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Case Report

A case report on pregnancy with uncorrected tetralogy of Fallot with pulmonary hypertension managed uneventfully at a tertiary health care in India

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ABSTRACT

Tetralogy of Fallot (ToF) is the most frequent kind of cyanotic congenital heart disease beyond the age of one year, accounting for around 10% of all congenital heart disease cases. Natural (non-corrective) survival into the fourth decade is quite unusual (3 %). There are various physiological and heamodynamic changes that occur in pregnancy, which makes causes complications in pregnancy and delivery in already compromised women, including women with uncorrected ToF. ToF is still a major source of maternal morbidity (62%) and even fatality (10%) in ToF patients, and it has a considerable impact on foetal outcome. Discussed below a case of pregnancy in a 26-year-old woman with uncorrected ToF, was diagnosed to have pulmonary hypertension, was admitted for safe confinement. She was taken up for elective LSCS with due risk and was later shifted to ICU for further management.

Keywords: Tetralogy of Fallot, LSCS, Maternal morbidity

INTRODUCTION

ToF is the commonest congenital cyanotic heart disorder, accounting for 5-6 % of them.[1]

It is characterized by a large ventricular septal defect, right ventricular outflow tract obstruction, right ventricular hypertrophy, and aorta overriding the interventricular septum.

The majority of ToF sufferers have reconstructive surgery when they are young.

Pregnancy in uncorrected ToF entails serious risks including increased maternal morbidity, mortality up to 15%, and poor perinatal outcome.[2]

We reported a case of pregnancy in uncorrected ToF with pulmonary hypertension with scoliosis which was successfully managed at a tertiary care centre with multidisciplinary approach.

CASE REPORT

A 23 years old, primigravida with uncorrected ToF was registered for antenatal care at 28 weeks at Bharati Hospital, Pune, which was a tertiary care referral center attached to medical college. The patient was asymptomatic at presentation.

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 and developed scoliosis of the thoraco-lumbar spine. She appeared to have a stunted growth with a height of 138 cms, and weighed 32 kgs, her BMI was 22 kg/m^2. She was married since 11 months.

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: Patient details.

Characteristics	
Age of presentation	23 years
Gestational age of presentation	28 weeks
Symptoms at time of presentation	None
BMI (kg/m^2)	22
Antenatal period	Uneventful
LVEF	55%
Mode of delivery	Elective LSCS
Anesthesia	General anesthesia
Post op	ICU management
Post op complication	Sub-acute intestinal obstruction

Her echocardiography at Bharati Hospital confirmed the findings of ToF. It revealed a large outlet malignant VSD of 14 mm size, with a bidirectional shunt with overriding of the aorta. There was non dysplastic pulmonary stenosis, right ventricular hypertrophy, severe pulmonary hypertension, severe pulmonary regurgitation with a left ventricular ejection fraction of 55%.

At 37 weeks a decision to perform elective CS was taken in joint consultation of the obstetrics unit along with anesthesiologist, cardiologist, intensivist and neonatologist. She was administered antennal corticosteroids.

She underwent a caesarean section under general anesthesia. Care was taken to prevent any fall in the systolic blood pressure and avoid any drop in the systemic vascular resistance. Post operatively she was electively ventilated and shifted to ICU. She was extubated the next day and shifted to obstetrics HDU after 48 hours post-delivery. Her cardiovascular status was never compromised. She developed sub-acute intestinal obstruction on 3rd post-operative day which was managed conservatively with nasogastric tube insertion.

She was started on torsemide and was discharged on 8th post-operative day.

She will follow up with cardiologist after 4 weeks post-partum 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.

The couple were advised to refrain from further childbearing with use of barrier contraceptives and were appraised about the option of vasectomy as a method of sterilization.

DISCUSSION

ToF is characterized by a large ventricular septal defect, right ventricular outflow tract obstruction, right ventricular hypertrophy, and aorta overriding the interventricular septum.[3]

Although survival into adulthood without correction is rarely reported, but development of congenital systemic–pulmonary collaterals may enable patients with pulmonary atresia to reach adult age as in our patient.[4]

Because of the concomitant hemodynamic changes, managing these patients throughout pregnancy is difficult. During pregnancy, the right-to-left shunt is increased due to an increase in plasma volume and a concurrent decrease in both pulmonary vascular resistance and systemic arterial resistance, which increases hypoxia and cyanosis.[4]

Chronic hypoxia causes secondary polycythaemia, which causes thrombopoiesis to be suppressed.[4] Chronic hypoxia triggers secondary polycythaemia which in turn depresses thrombopoiesis. Hypoxia and polycythaemia both are risk factors for respiratory tract infection which further worsens dyspnoea.[5] Maternal complications in these women include worsening cyanosis and dyspnoea, right ventricular failure, thromboembolism, and maternal mortality.[5]

Abortions, intrauterine growth restriction, preterm deliveries, and intrauterine death are only a few examples of foetal problems. Maternal cyanosis and polycythaemia are the main threats leading to fetal growth restriction especially with oxygen saturation below 85%. Multidisciplinary approach and strict vigilance are necessary for favourable maternal and fetal outcomes.[6]

CONCLUSION

ToF is a major congenital cyanotic heart disease that is usually diagnosed in childhood and is managed surgically, getting an adult with TOF is quite rare and such patients aren't advised to conceive and in pregnancy there are a lot of hemodynamical changes which does increase the load on the ailing heart, which could be fatal to the mother, thus by proper counselling and educating antenatal mothers with uncorrected ToF regarding the complications that can happen and giving them the option of MTP at the right time these adverse outcome could be avoided and if she continues pregnancy then a multidisciplinary approach at a tertiary care center is needed.

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