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Total anomalous pulmonary venous connection: preoperative anatomy, physiology, preoperative evaluation, surgical considerations and single centre clinical experiences

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Abstract

Objectives: The aim of the present study was to document the anatomical subtypes of total anomalous pulmonary venous connection (TAPVC) since it is critical in the surgical and medical management of this cardiac anomaly.

Methods: This retrospective study was conducted at a paediatric heart centre between February 2019 and December 2021. The study included 27 patients who underwent intracardiac repair of total anomalous pulmonary venous connection.

Results: The mean age of the patients were found 159.37±411.29 (range: 7.00–2160) days, the mean weight of patients were 4756.67±2988.78 grams. Mean oxygen saturation at the arrival of the hospital was 84.89±7.98 (range: 60–96) %. Mortality was seen in 7 (25.9%) patients. 2 of 7 patients (28.6%) had supracardiac type, 2 of 7 patients had (28.6%) cardiac type, 2 of 7 (28.6%) patients had infracardiac type and 1 of 7 patients (14.3%) had mixt type anomaly. Only 3 (11.1%) patients needed ECMO. One of these 3 patients (33.3%) had supracardiac type, and the remaining 2 of 3 patients (66.7%) had infracardiac type anomaly.

Conclusion: Total anomalous pulmonary venous connection is a rare cardiac anomaly seen in 7/100,000 live births. It constitutes approximately 1% of all congenital heart diseases. It can be found as an isolated cardiac defect or accompany other diseases; usually accompanying cardiac lesions are associated with the severity of the disease and poor prognosis. Understanding the anatomical subtypes of TAPVC is critical in the surgical and medical management of this disease.

Keywords: cardiac anomaly; pulmonary venous hypertension; total anomalous pulmonary venous connection

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Introduction

Total anomalous pulmonary venous connection (TAPVC) is a spectrum of cardiac anomalies that occurs when all of the pulmonary veins drain to a structure other than the left atrium.[1] The pulmonary veins return either to one of the systemic veins such as the brachiocephalic vein, ductus venosus, hepatic vein, the coronary sinus, or directly to the right atrium. In this pathology, there is a complete mixture of pulmonary and systemic venous return in the right atrium and an obligate right to left atrial shunting to sustain life.[2] There are multiple anatomical subtypes, and the presentation and outcomes are largely influenced by any point of obstruction to the pulmonary venous return to the heart. The aim of the present study was to document the anatomical subtypes of this pathology since it is critical in the surgical and medical management of this disease.

Materials and Methods

This retrospective study was conducted at a paediatric heart center, between February 2019 and December 2021. The study included twenty-seven patients who underwent intracardiac repair of total anomalous pulmonary venous connection.

Data including age, sex, type of TAPVC, surgical technique, preoperative right ventricle systolic pressure, oxygen saturation at the arrival of the hospital, duration of cardiopulmonary bypass (CPB) and cross-clamp (CC)

time, postoperative pulmonary artery pressure, duration of inotropic support, duration of ventilation, duration of stay in the intensive care unit (ICU), duration of stay in hospital as well as mortality rate, was all retrieved from the institutional databases and medical records of the patients.

Interquartile range (IQR) was used to express continuous data, whereas frequency and percentages were used to represent categorical variables. A p-value of less than 0.05 was considered statistically significant for all statistical analyses, which were carried out using the SPSS for Windows (Version 25.0, IBM Corp., Armonk, NY, USA).

Results

The mean age (day) of the patients were found to be 159.37±411.29 (range: 7.00–2160) days, the mean weight (g) of patients was 4756.67±2988.78 g, the mean of CC time (minutes) was 55.85±23.23 (range: 32–173) and the CPB time (minutes) mean was 96.74±37.12 (range: 41–226) minutes.

