CASE SERIES

Pregnancy Outcome in Cases of Uncorrected Tetralogy of Fallot: Case Series with Review of Literature

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Abstract

Tetralogy of Fallot (TOF) is one of the most common cyanotic congenital heart diseases (CHD), which presents with classical tetrad ventricular septal defect (VSD) with overriding aorta, pulmonary stenosis, and right ventricular hypertrophy. Women with uncorrected TOF have maternal hypoxemia with miscarriage, poor fetal growth, preterm delivery, or even fetal death with chances of maternal heart failure as its consequence event.

We are reporting three cases of pregnancy in women having uncorrected TOF who were managed successfully at our institute. Pregnant women with class III–IV NYHA are counseled for MTP, as done in the third case here. However, if the patient refuses, we can proceed with the pregnancy with total bed rest and supportive care, as done in the first two cases. Intense monitoring is required throughout pregnancy as hypoxia and polycythemia both are related to poor maternal and fetal outcomes. Our findings in all three cases were comparable to published cases and studies like done by Veldtman et al., Balci et al., and Drenthen et al. So, patients with TOF during pregnancy need multidisciplinary team care consisting of obstetricians, cardiologists, anesthesiologists, and neonatologists among others. At the initial booking itself, these high-risk patients should be admitted for evaluation of the severity of TOF and its effect on pregnancy.

Keywords: Congenital heart disease, Fetal hypoxia, Polycythemia, Maternal and fetal outcome, Tetralogy of Fallot.

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Introduction

One of the most common causes of cyanotic congenital heart disease (CHD) in fetuses is Tetralogy of Fallot (TOF), which constitutes 10% of the total congenital heart defects. Its four components are ventricular septal defect, overriding of aorta, and right ventricular outflow tract obstruction (RVOT), which can be subvalvar, valvar, and supravalvar, and right ventricular hypertrophy. The morphology of the normal heart and TOF is predicted in Figure 1.

Without surgical repair, 25–35% die within a year after birth, 40–50% die within 4 years of age, 70% by the age of 10 years, and 95% by 40 years of age. With complete repair, >85% survive to adulthood.[1] If satisfactory surgical correction is provided prior to pregnancy, significant maternal risks can be decreased with a dramatic increase in the fetal environment. Healthy women encounter multiple physiological cardiovascular changes during pregnancy, parturition, and postpartum phase, which are particularly hazardous for uncorrected TOF patients because of the increase in right-to-left shunt leading to decreased peripheral vascular resistance and blood loss during delivery, which ultimately causes more hypoxia and cyanosis. Uncorrected TOF, residual defect after surgery, and palliative surgery, if done for TOF, is a crucial cause of maternal morbidity (62.5%) and mortality (10%).[2]

Here, we are reporting three cases of females with pregnancy having uncorrected TOF who were successfully managed in our institute under a multidisciplinary approach.

Case 1

A 22-year unbooked, primigravida, married for 7 years with spontaneous conception, presented at 35 weeks + 3 days of pregnancy with complaints of dyspnea, easy fatigability, and H/O cyanotic on heavy exertion (NYHA grade III). She was a known case of uncorrected TOF with Rh –ve pregnancy with fetal growth restriction (FGR) and was on beta-blockers (Ciplar 10 mg TDS) daily. Diagnosis of TOF was confirmed after angiography 7 years back due to repeated cyanotic spells. Surgery was postponed because of coexistent pulmonary Koch's (given full course of ATT). She had undergone phlebotomy once in 2011.

On examination, her general condition was stable, SpO_2 – 76% on room air, pulse rate – 80/min (regular), BP – 110/60 mm Hg, central cyanosis, peripheral cyanosis, and clubbing were present. The chest was clear bilaterally with pansystolic murmur over the apex on auscultation. On obstetric examination, per abdomen, she was 32 weeks at the gestation age of 35^{+5} weeks, single live cephalic fetus. Her investigations were – Hb 21 gm/dL, HCT 65%, fetal echo was normal, PT – normal, and ICT – negative.

