Case Report

Supracardiac Total Anomalous Pulmonary Venous Connection

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Abstract

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart disease in which there is developmental absence of connection of all four pulmonary veins with the left atrium. To report a rare case and share our experience in surgery and post-operative management for supracardiac TAPVC. Patient with supracardiac TAPVC with atrial septal defect (ASD) secundum variety with rudimentary patent ductus arteriosus (PDA) underwent rechanneling of pulmonary veins to left atrium (LA) with gluteryldehye treated autologous pericardial patch closure of ASD with ligation of ascending vertical vein and ligation of rudimentary PDA.Post operatively there were no events of pulmonary hypertensive crisis, low cardiac output syndrome, right heart failure or conduction defect were observed and echocardiogram showed adequate pulmonary venous drainage with no residual shunt across the interatrial septum. Marked development in surgical results of TAPVC has been observed in recent years with declining mortality rate from 65% in early sixties to 5% in current surgical scenerio.

Key words: *Vertical vein; Total anomalous pulmonary venous connection; Pulmonary hypertensive crisis.*

Date of received: *10.10.2018.*

Date of acceptance: *25.02.2019.*

DOI: https://doi.org/10.3329/kyamcj.v10i2.42792

KYAMC Journal.2019;10(2): 118-121.

Introduction

TAPVC is a rare congenital heart disease accounting for 1% to 3% of all congenital heart malformation[1] in which all the pulmonary venous effluent from the lungs drains to the systemic venous system creating a large left to right shunt.[2] More than one third of cases have an anomolous connection to left brachiocephalic vein while most of them have a common venous confluence that drains into other sites such as coronary sinus, right atrium or superior vena cava.[3,4] Depending on the drainage site of the pulmonary veins, the defects may be divided into (i) Supracardiac (50%)-when pulmonary veins open into left innominate vein and superior vena cava (SVC) (ii) Cardiac (28%)-when they open into coronary sinus (iii) Infracardiac (14%)-when they open into portal vein and (iv) Mixed (7%) variety.[5,6] Pathophysiologically, these four types are subclassified according to whether the pulmonary venous return is obstructed or nonobstructed.[7] In this case report we describe and share our experience regarding surgical technique and post-operative outcome of supracardiac variety of unobstructed TAPVC.

Case Report

This ten years old boy presented with exertional dyspnoea and occasional exertioal chest pain for last three years duration. He had history of recurrent respiratory tract infection with poor feeding and growth since birth. On general examination, no abnormality was detected except poor nutritional status. There is a soft systolic murmur in the pulmonary area and a fixed wide splitting of the second heart sound. The chest radiograph revealed mild cardiomegaly and plethoric lung field. The electrocardiogram showed incomplete right bundle branch

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block. The echocardiography revealed atrial septal defect (secundum), anomalous pulmonary venous connection into third chamber, tricuspid regurgitation gradeII, pulmonary hypertension (PASP=44) with intact interventricular septum. There was no abnormality at atrioventricular valves and no PDA flow seen (Figure1). CT angiogram revealed large interatrial septal deffect (about 23.3 mm) with right to left intra cardiac shunt. Pulmonary artery (PA) developed from right ventricle (RV) at infundibular level measuring about 27.4 mm. Valve area was about 32.5 mm, right pulmonary artery (RPA) was about 20.5 mm and left pulmonary artery (LPA) was about 19.7 mm. Pulmonary veins united together to form single venous confluence on left side and ascending vertical vein developed from venous confluence arches over aorta crossing left to right side over aorta and opened into superior vena cava (SVC). Right subclavian vein, right internal jugular vein (IJV) open into SVC. Left subclavian vein, left IJV opened in the left side of pulmonary vein. No PDA, no MAPCA were seen. Coronary arteries were normal. No pleuro-pulmonary lesion was see and bony thorax and overlying soft tissue were normal. The impression of CT angoigram was supracardiac TAPVC with ASD with dilated RA, RV. (Figure 2)

