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Case Report

Pregnancy with uncorrected tetralogy of Fallot: a case report

Kiran Pandey¹, Sangeeta Arya^{1*}, Deepti Sharma¹, Chandra Madhur Sharma²

¹Department of Obstetrics & Gynecology, G.S.V.M. Medical College, Kanpur, U.P., India

²Department of Obstetrics & Gynecology, Rama Medical College, Kanpur, U.P., India

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***Correspondence:**

Dr. Sangeeta Arya,

E-mail: sangeetanilverma@rediffmail.com

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ABSTRACT

Tetralogy of Fallot (TOF) is the commonest form of cyanotic congenital heart disease, with overall incidence accounting for 10% of all congenital heart disease. Natural survival into the fourth decade is extremely rare (only about 3%). After corrective surgery, the life expectancy has increased, so increasing number of women with cyanotic congenital heart disease with pregnancy are coming to hospitals, thereby imposing a great challenge to obstetricians. We have discussed below a rare case of pregnancy with uncorrected TOF in a 24 year old woman.

Keywords: Pregnancy, Uncorrected tetralogy of Fallot, Cyanotic congenital heart disease

INTRODUCTION

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease accounting for 10% of all congenital heart diseases. The defects found in patients with TOF is caused by a single developmental defect which is an abnormal anterior and cephalad displacement of the infundibular (outflow tract) portion of the interventricular septum. Four anomalies arising from this defect are (1) pulmonary stenosis, (2) right ventricular hypertrophy, (3) overriding aorta, (4) non-restrictive ventricular septum defect. To reach adulthood, most patients with TOF require surgery, either palliative or reparative. Only few patients survive up to adulthood without correction of TOF.^{1,2} Natural survival into the fourth decade is extremely rare³ (only about 3%). After corrective surgery, the life expectancy has increased, so increasing number of women with cyanotic congenital heart disease with pregnancy are coming to hospitals, thereby imposing a great challenge to obstetricians.

CASE REPORT

A 24 year old woman, G₄P₁A₂ (with no live issue) was referred from a private practitioner to emergency room of our department with chief complaints of loss of fetal

movements and dyspnoea on less than ordinary activity (NYHA grade 3) for one day following 33 weeks of pregnancy. While taking elaborate history, since childhood she had often felt fatigue and dyspnoea even during ordinary activities for which she used to squat to get relief. Patient had off and on cough which improved with over the counter drugs but she never sought proper medical consultation for these complaints. Now during this pregnancy she had felt dyspnoea on exertion (NYHA grade 2) for 15-20 days during third month, for which she didn't consult anyone as she got relief by itself. There was no orthopnea, paroxysmal nocturnal dyspnoea, cough, wheezing, or swelling over body at that time. However during the seventh month of pregnancy, she again felt dyspnoea on ordinary activity along with orthopnea and on and off cough for which she consulted her obstetrician. Getting doubt of cardiac lesion she was referred to cardiologist where she was examined and investigated and diagnosed to be a case of cyanotic congenital heart disease (Tetralogy of Fallot).

Past obstetric history: She had Preterm home vaginal delivery of dead born baby at seven months gestational age and two abortions at three and four month of gestational age which were spontaneous followed by dilatation and evacuation of uterus three and two years

back consecutively with no history of any cyanotic spells during this period. On her General examination, she was fully conscious with Pulse Rate - 90/ minute, regular, normovolaemic, Blood Pressure - 130/90 mmHg, Respiratory rate - 20/minute, bilaterally symmetric chest movement with normal vesicular breath sounds, temperature 98.4°F, Central Cyanosis with Grade 3 clubbing (parrot beak appearance) was present with no peripheral edema. S1 was normal, S2 was single and loud, without gallop, and a faint continuous murmur was heard over pulmonary area on auscultation of the heart. Systolic click was present. Per abdominal examination showed fundal height 28 weeks, longitudinal lie, breech presentation, podalic pole was free, fetal heart sound was not localised and uterus was relaxed. On per vaginal examination multiparous os was present. Investigations showed Hemoglobin 17.5 g/dL, haematocrit 60.8%, WBC 6200/cc, platelet count 80,000/cc. Arterial blood gas analysis showed pH: 7.307, pCO₂: 24.3 mmHg, pO₂: 50.6 mmHg, saturation of O₂: 70%, HCO₃⁻: 12.6 mEq/l; showing uncompensated metabolic acidosis. Coagulation profile showed BT 3 min 10 seconds, clotting time 5 min 25 seconds, PT -12.9 seconds (control-12seconds), aPTT -36 seconds(control-26.6 seconds) INR- 1.14. Other blood chemistry, urine analysis, and serum electrolytes were within normal limit. Chest X-ray showed classical boot shaped heart (enlargement of right ventricle), with clear lung fields. Electrocardiogram showed sinus rhythm, QRS rate 90 /minute, right axis deviation (RAD), normal p wave, PR interval 0.2", QRS duration 0.06". Echocardiogram showed hypertrophy of interventricular septum and right ventricle, pulmonary atresia, large VSD, R to L shunt, overriding aorta ± 50%, minimal aortic regurgitation, minimal flow with Left aortic arch. Patent ductus arteriosus was present with left to right shunt, no coarctation of the aorta and no collaterals were seen from descending aorta. Ejection fraction was 81%.

