



Total Anomalous of Pulmonary Venous Connection- A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: Among Cyanotic cardiac diseases, Total Anomalous Pulmonary Venous Connection is the only condition involving venous system malformation. Total Anomalous Pulmonary Venous Connection is a rare congenital anomaly in which all 4 pulmonary veins have no direct communication with the left atrium.

Background: Almost one in a hundred newborn babies worldwide is affected by congenital heart disease. TAPVC is approximately 7-9 per 100,000 live births and it accounts for about 0.7- 1.5% of all congenital heart diseases. Congenital cardiac defects are the most common congenital anomalies in live births as well as the main cause of childhood Mortality and Morbidity in the developed nations.

Presentation of Case: A 5-month-old male child was admitted to 'Sri Satya Sai Sanjeevani Hospital, Naya Raipur, Chhattisgarh, India' on date 1 February 2021 with complaints of excessive sweating on the head while feeding, Tachypnoea, cyanosis and Lower Respiratory Tract infection. The child was diagnosed at 2 months of age and the child was brought to Sri Satya Sai Hospital for surgical management of TAPVC.

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Interventions: The treatment of patients was started immediately after admission. The surgical repair of obstructive mixed TAPV and PDA ligation was done under general anesthesia on date 02 February 2021.

Conclusion: In this report, we mainly focus on expert surgical management and excellent nursing care helped in managing the complicated case very nicely. The patient response was positive to conservative and nursing management. The patient was discharged without postoperative complications and satisfactory with recovery.

Keywords: Cyanosis; congenital heart disease; total anomalous pulmonary venous connection.

1. INTRODUCTION

Congenital heart disease affects nearly one out of every hundred newborn babies worldwide [1]. TAPVC is approximately 7-9 per 100,000 live births and it accounts for about 0.7- 1.5% of all Congenital Heart Diseases [2]. Congenital cardiac defects are one of the major congenital anomalies in live births as well as the main cause of infant Mortality and Morbidity in developed nations [3]. Among Cyanotic heart diseases, TAPVC is the only condition involving venous system malformation.² Total Anomalous Pulmonary Venous Connection is a rare birth defect in which all four pulmonary veins have no direct connection with the left Atrium [4]. For those who survive after birth with TPAVC, the PFO or ASD is essentially present.

Here we report a case of 5 months old baby boy with a total anomalous pulmonary venous connection.

2. PATIENT AND OBSERVATION

2.1 Patient Information

A 5-month-old male child was admitted to Sri Satya Sai Sanjeevani Hospital, Naya Raipur, Chhattisgarh, India on date 1 February 2021 with complaints of excessive sweating on the head while feeding, Tachypnoea, cyanosis and Lower Respiratory Tract infection.

The patient has a history of a recurrent episode of Lower Respiratory Tract infection, cyanosis,

excessive sweating on the head while feeding, Tachypnoea and the patient was diagnosed during taking treatment of lower respiratory tract infection. The child was diagnosed at 2 months of age and the child was brought by parents to Sri Satya Sai Hospital for surgical management of total anomalous of pulmonary venous connection.

2.2 Birth History

The patient weighed 2.5 kg and was delivered via Lower Segment Caesarean Section at full term.

2.3 Clinical Finding

On clinical findings the patient's general parameters height 61 cm, weight 5.7 kg, Body Mass Index 15.31 kg/m². In the neck central venous line was present, Pulmonary/- Cardiovascular sutures were present on the chest region due to surgical management. The pacing was present. Integumentary sutures were present on the chest region due to surgical management and the skin is dry. The musculoskeletal system he well-built.

2.4 Vital Signs

The vital sign of patient is temperature 100 F, pulse 140 beats/min, Respiration 30 breathe/min, Blood Pressure 75/36 mm of Hg.

