

## Tetralogy of Fallot with isolated levocardia in a young female

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### ABSTRACT

Tetralogy of Fallot is the most common cyanotic congenital heart disease. It consists of right ventricular outflow tract obstruction, a ventricular septal defect, abnormally located aorta and right ventricular hypertrophy. It usually occurs as an isolated anomaly with a normally placed heart and abdominal viscera. We present a case of a 19 years old female who presented with a prolonged history of shortness of breath (SOB) and cyanosis. After undergoing echocardiography and cardiac computed tomography angiogram (CTA), she was diagnosed to be a case of Tetralogy of Fallot (TOF) in association with situs inversus with levocardia also termed 'isolated levocardia'. The patient underwent surgical correction and she was asymptomatic with no residual cardiac defects on follow up after 6 months. Isolated levocardia is a rare condition that is usually associated with severe cardiac defects and a low life expectancy in untreated patients. It is unusual for it to be diagnosed in adults as in our case.

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Cardiovascular medicine; levocardia; tetralogy of fallot; congenital heart defects; TOF

## 1. Introduction

Scragg and Denny in 1952 reported the first documented case of TOF with situs inversus. That case was associated with dextrocardia [1]. Situs inversus is a condition in which the abdominal organs lie on the opposite side of the body. The heart may also lie on the opposite side and the term situs inversus with dextrocardia is used in such cases. Situs inversus commonly occurs with dextrocardia but rarely it may occur with a normally located left sided heart. Various terms have been used to describe the latter condition including 'situs inversus with levocardia', 'isolated levocardia' and 'situs inversus incompletus'. The estimated incidence of isolated levocardia is 1 per 22,000 in the general population and it ranges from 0.4% to 1.2% in congenital heart disease patients [2,3]. Most cases of situs inversus with dextrocardia have a morphologically normal heart and only 3–5% of such patients have cardiac anomalies [4]. On the other hand up to 95% of cases of isolated levocardia have associated cardiac deformities like right ventricular outflow tract (RVOT) obstruction, septal defects, inversion of cardiac chambers and transposition of cardiac chambers [5]. However, the association of the full picture of TOF with isolated levocardia is quite rare [6,7]. Patients with isolated levocardia frequently have significant cardiac anomalies and as a result have a lower life expectancy. However, if diagnosed and managed properly, they can have a better outcome.

## 2. Case presentation

A 19-years old girl from Afghanistan, who had been diagnosed with a cyanotic heart disease at an early age when a heart murmur and cyanosis were detected, presented with shortness of breath and acral cyanosis. Throughout her life, she had been experiencing increasing exercise intolerance, frequent episodes of cyanosis and substantial shortness of breath upon exertion. She had no chest pain or palpitations. Her medical history was otherwise notable only for recurrent chest infections i.e. pneumonia several times. Because of limited access to medical care, she had not undergone any medical therapy or surgical treatment for her cardiac condition. In April 2015 she was referred to Pakistan for treatment of her condition as it was worsening. Her body weight and height were normal and according to her age.

General Physical examination revealed mild cyanosis that was more apparent on the extremities and grade 2 clubbing. Cardiac auscultation revealed a systolic ejection murmur best heard at left upper sternal border and radiating to axilla and back. Her respiratory, abdominal, and neurological examinations were normal.

### 2.1. Investigations

The blood workup revealed hemoglobin of 18.7g/dl and a hematocrit of 57%. The rest of the red cell indices and white cells counts with differential counts

were in the normal range. Her liver function tests, renal function tests, clotting profile, and serum electrolytes were also normal. Pulse oximetry showed an O<sub>2</sub> saturation of 88%. Abdominal ultrasound revealed situs inversus.

Preoperative echocardiography revealed the anatomy of levocardia, a large subpulmonary ventricular septal defect (VSD), and severe subvalvular pulmonic stenosis with a gradient of 80 mmHg. It also revealed atrioventricular (AV) discordance with right anterior aorta arising from morphologically left sided right ventricle and left posterior pulmonary artery arising more than 60% from morphologically left sided right ventricle. There was associated congenitally corrected transposition of the great arteries and a double outlet right ventricle (DORV). The morphologically left sided right ventricle was also hypertrophied. There was a single left sided superior vena cava and an inferior vena cava that drained into right atrium. The interatrial septum was intact. Other findings included a left aortic arch and an intact interatrial septum.