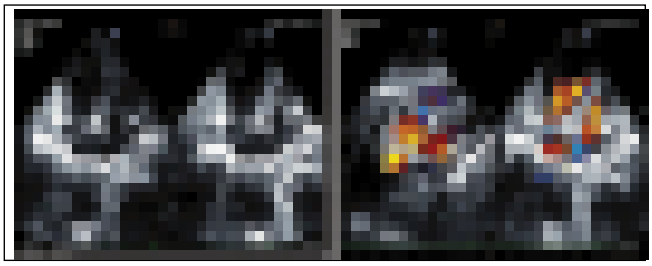

Fig 1: Echocardiography demonstrates supra cardiac TAPVC

Patient was planned for reconstructive surgery for rechanneling of four pulmonary veins into LA and closure of the atrial septal defect. CPB was established with aortic and venous cannulation (selective SVC and IVC). Vent was placed at right superior pulmonary vein. Control of ascending vertical vein was taken. Heart was arrested with intermittent antegrade cold bood cardioplezia.

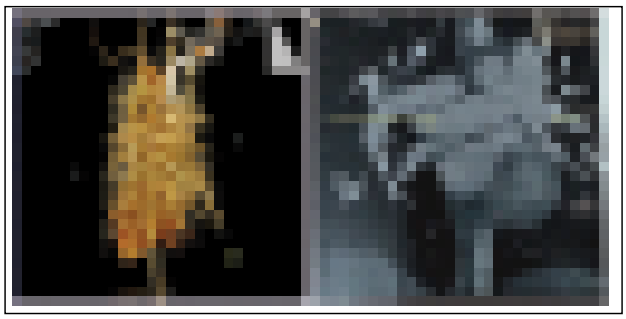

Figure 2: CT angiogram showing supra cardiac TAPVC.

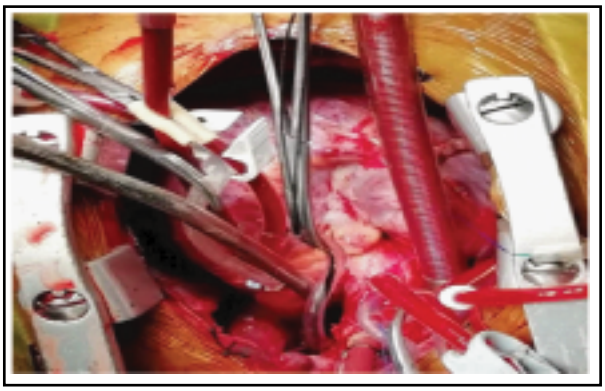

Figure 3: Per-operative view after rechanneling of four pulmonary veins to left atrium.

Intraoperatively there was a pulmonary confluent where all the four pulmonary veins drained and through ascending vertical vein pulmonary venous blood drained into SVC. SVC and ascending vertical vein were dilated. Aorta was small in size, main pulmonary artery (MPA) was distended and mildy tense but LPA and RPA were of adequate size. A large ASD measuring about 20 mm × 25 mm was found at fossa ovalis. A rudimentary PDA was found.

Rechanneling of TAPVC was done by making an incision over pulmonary confluent and adjacent LA auricle. Then anastomosis between both the chambers was done by continuous prolene 6(0) stitches. (Figure 3) Then right atriotomy was done and ASD defect was closed with gluteryldehye treatedautologous pericardial patch. After weaning from CPB, LA pressure, pulmonary pressure and systemic pressure were monitored with temporary occlusion of ascending vertical vein. The pressure readings were found acceptable and the ascending vertical vein was ligated. The rudimentary PDA was ligated before establishment of CPB. The postoperative period was uneventful and the patient was discharged in stable condition. The patient has been on regular follow-up. Postoperative follow up echocardiogram revealed normal pulmonary venous circulation with successful ASD closure with paradoxic IVS motion.Left atrium diameter was 32 mm, left ventricular diameter was 26 mm with normal pulmonary artery systolic pressure. (Figure 4)

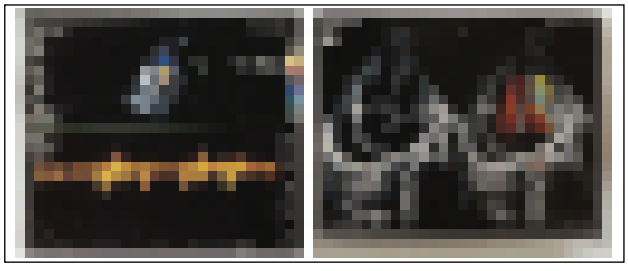

Figure 4: Echocardiography demonstrates communication of all pulmonary veins to left atrium.

