Case Report

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Pentalogy of Fallot

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Abstract

The pentalogy of Fallot is a variant of the more common tetralogy of Fallot, comprising the classical four features with the addition of an atrial septal defect or patent ductus arteriosus.

The five features therefore are:

1. Ventricular septal defect (VSD)

2. Right ventricular outflow tract narrowing or complete obstruction

3. Right ventricular hypertrophy

4. Over riding aorta

5. Atrial septal defect (ASD).

Keywords: Pentalogy of Fallot; VSD; Right ventricular outflow tract narrowing; Right ventricular hypertrophy; Over riding aorta; ASD.

Introduction

Tetralogy of Fallot, also known as Fallot's syndrome or Fallots tetrad, has four key features. A ventricular septal defect (a hole between the ventricles) and many levels of obstruction from the right ventricle to the lungs (pulmonary stenosis) are the most important. Also, the aorta (major artery from the heart to the body) lies directly over the ventricular septal defect, and the right ventricle develops thickened muscle. Because the aorta overrides the ventricular defect and there is pulmonary stenosis, blood from both ventricles (oxygen-rich and oxygen-poor) is pumped into the body. Sometimes the pulmonary valve is completely obstructed (pulmonary atresia). Infants and young children with unrepaired tetralogy of Fallot are often blue (cyanotic) as in the present case. The reason is that some oxygen-poor blood is pumped to the body. When the above condition is associated with atrial septal defect, it is called Pentalogy of Fallot.

Case Summary

5 month female baby was admitted in our hospital with complaints of breathlessness, cough since last 7days. She was having history of recurrent respiratory tract infections since she was 1month of age. She was having history of 1 episode of bluish discoloration of tongue, nail beds in last 1month. She had been treated for her previous illnesses by local practitioner by symptomatic treatment. Her parents had family history of 3rd degree consanguineous marriage. She was 6 months 3 weeks preterm vaginal hospital delivery with poor cry and birth weight of 750gms. Our case is 4th issue of her parents. First two baby were still births (1st male and 2nd female) We admitted the girl to our hospital in paediatric

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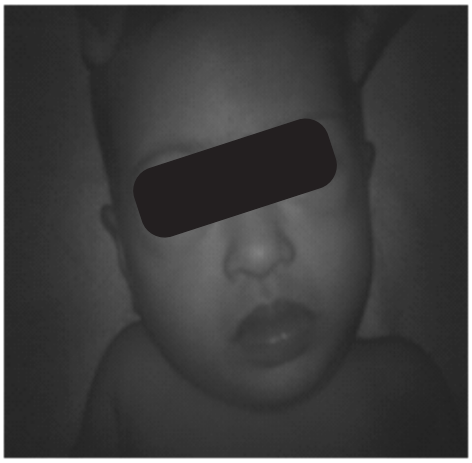
Cyanosis of Lips & Cyanosis of Tongue



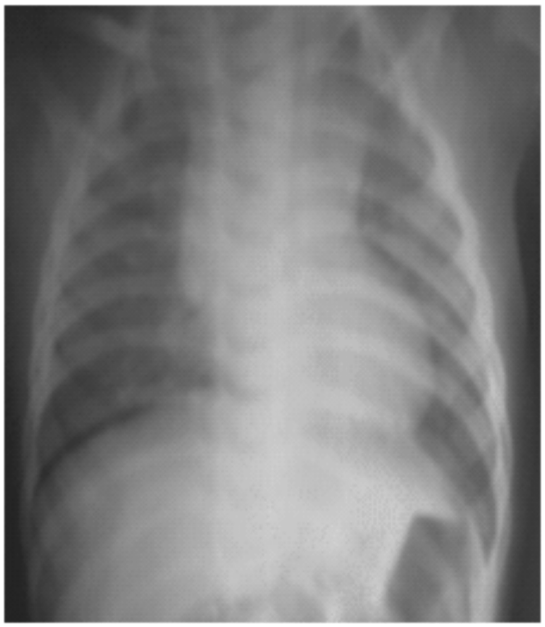
ward for further investigations & management.

