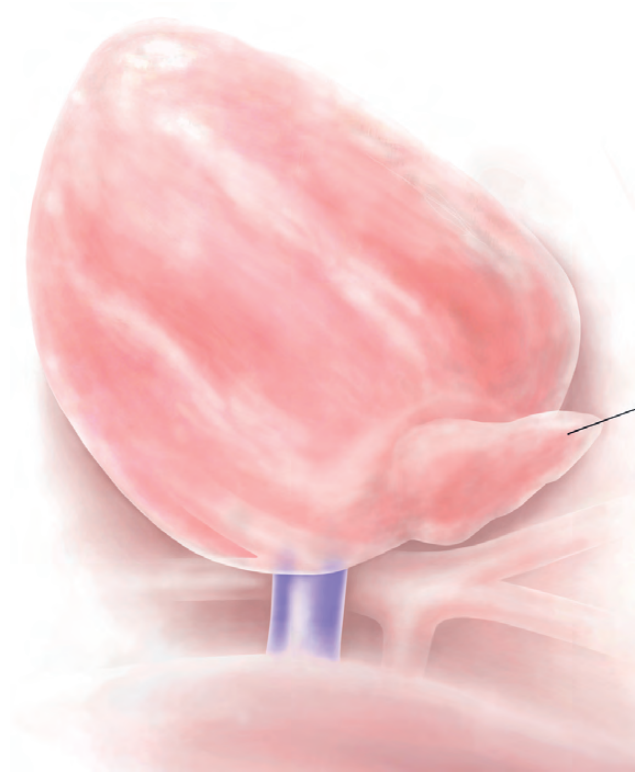

Figure 1: After instituting cardiopulmonary bypass and achieving cardiac arrest, the apex of the heart is luxated towards the right shoulder.

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).

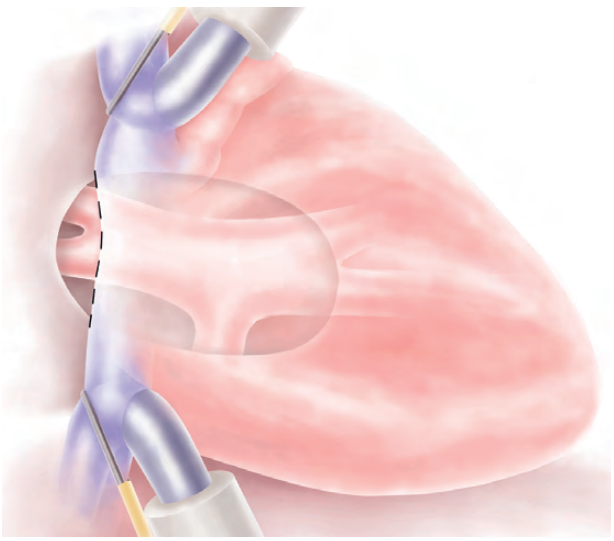

Figure 2: The right lateral aspect of the pericardial well is dissected along the interatrial groove and the course of the right upper and lower pulmonary veins can be followed.

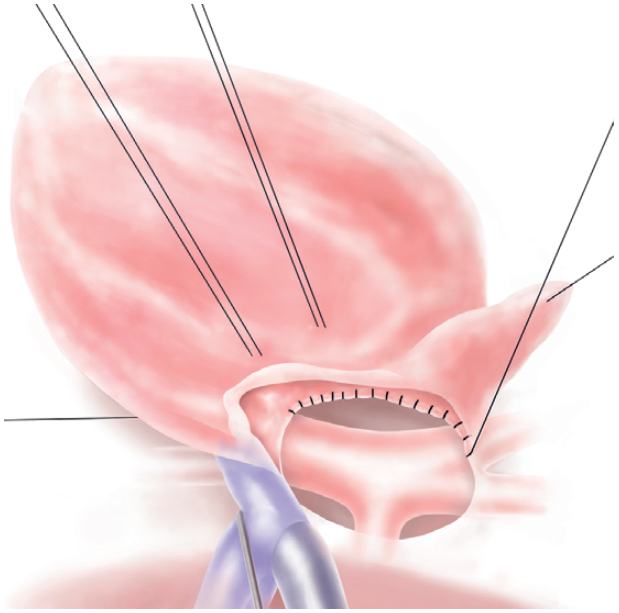

Figure 3: A left atriotomy parallel to the atrioventricular groove and extending from well into the left atrial appendage running up to the IVC is performed. A strip of left atrial wall is excised to widen the opening. The superior margin of the anastomosis is performed first starting from the far left end to the right under the SVC (suture marked black).