Mean oxygen saturation (%) of the patients at the arrival of the hospital was 84.89±7.98 (range: 60–96). The mean preoperative right ventricular systolic pressure (RVSP), mechanical ventilation time and inotropic support time was 63.59±27.37 (range: 25–150), 69.59±76.78 (range: 12–360), and 69.59±74.42 (range: 12–360) minutes respectively.

Finally, the mean postoperative oxygen saturation (%) of the patients was 93.07±.08 (range: 80–100), and the mean postoperative pulmonary artery pressure (PAB) was 27.59±12.94 (range: 17–85) mmHg.

Nine of the twenty-seven patients (33.3%) had supracardiac type, 11 of 27 patients (40.7%) had cardiac type, 6 of 27 patients (22.2%) had infracardiac type and 1 of 27 patients (3.7%) had mixed type anomaly.

Mortality was seen in seven patients (25.9%). Two of these patients (28.6%) had supracardiac type, 2 of 7 patients (28.6%) had cardiac type, 2 of 7 patients (28.6%) had infracardiac type and 1 of 7 patients (14.3%) had mixed type anomaly.

The distribution of the patients according to the number of inotropes that they received was as follows: seven patients (25.9%) received 2 types, 12 patients (44.4%) received 3 types, 8 patients (29.6%) received 4 types of inotropes.

Only three patients (11.1%) needed extracorporeal membrane oxygenator (ECMO). One of these 3 patients (33.3%) had supracardiac type, and the remaining two patients (66.7%) had infracardiac type anomaly.

Discussion

TAPVC is a rare cardiac anomaly seen in 7/100,000 live births.[3] It constitutes approximately 1% of all congenital heart diseases.[4] It can be found as an isolated cardiac defect or accompany other diseases. Accompanying cardiac lesions (heterotaxy syndrome, atrial isomerism, anatomical lesions with single ventricular physiology, etc.) are associated with the severity of the disease and poor prognosis.[5] Most patients do not have a family history of congenital heart disease, but case reports showing that it is seen in siblings and first-degree relatives have been described in the literature. In the case series published by Lucas et al.,[6] TAPVC opening into the portal vein was found to be more common in men (3.6:1). There is no known environmental fetal factor for TAPVC.

The failure of all pulmonary veins to separate from the splanchnic venous system and the concomitant leftward placement of the atrial septum, are thought to be the primary defect in the formation of TAPVC. Unilateral formation of the defect throughout development results in partial pulmonary venous return anomaly, and its occurrence in later stages results in common pulmonary vein atresia, cor triatriatum or abnormal union of the common vein with the left atrium, resulting in congenital pulmonary vein stenosis and an abnormal number of pulmonary veins.[7–10]

TAPVC consists of four anatomical subtypes: supracardiac, cardiac, infracardiac and mixed type, where the supracardiac type accounts for approximately 45% of all cases. All pulmonary veins form a sac and this common sac drains through the vertical vein into the brachycephalic vein, superior vena cava or azygos vein. This vertical vein typically courses anterior to the left pulmonary artery and enters the systemic venous circulation.[2] Although supracardiac TAPVCs are nonobstructive, obstruction can occur at two main points. In 40% of supracardiac TAPVCs, the vertical vein passes between the left pulmonary artery and the left pulmonary bronchus, and creates a stenosis. The second possible site of obstruction occurs where the vertical vein opens directly into the superior vena cava (SVC) or azygos vein, and may cause obstruction at different levels. The innominate vein and SVC are enlarged due to increased blood flow.[1,2] In our study 33.3% of the patients had supracardiac type anomaly.

Infracardiac TAPVC accounts for 25% of all cases and is the most common type of obstruction. After the pulmonary veins open into the common venous sac, they open into the ductus venosus, hepatic artery, inferior vena cava or portal vein at a level below the diaphragm via the vertical vein. Since the ductus venosus begins to regress

with birth, it causes obstruction that develops over time. In cases without stenosis, pulmonary venous return comes to the right side of the heart via the inferior vena cava (IVC) and passes through the patent foramen ovale (PFO) to reach the left side of the heart.[2] In our study, 22.2% of the patients had an infracardiac type anomaly.

