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Cardiology

A CASE STUDY ON TETRALOGY OF FALLOT

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ABSTRACT Tetralogy of Fallot is the most common cyanotic heart condition in children who have survived untreated beyond neonatal age, with the need for an intervention in the first year of life. Tetralogy of Fallot is a birth defect that affects normal blood flow through the heart. It happens when a baby's heart does not form correctly as the baby grows and develops in the mother's womb during pregnancy.It accounts for 7% to 10% of congenital defects, affecting males and females equally and occurring in 3 to 5 of every 10,000 live births.In this article I report a case of a six years old patient with diagnosis of Cyanotic congenital heart disease, Tetralogy of Fallot with good sized pulmonary artries, good biventricular funtion and sinus rhythm.

KEYWORDS : Tetralogy of Fallot, Congenital Heart Disease, Survival, Cyanosis

INTRODUCTION

Tetralogy of Fallot is a type of congenital heart defect. Congenital means that it is present at birth.Tetralogy of Fallot is made up of the following four defects of the heart and its blood vessels:

1. A hole in the wall between the two lower chambers—or ventricles—of the heart. This condition also is called a ventricular septal defect.
2. A narrowing of the pulmonary valve and main pulmonary artery. This condition also is called pulmonary stenosis.
3. The aortic valve, which opens to the aorta, is enlarged and seems to open from both ventricles, rather than from the left ventricle only, as in a normal heart. In this defect, the aortic valve sits directly on top of the ventricular septal defect.
4. The muscular wall of the lower right chamber of the heart (right ventricle) is thicker than normal. This also is called ventricular hypertrophy.

Because a baby with tetralogy of Fallot may need surgery or other procedures soon after birth, this birth defect is considered a critical congenital heart defect. Congenital means present at birth.

Causes And Risk Factors

The causes of heart defects (such as tetralogy of Fallot) among most babies are unknown. Some babies have heart defects because of changes in their genes or chromosomes. Heart defects such as tetralogy of Fallot also are thought to be caused by a combination of genes and other risk factors, such as the things the mother or fetus come in contact with in the environment or what the mother eats or drinks or the medicines she uses.Tetralogy of Fallot causes low oxygen levels in the blood. This leads to cyanosis (a bluish-purple color to the skin).The classic form includes four defects of the heart and its major blood vessels:Ventricular septal defect (hole between the right and left ventricles)Narrowing of the pulmonary outflow tract (the valve and artery that connect the heart with the lungs)Overriding aorta (the artery that carries oxygen-rich blood to the body) that is shifted over the right ventricle and ventricular septal defect, instead of coming out only from the left ventricle Thickened wall of the right ventricle (right ventricular hypertrophy) Tetralogy of Fallot is rare, but it is the most common form of cyanotic congenital heart disease. It occurs equally as often in males and females. People with tetralogy of Fallot are more likely to also have other congenital defects.The cause of most congenital heart defects is unknown. Many factors seem to be involved.

Factors that increase the risk for this condition during pregnancy include:

- Alcoholism in the mother
- Diabetes
- Mother who is over 40 years old
- Poor nutrition during pregnancy
- Rubella or other viral illnesses during pregnancy
- Children with tetralogy of Fallot are more likely to have chromosome disorders, such as Down syndrome, Alagille syndrome, and DiGeorge syndrome (a condition that causes heart defects, low calcium levels, and poor immune function).
- Tetralogy of Fallot is rare, but it is the most common form of cyanotic congenital heart disease. It occurs equally as often in males and females. People with tetralogy of Fallot are more likely to also have other congenital defects.

PATHOPHYSIOLOGY

To better explain the pathophysiology of TOF, we should explore the 4 lesions in greater depth:

VSD

VSDs are categorised according to their location on the ventricular septum. The smaller membranous septum is located on the superior aspect whilst the larger muscular septum is on the inferior aspect.When the VSD involves parts of the membranous and muscular septum, this is called a perimembranous VSD – this is the commonest type associated with TOF. Other VSDs associated with TOF are muscular VSDs and doubly committed VSDs (located near both pulmonary and aortic valves)

The VSD is normally of a significant size, which causes the systolic pressures between the ventricles to equalise. In mild TOF, the left ventricular pressures remain higher than the right ventricle, thus blood shunts from left-to-right through the VSD. These patients are normally acyanotic.

