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Case Report

Successful pregnancy outcome in uncorrected tetralogy of Fallot with bidirectional shunt

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ABSTRACT

Tetralogy of Fallot (TOF), a cyanotic congenital heart disease, is the most prevalent type, constituting 10% of all congenital heart conditions. During pregnancy and childbirth, patients with uncorrected TOF can experience deterioration, posing a significant risk to maternal health and even mortality. A 30 year old patient was referred from a private clinic in view of uncorrected TOF in the third trimester of pregnancy. She was G3P1A1 who reported at 37 weeks 2 days period of gestation to the emergency room with history of breathlessness on routine activities for 10 days (NYHA III) and easy fatigability for 4-5 months and pain abdomen. Her previous antenatal and pre pregnancy period was uneventful with no history of cyanotic spells, dyspnea or palpitations. Electrocardiography showed sinus rhythm, right atrial enlargement, right ventricular hypertrophy with sudden transition of QRS in V2. Her echocardiography confirmed the findings of TOF. It revealed a large peri membranous VSD with bidirectional shunt with 50% overriding of aorta. There was non dysplastic severe pulmonary stenosis, right ventricular outflow tract narrowing with right ventricular hypertrophy, severe pulmonary hypertension, with a left ventricular ejection fraction of 56%. She underwent emergency cesarean section under general anesthesia in joint consultation of the obstetrics unit along with anesthesiologist, cardiologist, intensivist and neonatologist. Her cardiovascular status was never compromised. She was started on torsemide and was discharged on 6th post-operative day. She followed up with cardiologist after 4 weeks postpartum for further management.

Keywords: Uncorrected tetralogy of Fallot, High-risk pregnancy, Bidirectional cardiac shunt

INTRODUCTION

Tetralogy of Fallot (TOF), a cyanotic congenital heart disease, is the most prevalent type, constituting 10% of all congenital heart conditions. In TOF patients, a single developmental defect leads to anomalies: abnormal anterior and cephalad displacement of the infundibular portion of the interventricular septum. The defect results in four anomalies: pulmonary stenosis, right ventricular hypertrophy, overriding aorta, and non-restrictive ventricular septal defect.[1,2] Surgical intervention-either palliative or reparative-is necessary for most TOF patients to reach adulthood.[3] Uncorrected TOF cases are relatively rare, and most reported instances occur in developing countries.

This prevalence might be attributed to limited awareness and resources for early correction of this congenital anomaly. During pregnancy and childbirth, patients with uncorrected TOF can experience deterioration, posing a significant risk to maternal health (with a 62.5% morbidity rate) and even mortality (10%). Additionally, these cases have notable effects on fetal outcomes4.

Managing such cases presents challenges for obstetricians, cardiologists, and anesthetists, partly due to the scarcity of literature on the topic.

CASE REPORT

A 30-year-old patient was referred from a private clinic in view of uncorrected TOF in the third trimester of pregnancy. She was G3P1A1 who reported at 37 weeks 2 days period of gestation to the emergency room with history of breathlessness on routine activities for 10 days (NYHA III) and easy fatigability for 4-5 months and pain abdomen (Table 1). Her previous antenatal and pre pregnancy period was uneventful with no history of cyanotic spells, dyspnea or palpitations. She was a known case of TOF diagnosed at 4 years of age, and was advised a surgical correction, which the family refused. She had normal developmental milestones. She had normal growth with a height of 156 cm, and weighed 56 kg, her BMI was 22.4 kg/m^2. She was married for 11 years.

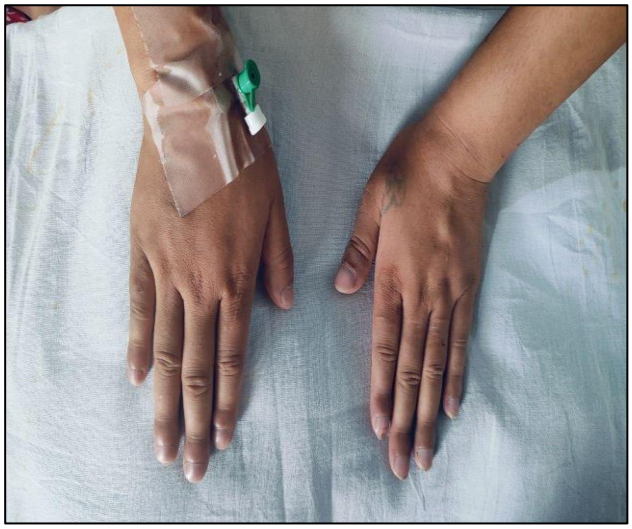

Figure 1: Clubbing of finger nails.

On examination patient was plethoric, had grade 4 clubbing, bilateral pitting pedal edema and cyanosis (Figure 1). Respiratory rate was 22 per minute. Cardiovascular system examination revealed a pulse of 88 beats per minute. The blood pressure was 120/80 mmHg, apex beat was localized in the 5th left intercostal space within the mid clavicular line and there was right parasternal heave and a systolic thrill. On auscultation, first and second heart sounds were normal, and grade 4/6 pan systolic murmur was heard which was loudest at the left lower sternal edge. Chest was clear. On abdominal examination, uterus was 32 weeks size with a single live fetus in longitudinal lie in cephalic presentation with decreased liquor [suggestive of fetal growth restriction (FGR)]. Investigations revealed that the hematocrit was 52%, hemoglobin was 14.5 gm/dl, platelet count was 270000/mm^3. O_2 saturation was 83% on room air. Her serum electrolyte, urea and creatinine levels were normal. Electrocardiography showed sinus rhythm, right atrial enlargement, right ventricular hypertrophy with sudden transition of QRS in V2. Her echocardiography confirmed the findings of TOF. It revealed a large perimembranous VSD with bidirectional shunt with 50% overriding of aorta. There was non dysplastic severe pulmonary stenosis, right ventricular outflow tract narrowing with right ventricular hypertrophy, severe pulmonary hypertension, with a left ventricular ejection fraction of 56%.

