

A Rare Association of Pentalogy of Fallot with Situs Inversus Totalis Complicated by Brain Abscess in an Adolescent—Case Report

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ABSTRACT

Tetralogy of Fallot (TOF) is the most frequently diagnosed congenital cyanotic heart disease. It is often associated with additional findings, such as atrial septal defect (i.e., pentalogy of Fallot) or right sided aortic arch. Association of this pentalogy of Fallot with situs inversus totalis is rarely reported in paediatric literature and it can cause technical challenges to intracardiac repair. We report the case of pentalogy of Fallot with dextrocardia and situs inversus presenting as parieto-occipital abscess in a 12-year-old child. Complicated cardiac malformation like this when associated with brain abscess pose several challenges to the anaesthetist due to the altered haemodynamics and warrant a meticulous anaesthetic as well as surgical plan.

Keywords: Atrial septal defects, Dextrocardia, Heart defects/congenital, Tetralogy of Fallot

CASE REPORT

A 12-year-old boy was admitted with complaints of fever, headache and vomiting of 20 days duration. There was no history of loss of consciousness or focal neurological involvement. There was a history of infrequent episodes of cyanotic spell and exercise intolerance since 3 years of age. No history of palpitation or chest pain was there. He was not taken to any tertiary care centre due to financial constraints.

On general examination there was plethoric conjunctiva, marked central cyanosis and grade 3 clubbing. Neurological examination revealed neck stiffness and positive Kernig's sign. There was no focal neurological deficit. On cardiovascular examination, apex was in right 6th intercostal space 2cm lateral to mid-axillary line. On auscultation grade 3/6 ejection systolic murmur was heard, best at the right upper sternal border without any radiation. Respiratory system examination was within normal limits and per-abdomen no organomegaly was detected.

Blood tests revealed a haemoglobin level of 20.4 g%, haematocrit, of 59.3% and a raised Total Leucocytes Count (TLC) of 17,200cells/cmm. Other serum biochemical parameters were within normal limits. A 15-lead electrocardiogram yielded results consistent with dextrocardia [Table/Fig-1]. Chest radiograph showed dextrocardia with total situs inversus, without any apparent cardiomegaly [Table/Fig-2].

Transthoracic 2D echocardiogram revealed the anatomy of dextrocardia with situs inversus and Pentalogy of Fallot (POF): Ostium primum ASD, Inlet Ventricular Septal Defect (VSD) with Right

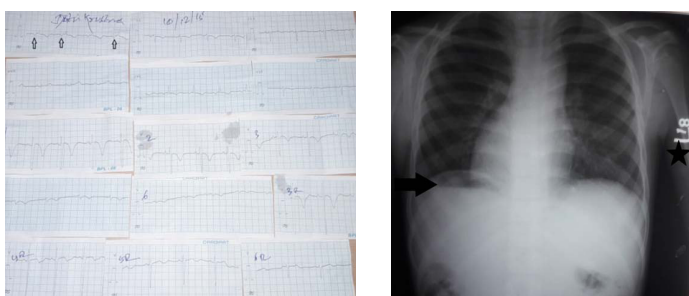
Ventricular (RV) outflow tract obstruction [Table/Fig-3]. There were no other anomalies like right sided aortic arch.

Computed Tomography (CT) scan of the brain revealed a well defined thin walled ring enhancing lesion in the right temporo-parietal lobe region (size 4.05cm× 3.02cm) without mass effect, probably pyogenic abscess [Table/Fig-4].

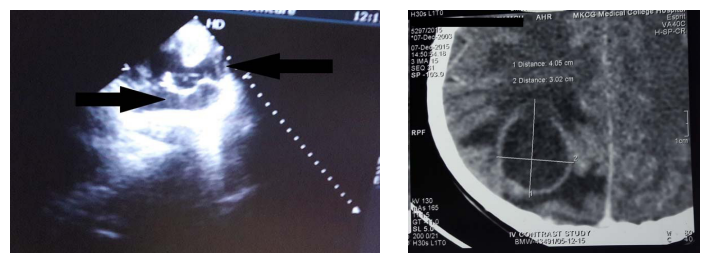
The patient was treated with parenteral antibiotics empirically (Ceftriaxone, Vancomycin for 6 weeks; Metroglol for 10 days) and steroids. After obtaining informed consent from parents, he was posted for emergency craniotomy and drainage of right temporo-parietal lobe abscess. Craniotomy was performed successfully and abscess was completely drained. Culture was sent and report showed *Enterococcus* species (sensitive to vancomycin, linezolid). Child is now asymptomatic with stable neurological picture. Patient was planned for surgical repair.