She was kept in propped up position with continuous oxygen inhalation. Injection ampicillin 1 gm 6 hrly, Injection Gentamicin 80 mg IV 12 hrly, clonazepam 0.25mg 1 tablet BD and Propranolol 20mg 1tablet BD were also continued as advised by cardiologist. Two units of FFP (fresh frozen plasma) and two units of platelets were transfused.

As Bishop's score was three, mechanical induction of labour was done by putting intracervical Foley's catheter with inflation of balloon by 30 cc normal saline. After four hours of it mild contractions of uterus began and Bishop's score improved to 5. During this period patient's pulse rate was 68 per minute and B.P. was 110/70 mm of Hg. After 10 hours, score improved to nine with 2-3 moderate contractions lasting for 20-30 seconds. After 16 hrs of induction intracervical Foley's was expelled out spontaneously followed by delivery of a dead, macerated female baby after half an hour.

After delivery her general condition was fair along with pulse rate of 82/min, regular and B.P. was 110/90mm of

Hg. Uterus was well contracted. Her postnatal period was uneventful and she was discharged on 7th postnatal day with further follow up in cardiology OPD. Risks of next pregnancy were well explained to husband and were advised to use barrier method for contraception.

DISCUSSION

The main characteristic of TOF is cyanosis. Cyanosis can result from three separate Mechanisms i.e. inadequate pulmonary blood flow, right to left shunting or intrinsic pulmonary disease. In TOF, cyanosis usually results from a right-to-left shunt at the level of ventricles and inadequate pulmonary blood flow. Elevated pressures in the RV from outflow obstruction and exposure to systemic pressure from overriding aorta lead to compensatory RV hypertrophy. Because of the outflow obstruction, blood ejected from RV crosses the VSD and enters the overriding aorta. This reduces the amount of pulmonary blood flow available for oxygenation and adds desaturated blood to the systemic circulation. Pressures in the Right Ventricle remain near to the systemic pressure.⁴

The likelihood of a favourable outcome for the mother with TOF depends upon the functional cardiac capacity of the woman and any surgical correction before conception, other associated complications that further increase the cardiac load and quality of medical care provided throughout pregnancy. Pregnant mothers with TOF are affected differently depending upon if their pathology remains uncorrected, have gone through palliative or definitive procedure or they have residual defects after surgical procedures.⁵

The principle danger for a pregnant woman with TOF is cardiac decompensation because of inability to meet the additional demands imposed by the physiological changes of pregnancy and parturition. The cardiovascular changes of pregnancy may unmask residual or recurrent TOF in patients with corrective procedures, who have been asymptomatic throughout their life after TOF repair. Women with uncorrected TOF are in a precariously balanced state. A state of hypotension (the fall in peripheral vascular resistance) as well as volume overload is poorly tolerated and specially become vulnerable to decompensation during the later months of pregnancy and postpartum when marked hemodynamic changes occur. Parturition is particularly critical time since the blood loss associated with the process may induce hypotension and eventually the right to left shunt making condition worse. Patients with TOF have an increased risk of fetal loss, and their babies are more likely to have congenital anomalies than in the general population.

Presbitero *et al.* demonstrated that the most important risk factor for adverse fetal outcome in cyanotic patients was the degree of cyanosis. These authors suggested that an arterial oxygen saturation >85% and a hemoglobin concentration <18 g/dl were more likely to result in live

birth, whereas hemoglobin concentrations >20 g/dl were associated with adverse fetal outcome. Chronic hypoxemia in such patients leads to adaptations to provide adequate tissue oxygenation i.e. polycythemia, increased blood viscosity, vasodilatation, hyperventilation and chronic respiratory alkalosis. Such adaptive mechanisms may limit cardiac reserve and Oxygen delivery during stress.

During pregnancy and labor most centres give antibiotic prophylaxis while oxytocin and ergometrine have unpredictable effects on the hemodynamic status and should be avoided in TOF patients. Close observation and continuous hemodynamic monitoring are mandatory during the delivery and upto one week postpartum. In our patient no anticoagulation was administered during the pregnancy or puerperium.

Our case suggests that pregnancy in a woman with congenital heart disease carries substantial risks for foetus and mother and favourable outcome is feasible only under appropriate medical care. But in spite of appropriate medical care these cases still remain an important cause of maternal morbidity (62.5%), and even

mortality (10%) and have significant effects on fetal outcome.

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