2.5 Diagnostic Assessment

Table 1. Showing investigations of the patient

Complete Blood Count (CBC)	Hemoglobin was 14.1 g/dl Random Blood Glucose was 98 mg/dl (60-150 mg/dl) Total Red Cell Blood Count was 5.24 x 10 ⁶ /μL Total White Blood Cell Count was 8.5x 10 ³ /μL Haematocrit (HCT) was 40.2% Mean Corpuscular Volume was 76.7 fL, Mean Corpuscular Hemoglobin was 28.1pg Mean Corpuscular Hemoglobin Concentration was 36.6 g/dL. Total Platelet Count was 102x 10 ³ /μL.
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Kidney Function Test (KFT)	Urea was 39 mg/dl (10-50 mg/dl) creatinine was 0.5 mg/dl (0.6-1.1 mg/dl)
Liver Function Test (LFT)	Alkaline Phosphate was 94 IU/L (39-117 IU/L) Total bilirubin was 0.6 mg/dl (1.1 mg/dl) SGOT was 50 IU/L (up to 37 IU/L) SGPT was 43 IU/L (up to 40 IU/L) Total Protein was 6.3 g/dl (6.6-8.7) g/dl Albumin was 4.2 g/dl (3.8-5.1g/dl)
Blood Group	A+ RH factor: Positive Sickling Test: Negative HIV Test: Non-Reactive HCV Test: Non-Reactive HBS Ag: Non-Reactive
Blood coagulation factor	Bleeding time: 2 min 35 second Clotting Time: 5 min 13 second
Atrial Blood Gases	Arterial Blood pH: 7.31 Partial Pressure of Carbon dioxide (pCO ₂): 54 mmHg Partial Pressure of Oxygen (pO ₂): 63mmHg Bicarbonate (HCO ₃): 27.2 millimoles/L
ECG Finding	ECG revealed Sinus Tachycardia, Slight ST-Elevation and Right Ventricular Hypertrophy.

Table 2. Treatment

Sr. No.	Name of Drug	Dose	Route	Frequency	Drug Action
1.	Injection Piptaz (Piperacillin sodium and Tazobactam sodium)	350 mg	IV	TDS	Antibiotic
2.	Injection Targocid	50 mg	IV	BD	Antibiotic
3.	Injection Pantoprazole	5 mg	IV	BD	Antacid
4.	Injection Paracetamol	80 mg	IV	TDS	Antipyretic
5.	Nebulization Levolin	$\frac{1}{2}$ Respule	Nasal	QID	Bronchodilator
6.	Injection Meropenem	200 mg	IV	TDS	Antibiotic
7.	Tablet Sildenafil	6.25 mg	Oral	QID	Vasodilator

2.6 2D ECHO Findings

In 2D Echo finding situs levocardia, AV-VA concordance, Dilated Right Ventricle, Dilated Right atrium, Right ventricular hypertrophy is present, Supracardiac TAPVC, moderate size Ostium Secundum ASD (6 x 7mm) with Left to right shunt, Trivial TR with PG- 40 mm Hg, Intact IVS, Mild PS, Doming PV with flow acceleration across valve gdt(Gradient) - 27 mm Hg, all 4 pulmonary veins behind LA draining left Innominate vein and draining to Left Ascending Aorta, Mild flow acceleration in verticle vein with MG- 5 mm Hg, AoV- 8.8 mm (+1.09z), MV- 14.3 mm(+0.38z), PV- 14.6 mm (+3.17z), mild PAH, Dilated MPA & confluent branch PA's, Left aortic arch with normal branching pattern, No LSVS/CoA, God biventricular contractility.

2.7 Therapeutic Intervention

General measures to check the vital sign (Temperature, Pulse, Respiration and Blood Pressure), to check the patency of the airway, to

maintain fluid and electrolyte balance, to prevent infection and prevent complications like a pulmonary hypertensive crisis, pulmonary edema, Rhythm disorder are mandatory. Health management includes medication administration, rest and a healthy diet.

3. MANAGEMENT

3.1 Surgical Intervention

The surgical repair of obstructive mixed TAPV and PDA ligation was done under general anesthesia on date 02 February 2021.