Preoperative CTA of the heart revealed similar findings and showed a normal origin of coronary arteries. (Figure 1–3).

An abdominal ultrasound was done which confirmed the situs inversus with a normal spleen on the right side and a normal liver and gall bladder on the left side.

## 2.2. Treatment

The patient underwent elective surgical repair. During surgical repair, a median sternotomy was performed and cardiopulmonary bypass was established with aortic and bicaval cannulation. Aorta was cross clamped and cold blood cardioplegia was



Figure 1. CT chest showing levocardia.

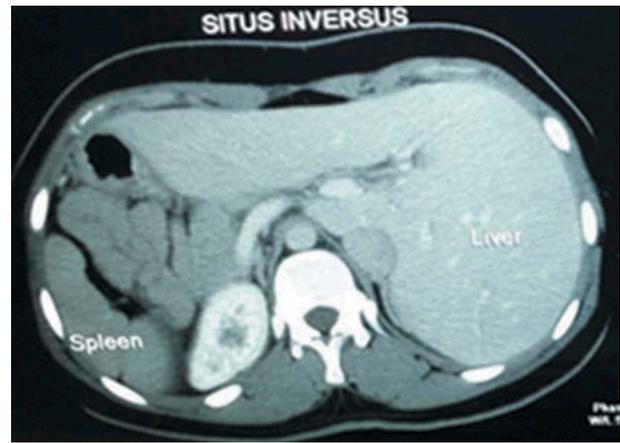


Figure 2. CT chest showing situs inversus.

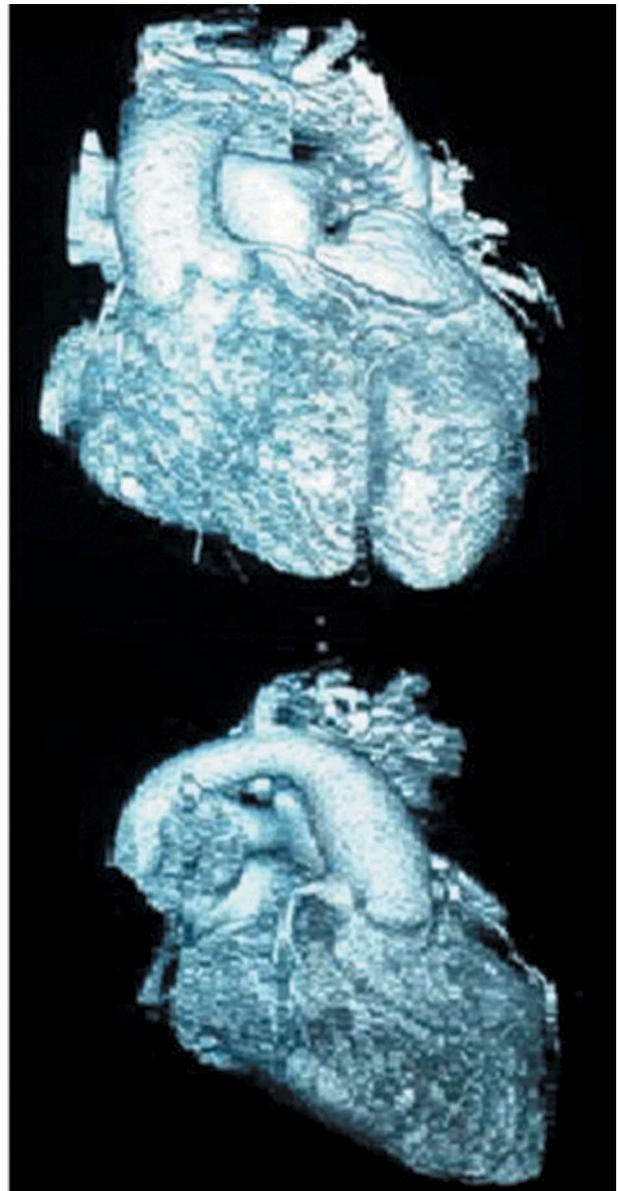


Figure 3. Aorta arising from the RV. Visible left main coronary artery.

