

Morphological dilemma: Anomalous pulmonary venous confluence or cor triatriatum—does it matter?



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Cardiac variant of total anomalous pulmonary venous return is a rare entity, whereby all the pulmonary veins drain directly to the right atrium or coronary sinus. The effective left heart blood flow channels through a small stretched patent foramen ovale and can often be confused with a variant of cor triatriatum. Cor triatriatum is a rare congenital cardiac anomaly where pulmonary veins drain to a persistent chamber above the left atrium with a membrane separating these two. There persists either a small aperture directly from the true to the accessory left atrium or none at all. Where there is no such aperture, it is often physiologically akin to the cardiac variant of total anomalous pulmonary venous return described above. Such morphological differentiation is often challenging in a clinical situation, but the effective treatment remains the same. It involves removal of the common wall between the two chambers and baffling the pulmonary veins effectively to the left atrium. We describe such a case where the pulmonary venous return is to the right atrium, managed recently in our centre, and discuss the morphological differences between these two.

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Keywords: Cardiac morphology, Cor triatriatum, Total anomalous pulmonary venous drainage

Introduction

Total anomalous pulmonary venous return and cor triatriatum are two of the rare congenital heart conditions, often resembling each other by morphological appearances. Diagnostic difficulties remain, and management is surgical though

at times difficult. This case report highlights one such report whereby urgent repair of total anomalous venous return highlights its close resemblance to cor triatriatum morphologically.

Case report

A 3-month-old child was evaluated for persistent desaturations and cyanosis. Echocardiogra-

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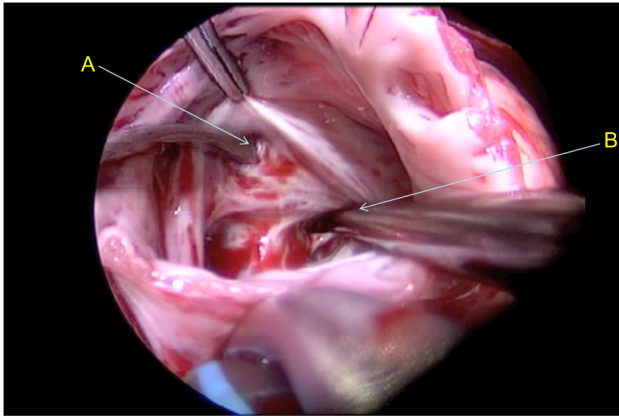


Figure 1. Intraoperative photograph showing atrial septum, fenestration draining pulmonary venous confluence (A), and coronary sinus (B).

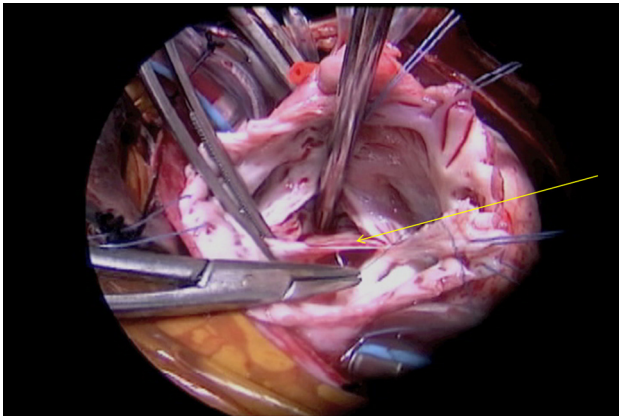


Figure 2. Intraoperative photograph after the intervening common wall was removed and the posterior wall repaired (arrow).

phy confirmed the presence of cardiac type of total anomalous pulmonary venous return (TAPVR) with all four pulmonary veins draining directly into the right atrium. The coronary sinus was normal and not unroofed. There was a small patent foramen ovale, with a restrictive flow across to the left atrium (LA). There was also evidence of severe pulmonary hypertension. The child was accepted for an urgent repair of TAPVR. At surgery, under standard cardiopulmonary bypass and bicaval cannulations, right atriotomy was performed to expose the intracardiac anatomy. All four pulmonary veins were directly draining into the posterior part of the right atrium, with no intervening septum. There was an oblique lie of atrial septum with a small superiorly placed defect providing an interatrial shunt (Fig. 1). This was incised to reveal the true LA and mitral valve. The primum septum was then completely taken

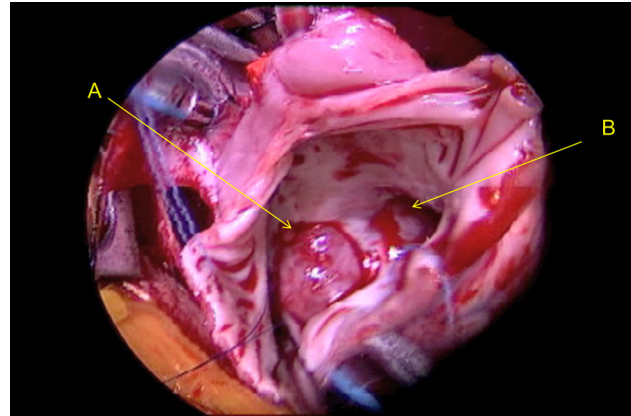


Figure 3. Intraoperative photograph showing completed repair with atrial septal patch (A) and tricuspid valve (B).

down, and an autologous pericardium was used to baffle the pulmonary veins to the true LA (Figs. 2 and 3). Postoperatively, there was an episode of pulmonary hypertensive crisis, necessitating commencement of inhaled nitric oxide and a delayed chest closure, but the child recovered well subsequently and discharged home in 12 days after surgery. Follow-up at 8 months revealed an asymptomatic child with normal cardiac function, with no evidence of any pulmonary venous obstruction.

Discussion

The cardiac variant of TAPVR is a rare type of congenital heart disease and constitutes 25% of TAPVRs [1]. The common pulmonary venous chamber can drain either into the coronary sinus, thereby channelling pulmonary venous outflow into the right atrium, or directly into the right atrium. When draining into the right atrium, blood flow into the left heart chamber depends on the fenestration at the interatrial septum or patency of the foramen ovale, in order to maintain the cardiac output. Similarly, in a variant of cor triatriatum, where a membrane separates the false and true LA with a large atrial defect above the level of membrane (Type IIa—Lucas–Schmidt classification), the flow into the LA will depend on the fenestration in the atrial septum towards the true LA in order to maintain the left heart output [2].

Diagnosis depends on clinical presentation, and quite often than not, a Type IIa cor triatriatum with a possible restrictive left heart flow will present early with cardiomegaly and pulmonary hypertension [3,4]. An unobstructed cardiac vari-

ant of TAPVR may present late clinically, on a comparative basis, though the final restriction depends on the true nature of atrial septal fenestrations allowing the shunt across to the left heart.

Though morphologically different, these two variants will look similar and involve a similar strategy to repair at surgery. Surgery will involve removal of the intervening membrane and providing an unrestrictive inflow from pulmonary chamber into the true LA, while the atrial septum is reconstructed with a patch. The surgical results are very satisfactory in both the groups [3,4].

The ongoing morphological controversy highlights that pathological and/or embryological mechanism may actually be identical between the cardiac variant of TAPVR and some of the types of cor triatriatum [2,5].

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