Intra-atrial repair for total anomalous pulmonary venous connection

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Abstract

The natural history of total pulmonary venous connection (TAPVC) is unfavorable per se. We describe a modified technique of intra-atrial repair in infants with supra- and infracardiac TAPVC. Twenty patients were treated. The median age at repair was 16 (range 3–62) days. Echocardiography and multi-detector row computed tomography were used to confirm the diagnosis. Our policy was to perform surgery on an urgent basis whenever there was a sign of severe pulmonary congestion or hypoxia. The procedures performed in cardiopulmonary bypass (CPB) were established in a standard fashion using bi-caval cannulation and moderate hypothermia. A novel modification of our surgical technique is the H-shaped instead of simple straight incision of pulmonary venous confluence and intra-atrial sewing. H-shaped incision of pulmonary venous confluence can increase the anastomotic area than simply straight-line incision and create a large anastomosis with maximal use of the venous confluence and atrial tissue. Intra-atrial repair can avoid torsion and rotation of the pulmonary veins. There were no operative deaths and no recurrent pulmonary venous obstruction was noted after a mean period of 2 ± 0.8 years (range: 12–20 months). Intra-atrial repair provides excellent results for primary repair of supra- and infracardiac TAPVC in infants.

Key words: Total anomalous pulmonary venous connection; Infant; Operation

1. Introduction

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital anomaly. The natural history of supra- and infracardiac TAPVC is unfavorable per se. Obstruction of the pulmonary venous pathway is a powerful predictor of adverse natural outcome and the tendency for pulmonary venous obstruction [1–3]. Although surgical outcome of TAPVC has improved, postoperative pulmonary venous obstruction is still associated with reoperation or death [4–7]. We describe our surgical experience with supra- and infracardiac TAPVC in infants by using a novel modification of intra-atrial repair.

2. Materials and methods

Twenty consecutive cases of TAPVC undergoing total repair by a novel modification intra-atrial technique were reviewed. The median age at repair was 16 (range 3–62) days and the mean body weight was 3.2 ± 1.0 (range 2.3–6.5) kg. Eight patients were newborns. Diagnosis was made by echocardiography. Multi-detector row computed tomography was used to confirm the diagnosis if necessary (Fig. 1). Drainage of pulmonary vein was infracardiac in six (descending vein to portal vein in four, to the ductus venosus in two) and supracardiac in 14 (Darling type IA: 10, IB: 4). We favor performing surgery on an emergent basis whenever there was a sign of severe pulmonary congestion or hypoxia.

Cardiopulmonary bypass (CPB) was established in a standard fashion using bi-caval cannulation and moderate hypothermia. Adequate exposure of the vertical pulmonary venous confluence located in the posterior mediastinum was provided. The posterior mediastinal pleura was incised over the pulmonary venous confluence. The posterior pericardium was opened, the pulmonary venous confluence and the descending vein were identified, and a vertical incision is made in the anomalous pulmonary venous confluence to decompress the pulmonary veins. The vertical vein was tied off and the previous incision on the common pulmonary vein was extended longitudinally from the cephalad aspect down to the diaphragmatic end. At the each end of the incision, a vertical incision stretched slightly along the edge of common pulmonary vein. The whole incision is H-shaped. A mirror incision was made on the left atrium. A right atriotomy was made and across the atrial septum to allow visualization of the pulmonary vein ostia and clear definition of the location. Intra-atrial repair was performed without heart vaigus (Fig. 2). The divided edge of the left atrial wall was...
subsequently connected to the pulmonary venous confluence by suturing the edges of the atrium to the vertical vein, staying away from the actual edge of the pulmonary veins. The anastomosis was constructed by using a 7/0 polypropylene suture. An autologous pericardial patch was used to close the enlarged atrial septal defect.

3. Results

This technique was successfully performed in 20 infants. All patients were weaned from CPB without difficulty. The median intubation period was 3 (range 0—14) days. The mean CPB time and aortic cross clamp time were 78.5 ± 18.2 and 39.0 ± 10.2 min, respectively. Postoperative course of the patients was satisfactory. The surgical results in the follow-up period (2 ± 0.8 years, range: 12–20 months) were evaluated in 15 patients. Improved cardiovascular function was confirmed by echocardiogram. There were no echocardiographic signs of pulmonary venous obstruction. Blood flow velocity in pulmonary venous anastomosis was between 0.5 and 1.1 m s⁻¹.

4. Discussion

We use a novel modified intra-atrial repair technique in primary repair of supra- and infracardiac TAPVC in infants. Based on our limited experience, we speculate that the main principles to reduce the risk of subsequent anastomotic stenosis of repair TAPVC are an adequate large and tension-free anastomosis and avoiding trauma to the pulmonary venous wall and endothelium. In our report, the H-shaped incision of pulmonary venous confluence like a clamshell, which can increase the anastomotic area than simply straight line incision and create a large anastomosis with maximal use of the venous confluence and atrial tissue.

There were no echocardiographic signs of pulmonary venous obstruction such as flow acceleration (>1.5 m s⁻¹), elevated right ventricular pressure, narrowing of the confluence or tributary veins, etc. Furthermore, patients with long-term follow-up are needed to study and draw definitive conclusions on its efficacy.

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References