In more severe disease, due to increased right ventricular pressure (secondary to PS – see below) the shunt direction reverses from right-to-left allowing mixing of deoxygenated ("blue") and oxygenated ("red") blood. This results in lower oxygenated ("purple") blood in the systemic circulation, hence patients are cyanotic.

Clinical Features

The history and clinical presentation depends on the severity of TOF which can be simplified into 3 categories:

Mild ('Pink' TOF)

These infants have mild PS/RVH and are usually asymptomatic. However, the disease normally progresses as the child and the heart grows thus by age 1-3 years they will develop cyanosis.

Moderate-Severe (Cyanotic TOF)

Infants born with moderate-severe PS may present in the first few weeks of life with cyanosis and respiratory distress. These infants may be prone to develop recurrent cyanosis and respiratory distress. These infants may be prone to develop recurrent chest infections or fail to thrive.

Extreme

These can be further divided into TOF with pulmonary atresia (10% of TOF patients) or absent pulmonary valves (6%).These are true 'duct dependent lesions' as the only way deoxygenated blood can flow into the lungs is through a patent ductus arteriosus (PDA). These infants are often detected on antenatal scans.

Examination

General: central cyanosis, clubbing

Palpation: thrill (depends on intensity of murmur) or heave (RVH)

Auscultation: Loud, single S2: due to closure of aortic valve in diastole with absent/reduced pulmonary valve closure (P2) depending

on the degree of stenosis.

Pansystolic murmur: best auscultated either mid or upper left sternal edge (LSE). The smaller the VSD the louder the murmur and vice versa.

Investigations Bedside:
ECG: may show signs of right axis deviation and RVH.

Bloods:
Microarray: if genetic syndromes suspected (e.g. dysmorphic features, multiple anomalies).

Radiological:
CXR: may show 'boot' shaped heart (RVH) and reduced pulmonary vascular marking

Echocardiogram: gold standard for confirmation of diagnosis.

Cardiac CT angiogram: to delineate anatomy especially if coronary abnormalities suspected (e.g. MAPCAs).

Cardiac MRI: in tertiary cardiac centres, MRI scans are of great value pre- and post- operatively to delineate the anatomy of the lesions and cardiac function. Useful if multiple aorto-pulmonary collateral arteries (MAPCAs) suspected. Interventional radiology:

Cardiac Catheter: this is performed under GA as an elective day case usually by the paediatric cardiologists. May be done as part of the pre-op assessment on the same day as the MRI to measure haemodynamics and delineate anatomy.

During childhood, most children do very well, with normal growth and development, but need regular cardiology care.

FUTURE CARE
1. Few children have symptoms, and most may participate in normal activities without restriction Medical care involves regular (yearly) evaluations by the cardiologist with electrocardiograms and echocardiograms to monitor for cardiac concerns that may occur late after repair
2. Continued or progressive obstruction in the right ventricle
3.Continued or progressive obstruction across the pulmonary arteries
4. Leaking (regurgitation) of the pulmonary valve
5. Rhythm problems
6. Heart-healthy lifestyle is encouraged
7. Healthy diet and appropriate weight for age
8. Regular exercise
9. Regular dental care is necessary
10. Higher incidence of learning and behavioral problems is observed
11. Attention deficit–hyperactivity disorder, learning disabilities, speech delays, etc
12. Early assessment and treatment
13. Adolescents are prepared to become more responsible for their own health care
14. Learn more about tetralogy of Fallot and long-term issues
15. Assume more responsibility for care over time
16. Most will achieve normal adult lifestyle (eg, work full-time, have children)

Case Report
A Six year old girl admitted to SMVDNSH with chief complaints of early fatiguability ,cynosis and history of syncope since 3 years of age . she was diagnostic to have cynotic congenital heart disease,Tatrology of fallot with good sized pulmonary arteries ,Good biventricular function and sinus rhythm and admitted in SMVDNSH for surgery .