She was managed conservatively, referred for a cardiothoracic surgeon's opinion, and was advised surgery 6 months post-partum. As she was asymptomatic, phlebotomy was deferred on

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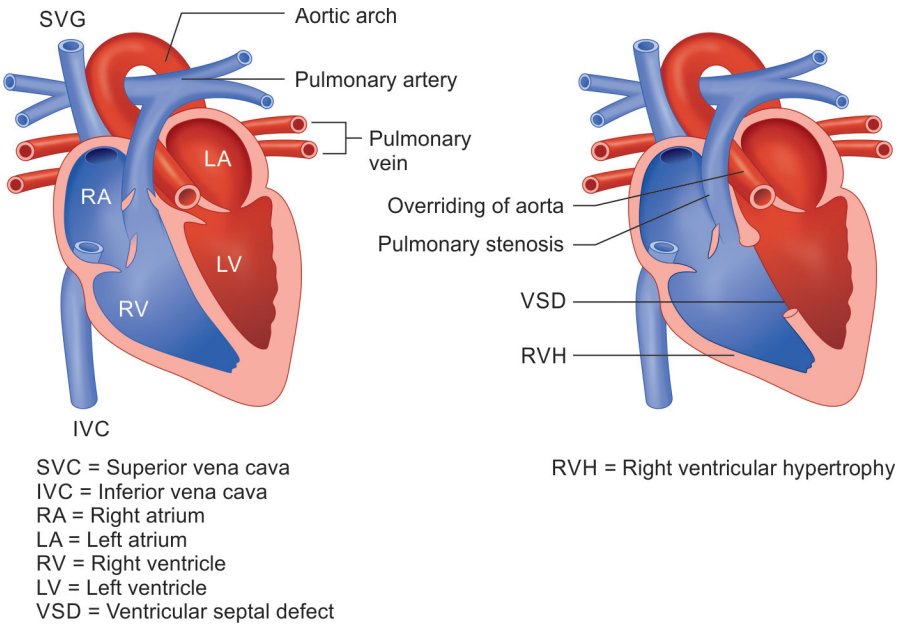

Fig. 1: Normal heart and TOF heart

cardiologist's advice despite high HCT. She was kept on intermittent oxygen therapy. Because of associated FGR, she was kept under close monitoring by modified biophysical profile and Doppler. She went into spontaneous labor, with an episode of intrapartum abruption (RPC 300 cc). She was delivered vaginally, a 1.84 kg live baby. She had mild atonic postpartum hemorrhage (PPH) (500 cc), managed medically with uterotonics. Her Hb was 19 gm%, INR 1.7, and platelet 74,000 after delivery. She was transfused 1 unit of packed cells, 4 units of fresh frozen plasma, and 4 units of platelets. Postpartum dyspnea on slight exertion was present, so she was kept on complete bed rest. Her SpO_2 was maintained at 80–85% with free-flow oxygen. The baby was kept in the nursery in view of FGR for 22 days. Both mother and baby's condition on discharge was good.

Case II

The second case was a 22-year-old primigravida with a married life of 2 years, unbooked, referred to our center as a known case of uncorrected TOF (NYHA-II). She was presented at 35 weeks, 4 days pregnancy, in our ANC OPD. Her 1, 2, and 3 trimesters were uneventful, except for one episode of a cyanotic spell. She was managed conservatively and was on tablet Ciplar (20 mg of BD). Her general condition was stable, PR – 90/min, BP – 110/70 mm Hg, cyanosis, and clubbing were present. The chest was clear. On cardiovascular examination, pansystolic murmur was present. On obstetrics examination, per abdomen, the uterus was 30 weeks, the fetus was clinically FGR, and the estimated fetal weight was 1.5 kg at 35-week gestation. Her Hb was 15.5 gm%, hematocrit – 61%, and rest of the routine investigations were within normal limits. Her USG Doppler biometry was showing 32 weeks of gestation age, asymmetrical FGR, AFI – 5 cm, and umbilical artery S: D ratio – 5.0.

Conservative management was done in consultation with a cardiologist. Fetal growth monitoring was done with serial Doppler and a modified biophysical profile. Reversal of flow in the umbilical artery was detected at 36^{+6} weeks, so, lower segment cesarean section was done under GA for fetal indication, explaining the maternal and fetal risk to the mother and close relatives. The baby's birth weight was 1.5 kg. Again, in this case, uterine atony occurred, which was managed medically (syntocinon plus carboprost) and with bimanual massage (800 mL of total blood loss). Her postoperative period was uneventful. The post-op patient was maintaining O_2 saturation (92%) on oxygen. Both baby and mother were discharged after 12 days in stable condition.