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 ostea, particularly if it appears that a discrete stenosis could exist. The vertical vein is interrupted using a ligaclip or a ligature (Fig. 5).

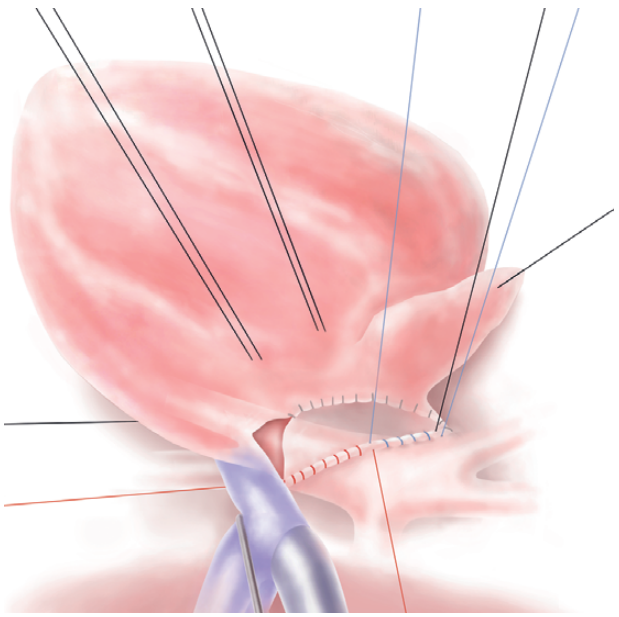

Figure 4: Breaking the inferior suture line (blue and red marked suture) helps in evenly correcting the discrepancy in circumference of the left atrium as well as the pericardial well.

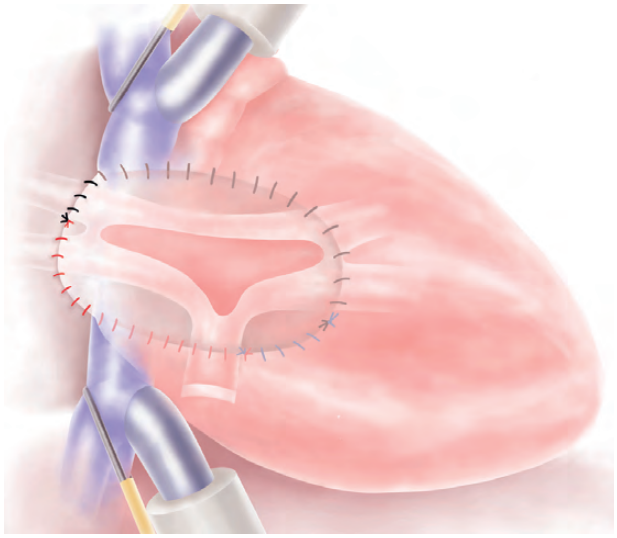

Figure 5: The descending vein, draining into the systemic vein under the diaphragm is clipped. During a short phase of circulatory arrest, the collector is opened with the incision extending into the descending vein. A small strip of the anterior wall of the collector is excised to prevent recoil. The right lateral part of the anastomosis between the parietal pericardium and the left atriotomy is completed.

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.

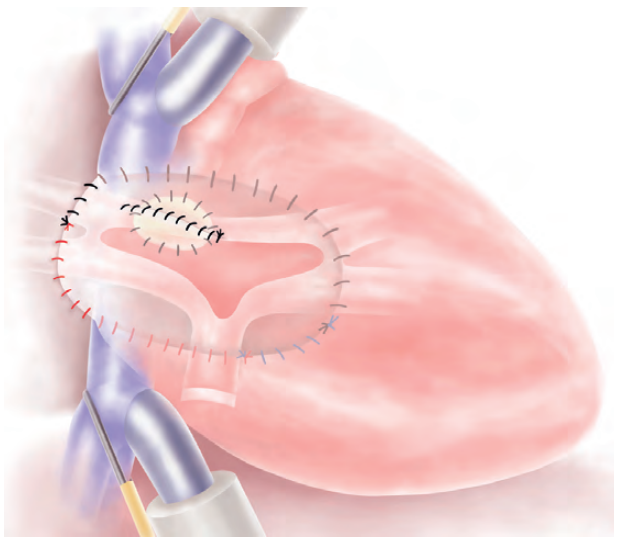

Figure 6: The atrial septal defect is closed, often using a piece of untreated autologous pericardium. A 2-mm fenestration is created and the right atriotomy closed.