Cardiac TAPVC occurs in approximately 20% of all cases. The common venous sac opens into the right atrium via the coronary sinus, which is dilated. In some cases, it can open directly into the right atrium. These cases are usually associated with heterotaxy syndrome and complex cardiac malformations. Oxygenated blood enters the left atrium via the PFO and this type of TAPVC is the least obstructed type. In our study, 40.7% of the patients had cardiac type anomaly.

The mixed type TAPVC is the rarest type of TAPVC, occurring in approximately 5% of patients. Such patients present in a severe and often severe form. Accurate preoperative diagnosis and clear anatomical definition are critical in planning the surgical strategy in these patients. The confusion in the diagnosis is usually due to the wide anatomical variations in mixed type TAPVC.[11] In our study 3.7% of the patients had mixt type anomaly.

At least fifteen different anatomical variations have been described. The most common form is the type in which the left upper lobe lung drains into the left vertical vein and all the remaining venous drainage of the lung into the coronary sinus. In the other most common form, the entire right lung drains into the coronary sinus, while the left lung drains into the vertical vein.[12] Chowdhury et al.[12] basically classified mix type TAPVCs into three general groups. Two by two pattern, three by one pattern and the bizarre pattern. The most common anatomical configuration is the latter, followed by 2 by 2, and the least common is the bizarre pattern. Obstruction occurs in approximately 40% of mixed-type TAPVCs.

In patients with TAPVC, obstructed or non-obstructed pulmonary venous drainage or the degree of obstruction dramatically changes the neonatal presentation of the disease. In patients with junctional obstruction, the main presentation is profound hypoxia and pulmonary hypertension. This pulmonary hypertension can sometimes be thought of as a persistence of the fetal circulation. X-ray radiograph shows pulmonary edema without cardiomegaly. The risk of developing circulatory collapse is high in patients with insufficient mixing at the atrium level. These patients may need ECMO until the correct diagnosis is made and surgical correction is made. Obstructed TAPVC is a surgical emergency. In non-obstructive types, varying degrees of desaturation are seen in the presence of heart failure and cardiomegaly; this is due to the wide left-right shunt. In some cases, the diagnosis is missed in the neonatal period.[11,12]

Two-dimensional echocardiography (ECHO) is sufficient to demonstrate anatomy in most TAPVC patients. Cardiac catheterization is rarely needed: in fact, it is often avoided as it may delay corrective cardiac surgery. Magnetic resonance and tomographic angiography may be useful for understanding the anatomy of the pulmonary connection. After diagnosis, surgical correction should be performed without delay for obstructed and non-obstructed TAPVCs. Because the disease does not have the potential to regress and palliative medical strategies do not provide an effective solution. Prostaglandins can be used to maintain ductal patency and provide systemic circulation, but they have no place in the correction of hypoxia or in the treatment of resistant acidosis. Similarly, nitric oxide is not effective in increasing oxygenation. Mueller performed the first surgical correction of TAPVC with pulmonary venous sac and left atrial appendage anastomosis in 1951, without using a cardiopulmonary bypass. In 1956, Lewis and Varco made the first complete correction of TAPVC, using the inflow occlusion technique. In the same year, Kirklin performed a TAPVC repair using cardiopulmonary bypass. Although all correction surgeries are performed using cardiopulmonary bypass today, there are surgical and central differences between the techniques.[13,14]