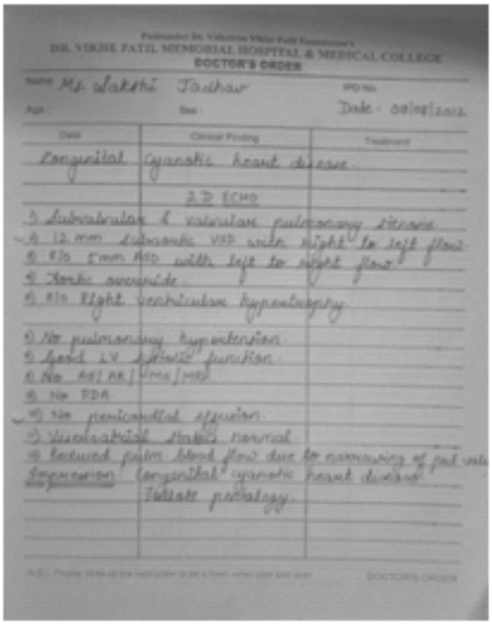
On examination, her pulse was 118beats/minute & blood pressure was 85/50 mmHg. Central cyanosis was present. On examination of cardiovascular system, she was having pansystolic murmur. The murmur was prominent over left parasternal, mitral & aortic areas. No parasternal thrill was present.All other systemic examinations were within normal limits.

Her investigations were as follows:

- Hb: 10.9 gm%
- TLC: 8400/cmm

X-ray Chest Showing Boot 2D-ECHO Report Shaped Heart with Cardiomegaly





DR. VIKHE PATIL MEMORIAL HOSPITAL & MEDICAL COLLEGE
DOCTOR'S ORDER

Name: Ms. Vakshi Jadhav
Age: Sex: Date: 09/05/2012

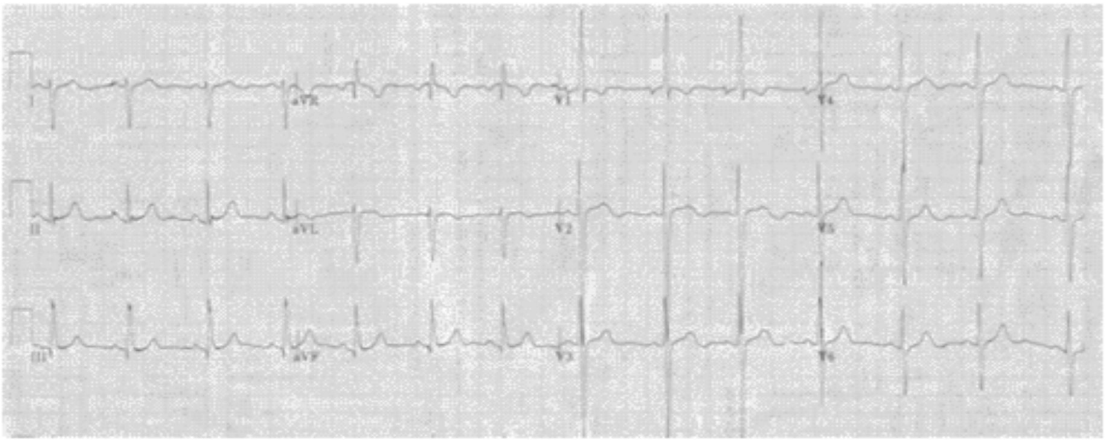
Date	Clinical Finding	Treatment
	Congenital cyanotic heart disease	
	2D ECHO	
	1) Subvalvular & valvular pulmonary stenosis	
	2) 12 mm diameter VSD with right to left flow	
	3) ASD 5mm with left to right flow	
	4) Aortic overriding	
	5) RVH Right ventricular hypertrophy	
	6) No pulmonary hypertension	
	7) Good LV systolic function	
	8) No AS/AR/MS/MR	
	9) No PDA	
	10) No pericardial effusion	
	11) Visceroatrial status normal	
	12) Reduced pulm blood flow due to narrowing of pul valve	
	Impression: Congenital cyanotic heart disease Tetralogy pathology	

