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Major AP Collateral from Sinus Node Artery in Case of Tetralogy of Fallot (TOF) - A Rare Entity

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Abstract: ***We are reporting a rare case of 17 - year - old female with tetralogy of Fallot (TOF), infundibular pulmonary stenosis, and coronary artery to pulmonary arterial fistulous communication arising from Sinus Node Artery and its surgical intraoperative management.***

Keywords: TOF, AP Collateral, Sinus Node Artery

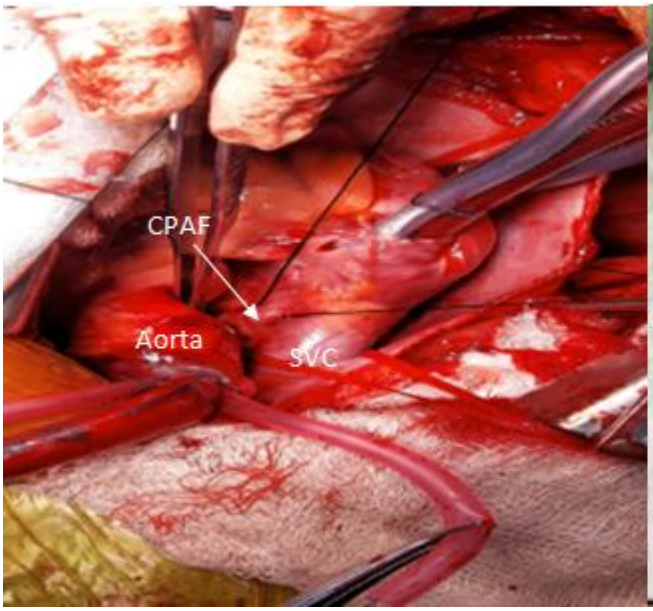
1. Introduction

There are few reports of large coronary artery to pulmonary arterial fistulous communications in the setting of tetralogy of Fallot. Here, we are reporting successful surgical management of a case having TOF with a coronary artery to pulmonary arterial fistulous communication arising from the Sinus Node Artery.

2. Case Report

A 17 - year - old female presented to our hospital with complaints of dyspnea (New York Heart Association Class III) and had been noted to be cyanotic since the age of 2 years. She also had history of hypercyanotic spells and hospital admission for the same. On examination, she was of average built with central cyanosis and grade II clubbing. Her pulse rate was 79/min with blood pressure of 104/62 mm Hg and baseline systemic arterial oxygen saturation (SaO2) was 71%. The cardiovascular system examination revealed normal S1, soft S2, and a continuous murmur best heard in the pulmonary area. . Chest roentgenogram showed normal sized heart with large aorta. Electrocardiogram revealed normal sinus rhythm with features of right ventricular hypertrophy. There was no evidence of myocardial ischemia. Two - dimensional echocardiogram revealed a large sub - aortic ventricular septal defect (VSD), RVH with severe infundibular pulmonary stenosis, and adequate sized branched pulmonary arteries with right sided aortic arch.

Cardiac CT Angiography revealed right sided aortic arch with mirror branching, large subaortic ventricular septal defect with RVH and tortuous and dilated SA Node artery with large aortopulmonary collateral arising from it. Catheterization and angiographic study confirmed the diagnosis of TOF with large sub - aortic VSD and aortic override. The aortic root injection showed the presence of a large communication from Sinus Node artery opacifying the pulmonary trunk. Selective coronary angiography confirmed the presence of a fistulous communication, arising from the SA Node artery opacifying the right and left pulmonary artery. Descending thoracic aortic injection revealed the absence of major large aortopulmonary collateral arteries (MAPCAs). Catheter intervention to achieve occlusion of the aortopulmonary collateral from SA Node artery by means of embolization was not performed to avoid risk of distal embolism, SA Node dysfunction and myocardial ischemia.