infused. The heart was decompressed with a right superior pulmonary vein (RSPV) vent. Aorta was opened, both coronaries were visualised and a subvalvular VSD was identified. Pulmonary artery was also opened and the VSD identified again. Interrupted pledgeted prolene sutures were placed from the aortic side to close the VSD. The aortic valve was intact and the pulmonary annulus was small so a pulmonary artery incision was extended across the pulmonary valve into the left ventricular outflow tract (LVOT). Also, a subpulmonic fibrotic ring was resected. Left ventricular outflow tract obstruction was relieved. A transpulmonary pericardial patch was placed. The aorta was cleaned and closed with prolene. The patient was weaned off cardiopulmonary bypass without any problem.

### 2.3. Outcome and follow-up

The patient's postoperative course was uncomplicated and she was discharged from the hospital on her sixth post-operative day on spironolactone, sildenafil, and aspirin.

The patient was evaluated one month and six months after her discharge. The patient had no cardiac symptoms and was gaining weight. Cardiac auscultation didn't reveal any significant murmur. Electrocardiogram (ECG) showed sinus rhythm. Two-dimensional echocardiograms showed no residual VSD, no pericardial or pleural effusion, and normal biventricular systolic function.

## 3. Discussion

In the USA, the incidence of TOF is 3.9 per 10,000 live births and it accounts for 7–10% of all congenital heart diseases [8]. It was initially described by Niels Stensen in 1672, by Edward Sandifort in 1773, and by the French physician Étienne-Louis Arthur Fallot in 1888 after whom it is named [9]. TOF consists of RVOT obstruction, a VSD, an abnormal position of the aorta, and right ventricular hypertrophy. The severity of the condition depends on the degree of RVOT obstruction. Mild obstruction may present as cyanosis and decreased exercise tolerance later in life while severe obstruction presents in the neonatal period. There may be other anomalies associated with TOF that include a right aortic arch (25%), atrial septal defect (10%), branch pulmonary artery anomalies (30%), and patent foramen ovale. There are many anatomical variants of TOF including TOF-pulmonary atresia, TOF-absent pulmonary valve, TOF-atrioventricular septal defects, and TOF-double outlet right ventricle as in our case [10]. Even though it is a relatively common congenital heart disease, its association with situs inversus is quite rare. A comprehensive literature search showed only two cases of TOF in association with isolated levocardia [8,10].

Our patient had severe RVOT obstruction, a VSD and double outlet right ventricle with both the aorta and pulmonary artery arising from the morphologically left sided right ventricle. The abdominal viscera of our patient were located on the opposite side of the body but were morphologically normal. The rest of the thoracic structures were also solitus i.e. on their normal position except for the superior and inferior vena cavae which were located on the left side and drained into the right atrium.

The underlying pathogenesis of isolated levocardia is not fully understood [11]. The condition may be diagnosed via a detailed history and physical examination, chest radiography, EKG, and cardiac imaging [2]. Cardiac imaging studies include computed tomography (CT) or cardiac magnetic resonance imaging (CMR). Cardiac CT is helpful in delineating the position of visceral organs, the cardiac apex, intracardiac anatomy and branching of the great vessel. However, cardiac CT involves significant exposure to radiation and may have harmful effects on children. CMR can be utilized instead in these patients as it does not involve exposure to ionizing radiation. Cardiac function and cardiac volumes can also be measured more accurately with CMR [12,13].

Surgical correction is advised for all patients with TOF. Even though some patients with uncorrected TOF can live long lives, surgical correction has consistently shown to improve survival [14]. Initially a two-step procedure used to be performed with an initial systemic-to-pulmonary shunt for palliation and a later complete repair that involved closure of the VSD and correction of the outflow tract obstruction. However, since the advent of cardiopulmonary bypass and cardioplegia, a single step procedure is now recommended and at an earlier age [15]. Asymptomatic patients are now recommended to undergo correction at 3–6 months of age and severely symptomatic patients should undergo correction even earlier than that [10,16]. Our patient was known to have a cyanotic heart disease but could not undergo surgical correction early in her life because of a lack of access to medical facilities. However, when her shortness of breath and cyanotic episodes became unbearable, her family shifted her to Pakistan for medical treatment.