Case III

This was a young woman with uncorrected TOF and was 21 years old. She was in the second trimester of her first pregnancy and had uncorrected TOF with Eisenmenger syndrome with NYHA grade III.

She was 15 weeks pregnant and because of pulmonary hypertension, she had advised termination of pregnancy by the doctor. She came to OPD for the termination of pregnancy. Her Hb was 18, hematocrit was 67%, and the rest of the investigation was normal. She was maintaining saturation at 94% on room air. A multidisciplinary team took care of her, including a senior obstetrician, senior cardiologist, and senior intensivist along with the experienced nursing staff. After cardiology consultation, she was given tablet Misoprost in divided doses per vaginally, and under vigorous monitoring, the pregnancy was terminated. The miscarriage happened uneventfully.

Review of Literature

Presbitero et al.[3] demonstrated a degree of cyanosis as the prime risk factor for adverse fetal outcomes in cyanotic patients. Gomez et al. found that cyanotic mothers had premature termination of pregnancy, however, the birth rate and Apgar were slightly deteriorated. In the study by Drenthen W et al.,[4] 3% had heart failure and 7% had arrhythmia during pregnancy in women with TOF. Veldtman et al.[5] did a study from the Mayo Clinic in 2004 on 147 patients with TOF, which was one of the largest studies and observed an increased risk of fetal loss and congenital anomalies, as shown in Table 1.

Table 1: Results of the study by Drenthen W et al.[4]

Maternal and neonatal outcome	*Repaired TOF*	*Unrepaired TOF*
Miscarriages	27%	20%
Cesarean deliveries	23%	5%
Premature deliveries	1.2%	0
SGA	8.5%	30%
Congenital anomalies	6%	6%
Mean infant weight	3200 gm	2570 gm

Table 2: Comparative result of Drenthen et al., Balci et al., and Veldtman et al.[4,5,10]

Maternal fetal outcome in corrected TOF	*Drenthen et al.*	*Balci et al.*	*Veldtman et al.*
Cardiac maternal event (arrhythmia)	12%	8%	7%
Miscarriage	19%	19%	27%
SGA	17%	19%	8.5%
Premature birth	11%	18%	1.2%
Cesarean delivery	28%	25%	23%
Congenital defects in newborn	2.2%	3%	6%
PPH	10%	12%	

A study by Kaur et al.[6] emphasized that there was a significant increase in small-for-gestational-age (SGA) babies in the uncorrected group (40%) in comparison to the corrected group (20%). Singh et al.[7] also reported no adverse maternal cardiovascular events in 40 successful pregnancies in 27 women with corrected TOF. Similar findings were observed by Meijer et al.[8] in the study done on 5 women who had 9 pregnancies in repaired TOF. About 4% infants with SGA were reported by Siu et al.[9]

After comparing the data of three major studies in the literature as shown in Table 2, we find a high rate of cardiac complications, miscarriages, and SGA babies. Preterm births were also more common but found less in the Veldtman et al. study. Lower segment cesarean section rate was also comparable in all three studies. The risk of congenital anomalies was also high in all three. PPH was also found in two studies in a significant number and also in our first two cases.

Discussion

Only a few patients survive to adulthood if TOF remains uncorrected, and with pregnancy, it is even rarer. The tendency of becoming pregnant increases in women with cyanotic CHD in their 3rd–4th decades.[5] In all three cases, patients who conceived were in their 20s with uncorrected TOF. Their survival is a rare example of the above-mentioned natural history of TOF.

Worsening hypoxia and cyanosis occur in patients with TOF during pregnancy due to an increase in right-to-left shunt because of a fall in peripheral vascular resistance, as shown in Figure 2.

With increasing gestation, the rate of mortality and morbidity also increases in patients with uncorrected TOF. This increase is much more in those with a history of syncope, polycythemia, and right ventricular hypertrophy. During pregnancy, 40% of women can develop heart failure. Maternal mortality can reach up to 10%. Fetal mortality rates up to 36% have been reported.[11,12] There is an inverse relationship between chronic hypoxia and polycythemia, both with the outcome of the pregnancy that has been noted in various studies.

