

Complex Supracardiac Total Anomalous Pulmonary Venous Connection — A Case Report

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We present the case of a 4-month-old female who had complex supracardiac total anomalous pulmonary venous connection, diagnosed by catheterization. Her right pulmonary veins drained into the left brachiocephalic vein and then into the superior vena cava. The left pulmonary veins drained into the hemiazygous and azygous veins and eventually into the superior vena cava. She had no other major structural abnormalities except for a secundum-type atrial septal defect and patent ductus arteriosus. Although several rare mixed types of total pulmonary venous connection have been reported in the literature, to our knowledge, the anomaly our patient had has not been described previously.

Key Words: Total anomalous pulmonary venous connection • Supracardiac • Mixed type

INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital cardiac malformation in which none of the pulmonary veins has a connection with the left atrium. More than one third of cases have an anomalous connection to the left brachiocephalic vein, while most of the rest have a common pulmonary venous confluence that drains into other sites such as the coronary sinus or right atrium.^{1,2} This report describes our experience of a rare variety of complex supracardiac TAPVC.

CASE REPORT

A 4-month-old female patient was referred to our clinic because she had peripheral cyanosis during crying

spells and was found to be polycythemic. On physical examination, she had mildly cyanotic lips and fingernails. A faint systolic murmur was audible over the left sternal border with a loud P2. She had no hepatomegaly, and her peripheral pulses were normal. A chest x-ray showed mild cardiomegaly with mild pulmonary venous congestion. Electrocardiography revealed right axis deviation, right atrial enlargement, and right ventricular hypertrophy. Echocardiography showed a dilated right atrium and ventricle, a small left atrium, patent ductus arteriosus, and mild tricuspid regurgitation. There was a secundum-type atrial septal defect with a pure right-to-left shunt. No pulmonary venous connection to the left atrium could be identified, and there was no abnormal pulmonary venous confluence. TAPVC was suspected and cardiac catheterization and angiography were performed. The respective O₂ saturations of the right brachiocephalic vein and IVC were 61% and 56%, respectively. There was a marked oxygen step-up in the right atrium (80%), as well as in the left brachiocephalic vein (84%) and the junction of the superior vena cava and azygous vein (95%). The pressure was 61/40 mmHg in the main pulmonary artery, 60/39 mmHg in the left pulmonary artery, and 70/44 mmHg in the right pulmonary artery. A selective right pulmonary arteriogram

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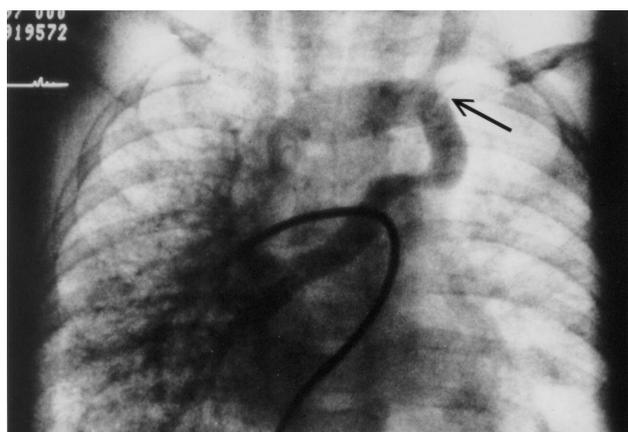


Figure 1. Right pulmonary veins draining into the left vertical vein obliquely and then into the innominate vein (black arrow) in the levophase of the selective right pulmonary arteriogram.

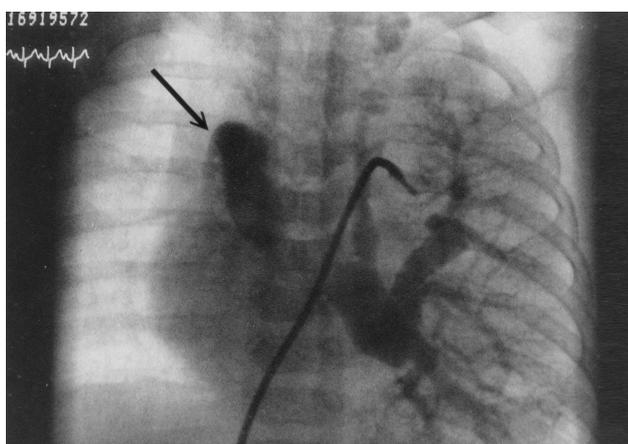


Figure 2. Left pulmonary veins draining into the hemiazygous and azygous veins and then into the superior vena cava (black arrow) in the levophase of the selective left pulmonary arteriogram.