- P – 25, L – 70, E – 03, M – 02, B – 00.
- Platelet count: 2.28 lacs/cmm.
- ESR: 12 mm at end of 1st hr.
- ECG revealed right ventricular hypertrophy.
- Chest x-ray showed boot shaped heart with cardiomegaly.
- 2D ECHO & Color Doppler of heart states presence of a ventricular septal defect of 12 mm diameter with bidirectional shunt, atrial septal defect(septum secondum defect) of 5 mm diameter with left to right shunt, presence of overriding of aorta & reduced pulmonary blood flow due to narrowing of pulmonary valve(valvular and subvavular stenosis). There was evidence of right ventricular hypertrophy (RV wall thickness -18 mm)

Discussion

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease in children but occurs rarely in adults. Its etiology is still not clear but its embryogenesis involves anterior deviation of the septal insertion of the infundibular ventricular septum resulting in

ECG Showing R Wave in Lead V1 with RS in V2 (Sudden Transition), Right Axis Deviation, No Q Waves in Lateral Leads Suggesting Decreased Pulmonary Blood Flow



mal-alignment of the outlet septum, ventricular septal deviation (VSD), pulmonary outflow tract stenosis and aortic over-ride. In addition, right ventricular hypertrophy is noted secondary to pulmonary stenosis.

Clinical presentation consists of cyanosis, clubbing of the fingers, polycythemia and exertional dyspnea. Cyanosis and polycythemia may be noted in the newborn. The extent of cyanosis depends on the balance of systemic and pulmonary vascular resistance, which depends on the severity of right ventricular outlet obstruction. The more severe the obstruction, the more blood flows into the left side. Therefore, the more severe the pulmonary stenosis, the more protection from lung disease is noted. Mild pulmonary stenosis may present with mild cyanosis or even acyanosis, termed pink TOF or acyanotic TOF. Patients with this condition may have lung disease and may expire in early childhood if no repair or palliative surgery is performed.

Signs and Symptoms

Tetralogy of Fallot results in low oxygenation of blood due to the mixing of oxygenated and deoxygenated blood in the left ventricle via the VSD and preferential flow of the mixed blood from both ventricles through the aorta because of the obstruction to flow through the pulmonary valve. This is known as a right-to-left shunt. The primary symptom is low blood oxygen saturation with or without cyanosis from birth or developing in the first year of life. If the baby is not cyanotic then it is sometimes referred to as a "pink tet." Other symptoms include a heart murmur which may range from almost imperceptible to very loud, difficulty in feeding, failure to gain weight, retarded growth and physical development, dyspnea on exertion, clubbing of the fingers and toes, and polycythemia.

Children with tetralogy of Fallot may develop "tet spells." The precise mechanism of these episodes is in doubt, but presumably results from a transient increase in resistance to blood flow to the lungs with increased preferential flow of desaturated blood to the body. Tet spells are characterized by a sudden, marked increase in cyanosis followed by syncope, and may result in hypoxic brain injury and death. Older children will often squat during a tet spell, which increases systemic vascular resistance and allows for a temporary reversal of the shunt.

Treatment

Complete surgical correction is now the most important and standard treatment of TOF. Several factors, such as old age, high hemoglobin level, pulmonary artery hypoplasia and a diminutive left ventricle, have been identified as risk factors for operative mortality in many previously published series. Palliative surgery includes the B-T shunt or Potts shunt, which constructs a

communicating shunt between the systemic and pulmonary circulation. However, the outcome is poor and it is no longer standard treatment for TOF except when the patient's condition means they are not suitable for repair. It may be a bridge from symptom relief to total correction. Medication is used for symptom relief only.

Prognosis

Mortality is about 3% in children and 2.5% to 8.5% in adults. The survival rate of patients who receive repair surgery is about 86% at 32 years follow-up and 85% at 36 years follow up; survival rates of un-operated TOF patients older than 10 years is about 30%, older than 20 years 11%, older than 30 years 6% and older than 40 years only about 3%.

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