The transventricular approach which was previously used is frequently replaced with a transarterial and a transpulmonary approach as was the case in our patient. It has been shown to result in lesser ventricular injury and scar tissue formation and reduces the incidence of long term complications like arrhythmias, conduction abnormalities, and dilation of the ventricles [2].

The prognosis for TOF patients after surgical correction is great with 85–90% survival into adulthood [10]. However the prognosis of situs inversus with levocardia is quite poor and only 5–13% of patients survive for more than five years mainly due to the severity of an associated cardiac abnormality [2].

#### 4. Conclusions

- (1) Tetralogy of fallot is a common congenital heart disease however its association with situs inversus and isolated levocardia is quite rare.
- (2) Isolated levocardia patients usually have associated severe cardiac anomalies and a reduced life expectancy unless surgical correction is performed.
- (3) Surgical correction of TOF is recommended at a much earlier age, even in asymptomatic patients and a single step procedure with a transarterial or a transpulmonary approach is preferred.
- (4) TOF patients who undergo surgical correction usually have a favorable outcome.

#### Disclosure statement

No potential conflict of interest was reported by the authors.

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#### References

- [1] Scragg JN, Denny M. Dextrocardia, Tetralogy of Fallot, and situs inversus: report of a case. *S Afr Med J*. 1952;26:1025–1028.
- [2] Vijayakumar V, Brandt T. Prolonged survival with isolated levocardia and situs inversus. *Cleve Clin J Med*. 1991;58:243–247.
- [3] Budhiraja S, Singh G, Miglani HPS, et al. Neonatal intestinal obstruction with isolated levocardia. *J Pediatr Surg*. 2000;35:1115–1116.
- [4] Winer-Muram HT, Tonkin IL. The spectrum of heterotaxic syndromes. *Radiol Clin North Am*. 1989;27:1147–1170.
- [5] Normal and abnormal cardiac position (situs anomalies). [cited 2014 Jul 30]. Available from: <http://www.fetalultrasound.com/online/text/7-004.html>.
- [6] Gojo S, Kawachi K, Fukutomi M, et al. [A successful operation of isolated levocardia with left atrial isomerism, polysplenia and tetralogy of Fallot]. *Nihon Kyobu Geka Gakkai Zasshi*. 1993;41:655–659.
- [7] Kumar M, Yadav NK, Nayak B. Isolated laevocardia with tetralogy of Fallot. *J Indian Med Assoc*. 1985;83:217–218, 224.
- [8] Centers for Disease Control and Prevention (CDC). Improved national prevalence estimates for 18 selected major birth defects—USA, 1999–2001. *MMWR Morb Mortal Wkly Rep*. 2006;54:1301–1305.
- [9] Animasahun BA, Madise-Wobo AD, Falase BA, et al. The burden of Fallot's tetralogy among Nigerian children. *Cardiovasc Diagn Ther*. 2016;6:453–458.
- [10] Starr JP. Tetralogy of fallot: yesterday and today. *World J Surg*. 2010;34:658–668.
- [11] Jo DS, Jung SS, Joo CU. A case of unusual visceral heterotaxy syndrome with isolated levocardia. *Korean Circ J*. 2013;43:705–709.
- [12] Bartram U, Fischer G, Kramer HH. Congenitally interrupted inferior vena cava without other features of the heterotaxy syndrome: report of five cases and characterization of a rare entity. *Pediatr Dev Pathol*. 2008;11:266–273.
- [13] Yoo S-J, Kim YM, Choe YH. Magnetic resonance imaging of complex congenital heart disease. *Int J Cardiac Imaging*. 1999;15:151–160.
- [14] Lindberg HL, Saatvedt K, Seem E, et al. Single-center 50 years' experience with surgical management of tetralogy of Fallot. *Eur J Cardio-Thorac Surg*. 2011;40:538–542.
- [15] Fraser CD Jr, McKenzie ED, Cooley DA. Tetralogy of Fallot: surgical management individualized to the patient. *Ann Thorac Surg*. 2001;71:1556–1563.
- [16] Apitz C, Webb GD, Redington AN. Tetralogy of Fallot. *Lancet*. 2009;374:1462–1471.