Patients with pulmonary venous obstruction should be operated immediately as soon as the diagnosis is made. Non-obstructive cases should be operated within the first two months. In patients with non-obstructive pulmonary venous connections, medical treatment is given for right ventricular failure, hypoxia and congestive heart failure. These patients are provided with inotropic support, effective diuresis and avoidance of high oxygen, as it reduces pulmonary vascular resistance and increases pulmonary congestion. Medical treatments are limited in obstructive type TAPVC. These patients are frequently intubated, the goal being to create respiratory alkalosis by keeping partial CO2 levels below 30 mmHg. These reasons generally require hyperventilation with 100% oxygen. To prevent metabolic acidosis, together with sodium bicarbonate, an inotrope is used to support ventricular functions and effective diuresis is provided for the treatment of pulmonary edema. In patients with insufficient right-left shunt, cardiac output is increased by increasing duct patency, by starting prostaglandin-E1. In some centers, cardiac catheterization is performed in patients with obstructive type TAPVC, both for diagnostic procedures and for palliation. These interventions are dilation of the restricted

atrial septum with balloon atrial septostomy and stenting of the obstructed vertical vein.[9,15,16]

In surgical repair of supracardiac TAPVC, the vertical vein is ligated through the innominate vein or SVC junction, freeing it from the surrounding tissues. In the presence of severe preoperative obstruction, sometimes leaving the vertical vein open may be beneficial for the prevention of postoperative pulmonary hypertension, but re-intervention may be required after significant residual left-right shunt. A horizontal incision is made in the posterior wall of the left atrium and the pulmonary venous sac, and these two structures are anastomosed to each other as wide as possible. The most critical factor here is proper anastomosis of the pulmonary veins without distortion to ensure long-term patency.[9]

In surgical repair of cardiac TAPVC, there is a vertical vein opening into the coronary sinus. The atrial septal defect (ASD) is first expanded towards the coronary sinus. An unroofed coronary sinus is created. If there is stenosis in the opening of the coronary sinus to the pulmonary venous sac, this area is widened and the stenosis is relieved. The large interatrial defect formed is closed with a patch, leaving the coronary sinus and pulmonary venous drainage to the left of the septum. In cases where the TAPVC opens into the right atrium, pulmonary venous drainage is diverted to the left atrium via the PFO by baffle.[9]

In infracardiac TAPVC, the vertical vein is typically extrapericardial and passes through the esophageal hiatus into the abdomen. The pulmonary venous sac extends vertically in a Y shape. When the pulmonary venous sac is reached, the vertical vein is ligated. After the pouch is opened with a vertical and Y-shaped incision, the supradiaphragmatic portion of the vertical vein is segmented and used to widen the left atrial anastomosis.[9,17]

The most common type of mixed type TAPVC is the type in which three veins drain posterior to the junction of the pulmonary veins and one drains separately into the systemic venous system. The repair is often in the form of establishing the connection of the left atrium to the pulmonary venous sac. Sometimes a single pulmonary vein that opens into the systemic veins can be left in place to prevent postoperative stenosis.[17]

Pulmonary hypertension may complicate the postoperative period in newborns with TAPVC, especially in patients with pulmonary venous obstruction in the preoperative period. There may be a need for nitric oxide during separation from CPB. Even in advanced cases, ECMO support may be needed due to failure to exit the CPB. In our study only 3 (11.1%) patients needed ECMO.

It is very important that there is no residual stenosis in the pulmonary venous anastomosis in these patients. In some cases, pulmonary hypertension may persist despite a non-obstructed anastomosis. In these patients, lymphangiectasias can be seen in lung biopsies. It is thought that the most important factor causing this situation is exposure to long-term pulmonary venous hypertension in the intrauterine period. These patients usually have diffused small pulmonary veins that are detected intraoperatively.[13,18]

Conclusion

TAPVC is a set of congenital cardiac defects with a wide range of anatomical characteristics that are linked by a shared pathophysiology, mostly determined by the presence or absence of pulmonary venous obstruction. An early diagnosis makes proper preoperative treatment and surgical correction easier.

Conflict of Interest

The authors declare no conflict of interest.

Author Contributions

All authors contributed equally to protocol/project development, data collection, data analysis, manuscript writing/editing.

Ethics Approval

The study was carried out in compliance with the Declaration of Helsinki's guiding principles and the protocol was accepted by the ethics committee of the Institution (No: E2-22-3047).

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