DISCUSSION

The POF is a variant of the more common Tetralogy of Fallot (TOF), comprising the classical four features with the addition of an atrial septal defect. Though it was mentioned by Niels Stensen and Edward Sandifort; later, a detailed analysis and description was made by the French physician Étienne-Louis Arthur Fallot in 1888, after whom it is named [1]. Situs inversus (also called situs transversus or oppositus) is a congenital condition in which the major visceral organs are on the opposite side as compared to their normal positions. In the most common situation, situs inversus totalis, there is complete transposition of all organs. Dextrocardia refers to a condition where heart is located on right side of the chest [2-4]. The estimated prevalence of congenital heart disease in individuals with situs inversus totalis is around 5–10% and the most common lesion is transposition of great vessels [5]. Ostium secundum type of Atrial Septal Defect (ASD) is found commonly



[Table/Fig-1]: ECG consistent with dextrocardia. Inverted p-waves in Lead-1 (arrow marks) and upright p-waves in Lead-aVF suggest situs inversus. Non progression of QRS complexes from V1 to V6 suggest dextrocardia. **[Table/Fig-2]:** Chest radiograph showing dextrocardia and situs inversus totalis. Arrow mark indicates fundic gas on right side and star mark indicates left side marked by radiologist.



[Table/Fig-3]: Transthoracic Echocardiogram showing components of pentalogy of Fallot. **[Table/Fig-4]:** CECT-Brain magnified film showing ring enhancing lesion possibly abscess of size 4.05cm× 3.02cm.

with POF than Ostium primum type [6]. Interestingly our patient had an ostium primum type of ASD.

After the initial diagnosis of dextrocardia we can treat patients with TOF and dextrocardia like any isolated TOF, with use of similar surgical procedures and postoperative monitoring. Old concept was, initial palliative systemic-to-pulmonary arterial shunt surgery, followed by definite repair surgery (usually when the patient was 3–5-year-old) to close the Ventricular Septal Defect (VSD) and resection of the Residual Volume (RV) outflow musculature [7,8]. But current school of thought is to go for a complete single-staged repair as early as 3 to 6 months of age, or even sooner if symptoms develop [7,8]. Decreased chance for development of aneurysm, ventricular dysfunction and rhythm changes make transatrial-transpulmonary approach a superior one as compared to transventricular approach [7].

Here, the altered spatial orientation of the cardiac structures is the major concern as far as patch closure of the VSD is concerned. The surgeon has to approach the VSD at a different angle in dextrocardia as compared to levocardia. Hence, there is a high chance for postsurgical heart block. Patients with cyanotic CHD are prone to developing brain abscesses and TOF is the leading cause for brain abscess [9]. This fact can be explained by the lack of filtering of blood by lung (due to right to left shunt) and hence organisms get an entry to brain. These septic foci later coalesce to form abscess.

Ideal anaesthesia management for a child with POF presenting for non cardiac surgery is very difficult. Ratio of Systemic Vascular Resistance (SVR) to Pulmonary Vascular Resistance (PVR) determines the amount of blood through R-L shunt. Greater the PVR and lesser the SVR, greater will be the shunt [9,10]. Therefore, the aim should be to reduce peri-operative test spells due to infandibular spasm, as it will worsen R-L shunt. Increased heart rate and effective myocardial contractility escalates the spasm, while hypotension (systolic BP <60mmHg) decreases the infandibular spasm. Similarly, hypotension (BP < 60mm Hg) can decrease SVR. Therefore, certain anaesthetic goals have to be aimed so as to avoid peri-operative cyanotic spells [10]. A case reports the incidence of a five-year-old child with brain abscess and an uncorrected POF, who underwent a successful emergency craniotomy [11].

CONCLUSION

Situs inversus totalis (situs inversus plus dextrocardia) can be very rarely associated with pentalogy of Fallot. In such a scenario altered spatial orientation of cardiac structures complicates the cardiac surgery and unfavourable hemodynamic complicates the non-cardiac surgeries like craniotomy for brain abscess.

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