Discussion

Normal embryologic development of the pulmonary venous system involves creation of a connection between the left atrium and the pulmonary venous plexus, and subsequent regression of systemic-to-pulmonary venous connections. Inappropriate connection of the pulmonary venous system to the systemic venous system is termed anomalous pulmonary venous drainage. Total (totally) anomalous pulmonary venous connection (TAPVC) is a cardiac malformation in which there is no direct connection between any pulmonary vein and the left atrium; rather, all the pulmonary veins connect to the right atrium or one of its tributaries. Although not part of the malformation, a patent foramen ovale or atrial septal defect is present in essentially all persons with TAPVC and is necessary for survival after birth. No matter what the final connection or termination maybe, individual right and left pulmonary veins usually converge to form a common pulmonary venous sinus, which in turn connects to the systemic venous system. Except for an atrial communication, most infants presenting with severe symptoms from TAPVC have either no associated condition or a small or large patent ductus arteriosus. Patent ductus arteriosus is present in nearly all infants coming to operation in the first few weeks of life with pulmonary venous obstruction and, overall, in about 15% of cases. Ventricular septal defects occasionally occur. However, more than one third of cases coming to autopsy, few of which are infants, have other major associated cardiac anomalies.[8] These include tetralogy of Fallot, double-outlet RV, interrupted aortic arch and other lesions.[9] The combination of TAPVC with other major cardiac anomalies is especially likely to occur when there is atrial isomerism.[10] Other associations have been identified. Esophageal varices can occur in obstructed TAPVC, and these are likely caused by obstructed veins. Hypoplasia of the small pulmonary arteries has recently been identified in obstructive TAPVC.[11] Patients without significant pulmonary venous obstruction present in infancy or early childhood with signs and symptoms related to the presence of a large left-to-right shunt. These patients have dyspnea, poor feeding, and poor growth. They may have cyanosis on examination, but this manifestation is usually mild. Other findings include a second heart sound that is split and a systolic flow murmur caused by increased flow across the pulmonary valve.

Chest radiograph findings are increased pulmonary vascularity as a result of the large left to right shunt created by drainage of pulmonary venous return into the right side of the heart, prominence of the pulmonary artery shadow and the right atrium silhouette often exists. In supracardiac drainage, the prominence of the upper mediastinal silhouette can create the classic "snowman" or figure-eight appearance. Two-dimensional (2D) echocardiography is remarkably accurate in assessing the morphology of TAPVC. Along with Doppler color flow interrogation, it is almost always diagnostic. Echocardiographic features include criteria for RV diastolic overload and an echo-free space posterior to the left atrium. After sternotomy and anterior pericardotomy, the common pulmonary venous sinus, lying behind the pericardium, is identified after lifting up the apex of the heart for a moment to visualize the retrocardiac portion of the pericardium. The right pulmonary artery, running parallel and just cephalad to the sinus, is also identified to avoid confusing it with the common pulmonary venous sinus. One method approaches the common pulmonary venous sinus from the right side of the heart. The posterior pericardial reflection is opened and the common pulmonary venous sinus is mobilized and opened. The posterior left atrial wall is opened, and the anastomosis is then made between the common pulmonary venous sinus and left atrium. The continuous suture line must not be pulled up so tightly as to purse-string the anastomosis and narrow it. The right atrium is opened, the foramen ovale closed, and the atrium closed. The remainder of CPB and reestablishment of myocardial perfusion are completed. A second method of repairing TAPVC is similar to that just described, but the common pulmonary venous sinus is exposed from the left side of the heart by lifting the cardiac mass out of the pericardial sac by retracting the cardiac apex anteriorly and rightward. This is best achieved by placing a retracting suture into the apical myocardium.

Conclusion

The long-term survival of surgical correction of TAPVC is good. The result of the surgical correction of this anomaly is associated with acceptable morbidity and mortality depending on early referral and surgery without progression of the pulmonary vascular pathology.

Acknowledgement

We are extremely grateful to Professor Dr. Lutfar Rahman, Chief and HOD of cardiac surgery, cardiac OT and cardiac ICU staff of Khwaja Yunus Ali Medical College and Hospital.

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