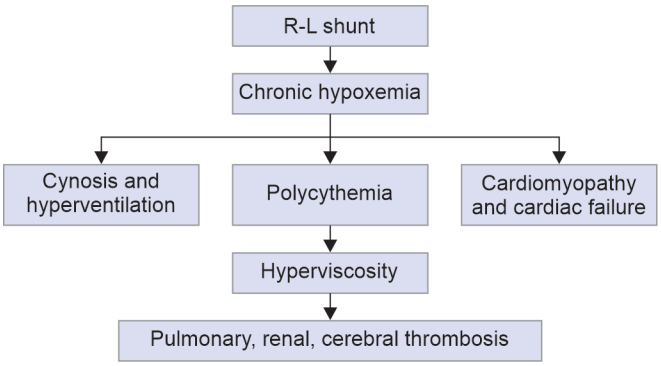

Fig. 2: Pathophysiology of TOF

Pulmonary artery abnormalities (PR) adversely affect maternal cardiac output at rest or exercise. Depressed placental blood flow subsequently leads to FGR and SGA as found in both our antenatal cases. Reversal of end-diastolic flow in the umbilical artery developed in our second case. The risk factor for the fetal outcome is the degree of cyanosis. If arterial oxygen saturation is >85%, more chances of live births are there. Hb more than 20 gm/dL and hematocrit >65% are associated with adverse fetal outcomes.[3] The incidence of miscarriage, poor fetal growth, preterm delivery, and fetal death increases due to maternal hypoxemia. In all three patients here, hematocrit was above 60%, but all were stable. The risk of heart failure increases during pregnancy and parturition as a consequence of increased volume load, leading to significant residual RVOT obstruction, valve dysfunction or ventricular dysfunction, and arrhythmias. This explains why our patient became more symptomatic during pregnancy. Fetal growth restriction encountered in both antenatal patients can be explained by chronic hypoxemia. All three patients also had polycythemia, which is a physiological mechanism to compensate for low tissue oxygen distribution. According to the CARPREG score, the risk of cardiovascular and maternal complications in our patients was 27%. According to the literature, cardiac defects in infants of TOF patients span from 3 to 17%. Definitive management is surgery only, which cannot be done in pregnancy (BLALOCK TAUSSING SHUNT). Therefore, comprehensive management is essential throughout pregnancy, labor, and postpartum.

Management starts in the preconception period itself. Women should be given adequate counseling regarding their cardiovascular risk and advised against pregnancy. Medications may be required for long-term CCF and abnormal atrial or ventricular arrhythmias. Vigilant and vigorous evaluation should be done to grade the severity of pulmonary regurgitation with consideration of pulmonary valve replacement before pregnancy in severe PR. Patients with severe PHT may require termination. In the first two cases, patients reported very late in the third trimester.

These patients require intensive monitoring and should have regular follow-up visits with the cardiologists. Additional investigations that need to be done are blood gas analysis (especially pO_2 and saturation of O_2), echo, and hematocrit. Strict surveillance of fetal growth and well-being needs to be done using fetal USG/biometry, Doppler velocimetry, and cardiotocography.

The multidisciplinary team should be involved for managing labor and delivery in such patients well in advance. Vaginal route is preferred for delivery in women with TOF with cesarean section done only in obstetric indications, as blood loss is significantly less in vaginal delivery (400–500 cc) in comparison to cesarean section (800–1000 cc). Good pain management should be done for labor and delivery to minimize maternal cardiac stress. Antibiotic prophylaxis is to be given to prevent bacterial endocarditis as we have given in our all three patients too. The second stage of labor needs to be shortened by instrumental delivery. To avoid the hypotensive effect of regionally administered anesthetic drugs, general anesthesia is preferred. Administration of loading fluids before the procedure is recommended to avoid the hypotensive effect. Our first patient delivered vaginally, whereas in the second one, cesarean section was done under GA, and the cesarean was done for obstetric indication. All the patients are managed well with the involvement of a multidisciplinary team.

Conclusion

Tetralogy of Fallot carries a substantial risk to mother and fetus, which can be improved by surgical correction preconceptionally. Detailed prepregnancy evaluation with joint supervision of a high-risk obstetrician, a cardiologist, anesthetist, and neonatologist, and a high-risk trained nurse, CTVS surgery should be done. Vaginal delivery is the preferred mode of delivery, which needs to be cut short. Hypotension and dehydration should be avoided in these patients. With the collaboration of the multidisciplinary team, the prognosis of these patients during pregnancy is excellent.

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