3.2 Nursing Management

The postoperative patient was on a ventilator for three days. For that, the patient was postoperatively under strict observation of on-duty staff. Intravenous fluid administered as per calculation & Administered medication as prescribed. Observation and reading of the

character of the drainage were done postoperatively. Packed Red Cell & Platelets were given, care of the wound and the daily dressing was done. Intake and output were maintained 2 hourly. Vital signs were recorded strictly. Monitor Blood Pressure, central venous pressure and Oxygen Saturation of the patient. Check the proper position of the transducer, check for a surgical site for a sign of bleeding and infection. The overall response to treatment of the patient was positive and patient condition improved progressively. The patient was shifted into a step-down ICU from ICU after recovery. The excellent nursing care and the patient parents reported to the nursing staff. Complete discharge procedure was explained by nursing staff to the patient's parents along with medication prescribed at home as advised by the cardiologist. The patient was discharged from the hospital after 7 days of surgery without any complications.

3.3 Therapeutic Diet Plan

Provide high caloric formula milk or breast milk to the infant.

4. DISCUSSION

This was a very rare case of congenital heart defects. CHDs are anatomically, clinically, epidemiological and developmentally heterogeneous [1]. Total Anomalous of Pulmonary Venous Connection is a rare birth defect in which all four Pulmonary veins have no direct connection with the left Atrium [4]. The incidence of TAPVC is 0.008% of live births, but it occurs 0.7- 1.5% in cases of congenital heart disease [5]. The most commonly used classification system of TAPVC is Darling Classification and which was introduced in 1957. TAPVC is classified into four categories according to the sites where the abnormal connection occurs. Type I TAPVC: Supracardiac type, which is the most common type, Type II TAPVC: Cardiac type, Type III TAPVC: Intracardiac type and Type IV TAPVC: Mixed type. 1,2 In our case represents the Supracardiac type of TAPVC [6,7].

The pulmonary veins most commonly drain to a confluence posterior to the left atrium in the Supracardiac type. 1 However in our case all 4 pulmonary veins behind LA draining left Innominate vein and drain to Left Ascending Aorta [8,9].

Obstruction may occur in any type of TAPVC but is most commonly seen in the infra-cardiac type. There are various complications of obstructed TAPVC leading to hypoxemia, pulmonary hypertension and pulmonary vascular obstructive disease [5,10]. However, our case represents Supracardiac type of TAPVC with low O₂ saturation, mild cyanosis but no signs of pulmonary hypertension as the patient had PDA.

The cardiac pathology in patients with TAPVC shows interatrial septal defects, cardiomegaly, prominent pulmonary trunk, increased pulmonary vascularity on x-ray, low oxygen saturation ranging from 80% to 90% on the oximeter. However, in our case preoperatively oxygen saturation was 89% on the oximeter [11]. In the patient with TAPVC, there is right atrial enlargement with right ventricular hypertrophy [11]. Clinically patients with TAPVC may have a congestive cardiac failure or profound cyanosis due to interatrial communication and patency of pulmonary venous flow [12]. However, in our case, Supracardiac type of TAPVC was present; there was RA and RV dilatation, low oxygen saturation and mild cyanosis were present. There were no signs of congestive cardiac failure.

Due to high pulmonary vascular resistance and shunting blood flow through the foramen oval, TAPVC does not cause any problems during fetal development. However, because the heart of a TAPVC patient does not have a direct pulmonary veins connection to the left atrium, severe pulmonary congestion and pulmonary hypertension may result after the ductus arteriosus is closed [1]. However, in our case pulmonary hypertension was not revealed as the patient had PDA.

5. CONCLUSION

In recent years, an increasing number of studies on prenatal diagnosis and postnatal management of TAPVC has been published.

This case report is a contribution to the knowledge of rare congenital malformation of the heart.

CONSENT

While preparing case reports for publication parental informed consent has been taken from parents.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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