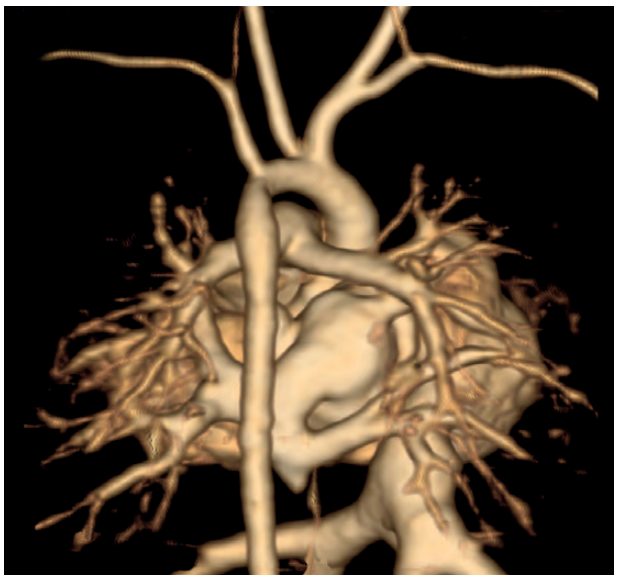

Figure 7: MR angiocardiography showing a wide neo-left atrium without pulmonary vein stenosis 4 months after primary sutureless repair for an obstructive infracardiac TAPVR.

The follow-up was 100% complete, with a median duration of 652 (range 370–1023) days.

RESULTS

There was no operative mortality and no late death. No patient required reoperation. Two patients with an obstructed TAPVR and preoperative hazy lungs were supported with an elective postoperative ECMO support. ECMO could be weaned off after 48 and 72 h, respectively. Two patients (one supracardiac TAPVR and one infracardiac obstructive TAPVR) had their vertical veins closed on postoperative days 3 and 11 because of a progressive significant left-to-right shunting. Postoperative echocardiography showed unobstructed pulmonary venous flow in all patients. Recurrent pulmonary venous stenosis was not seen during the follow-up in any patient. All sutureless anastomoses were echocardiographically confirmed to be wide and facilitating laminar pulmonary venous flow. One patient, following the repair of an obstructive infracardiac TAPVR with early postoperative severe PHT (two-third of systemic pressure) demonstrated no postcapillary obstruction (at the individual pulmonary veins or at the atrio-pericardial connection) on magnetic resonance angiography at 4 months (Fig. 7). PHT fluctuated between one-third and two-third of systemic pressure during the first 6 months after repair. The latest follow-up at 14 months showed a continuous decrease of pulmonary pressure to around one-third of systemic pressure. All patients were clinically healthy and thriving at the follow-up.

DISCUSSION

While conventional repair of TAPVR has served well with improving results, the late morbidity of PVO and PHT (5–18%) has been the focus of efforts aimed at improvement. Sutureless repair for the treatment of post-repair discrete PVO is an elegant operation described by Najm [10] and Lacour-Gayet [11]. However, the mortality in this difficult cohort of patients is high (40–66%). This has led to efforts to pre-empt and prevent the development of PVO right at the outset. To this end, various groups [4, 6] have pursued the use of sutureless repair for the primary correction of TAPVR. In view of the early positive results and intuitive advantages in terms of avoidance of handling of pulmonary veins (avoidance of any traction/distortion and avoidance of a full thickness fibrous scar), we have been pursuing this technique for primary correction of all supra- and infracardiac TAPVR. We have introduced modifications that allow us to perform a wide anastomosis, while avoiding deep hypothermia and minimizing the duration of circulatory arrest to a few minutes. In spite of early good results, conclusive mid- and long-term results are eagerly awaited, to see if the theoretical advantages prove to be true.

Pulmonary venous obstruction

Jenkins *et al.* 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 and 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, with 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.

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