



# TETROLOGY OF FALLOT; WITH DEXTROCARDIA AND SITUS INVERSUS: A CASE REPORT.

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**ABSTRACT:** Tetralogy of Fallot is the most common cyanotic congenital cardiac diseases. It is also associated with other cardiac abnormalities; however, its association with dextrocardia and situs inversus is rarely reported. We report a case of these findings in a 9-year-old boy, describe the patient's postoperative course, and review the medical literature relevant to these combined conditions.

**Key words:** dextrocardia, heart defects/congenital, situs inversus, tetralogy of Fallot.

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## INTRODUCTION

Tetralogy of fallot (TOF) is the most common congenital cyanotic heart defect with an incidence of 3.3 per 10, 000 live births. First diagnosed in 1888 by French physician Etienne-Louis Arthur Fallot and corrected for the first time in 1954 by Alfred Ballock.<sup>1,2</sup> There are variable other congenital abnormalities being reported with TOF. Dextrocardia is one of them. We report a boy with TOF associated with dextrocardia and situs inversus (SI), discuss the patient's postoperative course, and review the relevant medical literature.

## CASE REPORT

A 9 years old boy of sub-continent origin with 19kg weight and 116 cm came for the first time at the age of 7 in our hospital. He has the complaints of worsening of shortness of breath from mild to moderate exertion, there was no history of cyanosis and cyanotic spells. Past history was significant for only one time episode of severe chest infection at the age of 6 months. On examination there was no cyanosis or clubbing. Heart beat was found on the right side of chest. Cardiac auscultation showed a grade 2/6 systolic ejection murmur, best heard at the right upper sternal border and radiating to the axillae and back. His CBC revealed a hematocrit of 38.2. Chest X ray showed a right sided apex of heart. ECG showed findings of dextrocardia.

Abdominal Ultrasonography revealed situs inversus Echocardiography confirmed diagnosis of dextrocardia. Others findings included patent foramen ovale with left to right shunt, large perimembranous VSD with overriding of aorta 50%. It was associated with a bidirectional shunt. So a diagnosis of a pink TOF with dextrocardia and situs inversus was made. Cardiac catheterization showed single left sided superior vena cava. Right ventricular angiogram showed infundibular and valvular pulmonary stenosis. Right ventricular outlet tract angiogram (RVOT) showed adequate sized main and branches of pulmonary artery. Inferior vana cava (IVC) angiogram showed right sided IVC. Below heart it turned to left.

He underwent cardiac surgery with median sternotomy and routine cardiopulmonary bypass establishment, with surgeon standing on left side of patient pulmonary artery was opened. Pulmonary valve was normal so left as such. Right ventricular outlet tract was resected through pulmonary artery and right ventricle. Perimembranous subarteriolar VSD was closed with Dacron patch using interrupted suture technique. Pulmonary artery was closed with autologous pericardial patch. Total cross clamp time was 139 minutes and cardiac bypass time was 182 minutes.



Figure-1. Chest x ray showing right sided cardiac apex

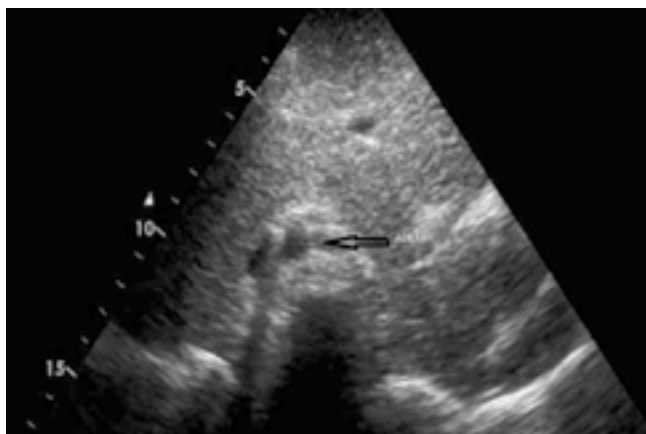


Figure-2. Echocardiography showing right sided aorta

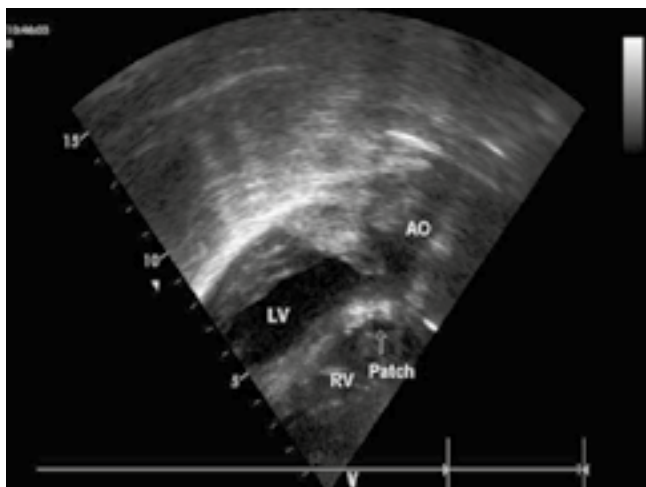


Figure-3. Post-surgical repair echocardiography showing Dacron patch in place

The patient made smooth and uneventful recovery. He was extubated uneventfully. He mobilized well and sent home on 7<sup>th</sup> postoperative day in good condition. His post-operative echo showed no residual VSD, mild tricuspid regurgitation, mild insignificant RVOT obstruction= 15mm of Hg, mild pulmonary regurgitation and good biventricular function.

## DISCUSSION

TOF is known to be associated with other cardiac abnormalities. Dextrocardia, patent ductus arteriosus, total anomalous pulmonary venous connection and unroofed coronary sinus are few to name. But incidence of association with each of these abnormalities is considerably low.<sup>3,4,5,6</sup> In 1952, Scragg and Denny reported the first documented case of TOF with situs inversus.<sup>7</sup> In a study by Abraham KA, 147 adults with TOF were studied. Only 2 patients had dextrocardia (prevalence, 1.4%).<sup>8</sup> In a study of 63 patients with dextrocardia by Ewans WN yielded 1 patient with TOF and situs inversus (prevalence, 1.6%).<sup>9</sup> In the experience from Green Lane Hospital, New Zealand between 1968 and 1978 only three out of 205 patients (1.5%) had dextrocardia and SI.<sup>10</sup>

Owing to the reverse anatomy, we have done correction with surgeon standing on left side of patient. This approach is also advocated by Talwar S.<sup>11</sup> Even though conduction system in such a heart travel through the same inferior margin of VSD; altered anatomical approach may be the reason for heart block in one of the study.<sup>12</sup> There was no heart block in our study.

Although our patient was of higher age as compared to patients who are routinely operated for TOF. But still there was no significant right ventricular dysfunction in our case. On literature review, case reports found were of blue TOF. While in our case it was a pink TOF. This characteristic and his favorable anatomy might be the reason for his uneventful recovery in our case.

To summarize, operating patient of TOF with dextrocardia is similar to operating patient with routine anatomy except with surgeon operating from the left side, Surgeon should be cautious


of conducting bundle being taken in the stitch during VSD patch closure because of altered spatial orientation.

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## AUTHORSHIP AND CONTRIBUTION DECLARATION

Sr. #	Author-s Full Name	Contribution to the paper	Author=s Signature
1	Dr. Tariq Waqar	Conceived designed the manuscript and is assumbash for arginality of this report	
2	Dr. Muhammad Usman Riaz	Helped is writing as reviewed this case report	