showed right pulmonary veins draining into obliquely the left vertical vein and then into the left brachiocephalic vein and the superior vena cava (Figure 1). The right pulmonary venous confluence was very stenotic at the junction with the vertical vein. A left pulmonary arteriogram showed the left pulmonary venous confluence draining into the hemiazygous and azygous veins and thence into the superior vena cava (Figure 2). The overall distribution of the pulmonary venous connection is illustrated in Figure 3. The associated cardiac anomalies were secundum-type atrial septal defect and patent ductus arteriosus. Unfortunately, the patient died one day after operation.

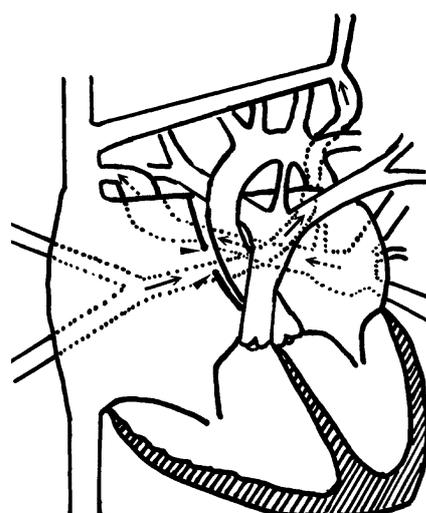


Figure 3. Illustration of the anomalous distribution of the pulmonary venous connections. The solid line with an arrow indicates the route of the right pulmonary venous return, and the dotted line with an arrow indicates the route of the left pulmonary venous return. The ostium of secundum defect is marked by two black arrowheads.

DISCUSSION

The incidence of TAPVC is 0.008% of live births, but it occurs in 2% to 3% of cases of congenital heart disease.^{3,4} Darling and associates divided these anomalies into four subtypes based on the site of drainage of the pulmonary venous flow: Type I, anomalous connection at the supracardiac level; (45% of cases) Type II, anomalous connection at the cardiac level (26%); Type III, anomalous connection at the infracardiac level (24%); and Type IV, mixed, with anomalous connections at two or more of the above levels (5%).^{2,5} Various mixed types have been reported.⁶⁻⁹ Grace et al. reported a patient with pulmonary drainage above, below, and into the heart.¹⁰ To our knowledge, the complex supracardiac drainage in our case has not previously been reported.

TAPVC is generally considered to result from early atresia of the common pulmonary vein while pulmonary to systemic venous connections are still present.^{1,11} In our case, during embryogenesis there may have been persistent communication between the primitive splanchnic plexus and the common cardinal veins bilaterally, followed by agenesis or atresia of the common pulmonary vein. We were unable to delineate clearly the entrance of the aberrant pulmonary venous connection by echocardiography. Catheterization was necessary to demonstrate the unique

pattern of TAPVC in this case. Imoto et al. reported that the diagnostic sensitivity of echocardiography alone for mixed TAPVC was 67%, while that of catheterization was 100%.⁷ This case is a reminder that, if the patient can tolerate it, cardiac catheterization should be performed when TAPVC is suspected but cannot be confirmed echocardiographically. This may be the only way to ascertain the exact route of the pulmonary venous drainage.

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複雜的心上型全肺靜脈回流異常 — 一個病例報告

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我們發表一位四個月大的小女孩，經由心導管檢查證實其患有複雜的心上型全肺靜脈回流異常。她的右肺靜脈注入左無名靜脈，然後回流至右上腔靜脈；而其左肺靜脈則注入奇靜脈系統最後回流至右上腔靜脈。除了心房中隔缺損及開放性動脈導管之外，小女孩並無其他重大心臟構造上的異常。文獻上雖有其他稀有的混合型全肺靜脈回流異常發表；但就我們所知，此位小女孩的患病型態應未有他人描述過相同的類型。

關鍵詞：全肺靜脈回流異常、心上型、混合型。