She had a spontaneous conception and her antenatal period was uneventful. Her antenatal care was under close observation of the high-risk pregnancy unit and cardiologist. She was not on any cardiac drugs. Her TIFFA scan and fetal 2D ECHO did not reveal any abnormality.

Table 1: Characteristics.

Characteristics	Results
Age of presentation (in years)	30
Gestational age of presentation	37 weeks 2 days
Symptoms at time of presentation	Pain abdomen
BMI (kg/m^2)	22.4
Antenatal period	Uneventful
LVEF	56%
Mode of delivery	Emergency LSCS
Anaesthesia	General anaesthesia
Post op	HDU management
Post op complication	Nil

Decision to perform emergency caesarean section was taken in joint consultation of the obstetrics unit along with anesthesiologist, cardiologist, intensivist and neonatologist at her presentation. She underwent a caesarean section under general anesthesia. Bilateral tubal ligation was done by modified Pomeroy's technique. Care was taken to prevent any fall in the systolic blood pressure and avoid any drop in the systemic vascular resistance. Post operatively shifted to obstetrics HDU. Her cardiovascular status was never compromised. She was started on torsemide and was discharged on 6th post-operative day.

She followed up with cardiologist after 4 weeks postpartum for further management. Decision of correction of the VSD will be taken after the resolution of pulmonary hypertension and after assessment of the cardiac status.

DISCUSSION

Uncorrected TOF in adulthood is quite rare, with only a few patients surviving to adulthood without surgical correction. When TOF occurs during pregnancy, it becomes even more uncommon. Pregnancy exacerbates hypoxia and cyanosis in these patients.

The underlying mechanism involves an increase in the right-to-left shunt due to decreased peripheral vascular resistance caused by vasodilation. Consequently, there is reduced pulmonary blood flow and an increase in aortic blood flow, leading to decreased arterial oxygen pressure

and saturation.[5-7] Mortality and morbidity rates are higher in uncorrected TOF patients during gestation, especially those with a history of syncope, polycythemia, and right ventricular hypertrophy. [8] The risk is further elevated when arterial oxygen saturation levels at rest fall below 85%.

This explains why our patient experienced worsened symptoms during pregnancy. Chronic hypoxemia also contributes to FGR observed in our patient. Additionally, our patient presented with polycythemia and thrombocytopenia. In uncorrected TOF, secondary polycythemia results from chronic hypoxemia due to the right-to-left cardiac shunt. It serves as a physiological compensatory mechanism to distribute oxygen more effectively to tissues.

Uncorrected TOF poses a life-threatening risk for both the mother and the fetus. It falls into the moderate-risk category for pregnancy with heart disease (Class 2A), and the mortality rate ranges from 5% to 15%.

The CARPREG score, a widely used risk assessment tool, indicates that our patient had a 27% risk of cardiovascular and maternal complications.[9,10] In all cases of cyanotic congenital heart diseases, there is an increased risk of fetal death (45-50%) and higher rates of premature delivery (30-50%) and FGR.[4,6] Maternal hypoxia contributes to these adverse outcomes. When hypoxemia is severe enough to raise hematocrit levels above 65%, pregnancy loss becomes nearly inevitable (close to 100%).[7,12] Additionally, infants born to TOF patients have an incidence of cardiac defects ranging from 3% to 17%.[13]

Given these risks, comprehensive management is crucial. Intensive monitoring is necessary throughout pregnancy, labor, and the postpartum period. Specific observations during pregnancy include blood gas analysis (especially measuring pO_2 and oxygen saturation) and monitoring hematocrit and hemoglobin levels. Fetal growth and well-being should be carefully assessed using techniques such as fetal ultrasound (USG/biometry), Doppler velocimetry, and cardiotocography.

In women with uncorrected TOF, vaginal birth is the preferred mode of delivery6.[14,16] Caesarean section is reserved for obstetrical indications. The rationale behind this preference lies in the fact that vaginal delivery typically results in less blood loss (around 400-500 cc) compared to caesarean section (800-1000 cc). Additionally, the use of anesthetic drugs during surgery can lead to hypotension.

In our patient, a caesarean section was performed due to obstetrical reasons. When caesarean delivery is necessary, general anesthesia is preferred over regional anesthesia to avoid the hypotensive effects of locally administered anesthetics.[14-16] Administering a large volume of fluids before the procedure helps mitigate this risk. Interestingly, in our patient, regional anesthesia was successfully employed.

Infective endocarditis prophylaxis is recommended and was appropriately administered in our patient. It's worth noting that approximately 15% of TOF patients have a deletion of the short arm of chromosome 22 as the genetic cause of the disease. There is a 50% probability of transmitting this condition to offspring.[17]

At delivery, the infant faces a risk of apnea, so having a senior pediatrician available for neonatal resuscitation is crucial. Overall, delicate multidisciplinary teamwork is essential for managing patients with uncorrected TOF during pregnancy. Without optimal intensive obstetrical and medical care, the prognosis for these patients remains poor, with high morbidity and mortality rates.

CONCLUSION

TOF is a significant congenital cyanotic heart disease typically diagnosed in childhood and managed surgically. While it's rare for adults with TOF to conceive, pregnancy poses unique challenges due to hemodynamic changes that strain the ailing heart. Proper counselling and education for antenatal mothers with uncorrected TOF are crucial. They should be informed about potential complications and given the option of medical termination of pregnancy when appropriate. If the pregnancy continues, a multidisciplinary approach at a tertiary care center is essential to optimize outcomes.

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