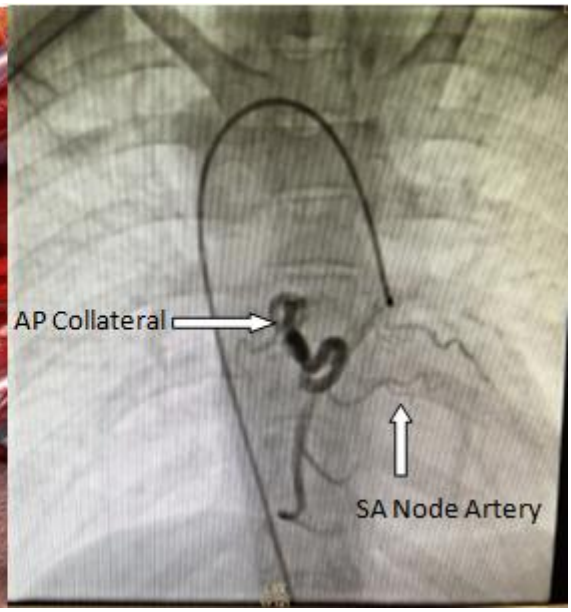


Figure 1 **Figure 2**

Figure1: Operative Photograph showing origin of Sinus Node Artery and A Fistulous tract [CPAF]originating from proximal part of SA Node Artery, medial to SVC, coursing over LA root and towards the hilum before terminating into Pulmonary Artery. The Fistulous tract was looped and ligated using 2 - 0 silk suture.

Figure 2: Pre - Op image of cathstudy showing origin of Sinus Node artery and AP collateral artery arising from SA Nodal artery.

Abbreviations

MAPCAs	Multiple large Aortopulmonary collateral arteries	LA	Left atrium
TOF	Tetralogy of Fallot	SVC	Superior Vena cava
RV	Right Ventricle	SA Node	Sino atrial Node
RVOT	Right ventricle outflow tract	VSD	Ventricular septal defect

Intraoperatively, the fistulous communication was found to be originating from the SA Node artery medial to SVC, coursing over the LA root and towards the right hilum. Before cardiopulmonary bypass, the fistulous communication was dissected medial to SVC and looped with 2 - 0 silk suture and ligated distal to the origin of sinus nodal artery prior to cardioplegic arrest. Intracardiac repair of TOF was performed using Dacron patch closure of the VSD, pulmonary valvotomy, and right ventricular outflow tract (RVOT) resection. . Ligation of the fistulous tract was accomplished without compromising Sinus Node coronary arterial blood flow. Post operative rhythm was NSR (Normal Sinus rhythm) and there was no evidence of SA Node dysfunction. The patient's postoperative recovery was uneventful. Two months following surgery, she is asymptomatic and without medications. Postoperative echocardiogram showed no residual VSD, no RVOT gradient, and normal biventricular function.

3. Comments

The cause of congenital coronary arteriovenous fistula is believed to be a failure of obliteration of the intramyocardial trabecular sinusoids with anomalous development of the intratrabecular spaces, through which blood is supplied to the myocardium during intrauterine life. Small coronary to pulmonary artery fistula in setting of TOF had been described. [1] However, the reports of large communication between pulmonary artery to coronary opacifying the whole trunk from the coronary injection have been recent only. [2, 3]The congenital coronary artery to pulmonary artery fistula is a rare anomaly with a reported prevalance of 1 in 50000. [4] However, congenital coronary artery to pulmonary artery fistulas are reported to occur commonly in patients with VSD and pulmonary atresia with reported incidence of 10%. [5]

In one report there was a fistulous communication reported between left main coronary artery to main pulmonary artery in a single coronary artery system. [2] The other report there was collateral from left circumflex artery to the right pulmonary artery. [3]

Physiologically such communications in TOF helps in increasing pulmonary blood flow and reduction in degree of cyanosis. The surgical importance of coronary artery fistula to pulmonary artery is that the communication needs to be identified and ligated before commencing cardiopulmonary bypass to prevent loss of volume to pulmonary vascular bed. Otherwise, there will be loss of cardioplegia solution to pulmonary circulation and incomplete myocardial protection during administration of cardioplegia.

4. Discussion

The identification of such communication is important preoperatively in patients of TOF especially when echocardiography reveals severe infundibular stenosis but the patient is not so cyanotic.

This case report demonstrates the rare finding of a fistulous connection between the coronary arterial circulation and the pulmonary arteries, with origin of the fistula from the SA Node Artery in the setting of TOF with careful ligation and obliteration of Pulmonary flow in it just distal to origin of SA Node blood supply.

Decisions to be made include the choice between obliteration of the fistulous connection by trans catheter technique or at the time of surgery. Embolisation of such a collateral is avoided as it may cause myocardial ischemia and arrhythmia during procedure and post coil inherent risk of SA Node dysfunction.

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