On 04/12/2019 the patient got admitted with diagnosis of cyanotic congenital Heart disease, Tetrology of fallot with good sized pulmonary arteries ,gs.ood biventricular functions and sinus rhythm.

Intracardiac repair of tetrology of fallot with transannular pericardial patch,augmentation of main pulmonary artery with pericardial patch and PDA ligation done on 04/12/2019.

Intra-operative course was uneventful. Patient was shifted to ITU in heamodynamically stable condition with Dopamine 5mcg/kg/min and dobutamine 5mcg/kg/min support Patient was extubated on POD-0 .Inotropes were gradually tapered off on POD-2 .patient was shifted to ward on POD-3 after removing all lines and drains.Patient was recieved 1 whole blood ,1 PRBC,2 RDP and 2FFP during hospital stay.Patient was discharged on POD-5 in stable condition.At the time of discharge her vitals were normal.

Medication At Discharge

Sr. No.	Trade name	Generic name	Action	Indication	Side effect
1.	Syp. Digoxin 75mcg	Digox, Lanoxin	to treat certain heart problems such as heart failure.	To be taken orally once a day morning till further order	vomiting, diarrhea, stomach pain;fast, slow, or uneven heart rate; a light-headed feeling, like you might pass out;bloody or black, tarry stools;confusion, weakness, hallucinations, unusual thoughts or behavior;
2.	Syp. Furosemide 10 mg	Lasix	Furosemide is used to treat edema associated with a number of disorders, including congestive heart failure, nephrotic syndrome, and hepatic cirrhosis; it also has been used as an adjunct in the treatment of acute pulmonary edema.	To be taken orally twice a day morning and afternoon till further order	N nausea & vomiting.diarrhea. constipation.stomach cramping.feeling like you or the room is spinning (vertigo)dizziness.headache.blurred vision.
3.	Tab. Aldactone 6mg	Spironolactone	Spironolactone is used in the treatment of Hypertension (high blood pressure), Edema, low potassium and Heart failure. It treats oedema (fluid overload) associated with heart, liver, kidney or lung disease.	To be taken orally once a day morning till further order	Nausea, Vomiting, Leg cramps, Dizziness, Drowsiness, Confusion, Breast enlargement in male, Increased creatinine level in blood
4.	Tab. Enalapril 1.5 mg	Vasotec	highbloodpressure (hypertension), left ventricular dysfunction, and congestive heart failure	To be taken orally twice a day morning and afternoon till further order	a light-headed feeling, like you might pass out; chest pain;jaundice (yellowing of the skin or eyes);little or no urination; fever, chills, sore throat; or.high potassium--nausea, weakness, tingly feeling, chest pain, irregular heartbeats, loss of movement.

5.	Tab. Junior Lanzole 15mg	Lansoprazole(Prevacid)	Treatment ofGastroesophageal reflux disease (Acid reflux)Treatment of Peptic ulcer disease	To be taken orally once a day morning for 1 week	Nausea Headache Flatulence Diarrhea
6.	Syp. Ibugesic Plus 7 ml	Acetaminophen or Paracetamol	Ibugesic Plus Suspension 60 ml belongs to a class of painkillers called as non-steroidal anti-inflammatory drugs (NSAIDs). It relieves symptoms of muscle pain, arthritis pain, reduces fever.	To be taken orally sos for pain and fever	Nausea Vomiting Abdominal pain Heartburn Diarrhea

DISCUSSION

Without operation, few patients with ToF reach adulthood with an average life expectancy of 12 years. Ten per cent may survive to their 30s but only 3% reach their 40s or older. However, patient had an excellent post-operative and two-year follow-up profile. Thorough physical examination of newborns and a screening echo in the early life may aid in detecting the disease earlier.

CONCLUSION

TOF is a congenital heart defect that results in decreased blood flow to the lungs and is successfully repaired by surgery in infancy, allowing most patients a normal lifestyle. Lifelong follow-up is important to watch for problems such as an abnormal heart rhythm, leaking of the pulmonary valve, or poor function of the right ventricle. Continued research leading to new knowledge and treatments for congenital heart disease will improve the care of children in the future.

CONSENT

Consent was obtained from the patient family and they was assured about the confidentiality of the data obtained from them.

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