International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Gupta K et al. Int J Reprod Contracept Obstet Gynecol. 2014 Sep;3(3):799-802
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: 10.5455/2320-1770.ijrcog20140935

Case Report

Successful outcome of pregnancy in uncorrected tetralogy of fallot

Kashika Gupta*, Bindu Bajaj, Banashree Das

Department of Obstetrics & Gynecology, VMMC & Safdarjung Hospital, New Delhi-110029, India

Received: 24 June 2014
Accepted: 5 July 2014

***Correspondence:**
Dr. Kashika Gupta,
E-mail: drkashika@gmail.com

ABSTRACT

Tetralogy Of Fallot (TOF) is the most common cyanotic congenital heart disease. Most commonly, it is detected in the first year of life and it is rare for patients with the disease to reach adulthood without corrective surgery. Pregnancy in patients with uncorrected TOF is rare and is associated with high morbidity and mortality rates. This is due to increased maternal hypoxemia and cyanosis because of fall in systemic vascular resistance and rise in cardiac output which exacerbates the right to left shunt. Its management poses a challenge to the clinician because of the rarity of its occurrence and the paucity of literature. 22 year old woman, G_3A_2 with uncorrected TOF presented at 36 weeks gestation to the emergency room with breathlessness on routine daily activities (NYHA III) and severe fetal growth restriction. The patient had higher hemoglobin and hematocrit levels and lower platelet count. Oxygen saturation was low. Pregnancy was terminated by caesarean section under spinal anesthesia. Post caesarean patient had a febrile course and falling oxygen saturation (nadir 45.4%) which was intensively managed in consultation with cardiologist and physicians. She was discharged along with her baby in satisfactory condition on post-operative day twelve. Uncorrected TOF in pregnancy poses a therapeutic dilemma to the obstetrician, cardiologist and anesthetist. Intensive multidisciplinary management is essential to optimize the fetomaternal prognosis. With adequate care, good outcome can be achieved.

Keywords: Tetralogy of fallot, uncorrected, Pregnancy, Cyanotic congenital heart disease

INTRODUCTION

The prevalence of Tetralogy Of Fallot (TOF) is 3.4 per 10000 live births.[1] It is the most common type of cyanotic congenital heart disease.

Tetralogy of fallot has four distinct anatomic features: pulmonary stenosis; ventricular septal defect; hypertrophy of the right ventricle and rightward deviation of the aortic valve, so that it overrides the ventricular septum.

In the present era of ultrasonography, majority of these patients are diagnosed in intrauterine life, and most are identified and treated in first year of their life.[2,3] To reach adulthood, patients with TOF need surgery, either palliative or reparative. However, few patients may reach adulthood without correction of TOF. Uncorrected TOF in pregnancy is a rare entity; with few cases reported till date. Most of the cases reported, are from developing countries; possibly because of lack of awareness and resources for correction of this congenital anomaly at an early age. Patients with uncorrected TOF may deteriorate during pregnancy and parturition. These remain an important cause of maternal morbidity (62.5%), and even mortality (10%) and have significant effects on fetal outcome.[4]

Management of these cases poses a challenge for the obstetrician, cardiologist and the anesthetist partly due to paucity of literature.

CASE REPORT

A 22 year old patient was referred from a private clinic in view of uncorrected TOF in the third trimester of pregnancy. She was G_3A_2 who reported at 36 weeks 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. Her previous antenatal and pre pregnancy period was uneventful with no history of cyanotic spells, dyspnea or palpitations.

On examination; patient was plethoric, had grade 4 clubbing, bilateral pitting pedal edema and cyanosis. Respiratory rate was 20 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 30 weeks size with a single live fetus in longitudinal lie in cephalic presentation with decreased liquor (suggestive of fetal growth restriction).

Investigations revealed that the hematocrit was 52%, hemoglobin was 17 gm/dl, platelet count was 68000/mm^3. O_2 saturation was 84% on room air. Her serum electrolyte, urea and creatinine levels were normal.

Electrocardiography showed sinus rhythm, Right Atrial Enlargement (RAD), right ventricular hypertrophy with sudden transition of QRS in V_2.

Echocardiography showed a sub aortic ventricular septal defect with right to left shunting, <50% aortic override of septum. Mitral aortic continuity was maintained. Left ventricular ejection fraction was 60%. There was also right ventricular hypertrophy with dilated right atrium, with a narrow pulmonary artery and low pulmonary blood flow.

Obstetrical USG revealed a single live fetus in cephalic presentation with severe asymmetrical Fetal Growth Restriction (FGR) with umbilical artery S/D (systolic/diastolic) ratio of 6, brain sparing effect and AFI (amniotic fluid index) 9 cm. Estimated fetal weight was 1.45 kg.

In view of severe FGR with Doppler changes, the patient was taken up for elective caesarean section under spinal anesthesia. Intra operatively, liquor was meconium stained. A boy baby of 1.3 kg was delivered (APGAR score 8, 9). Inspite of all preventive measures, patient had atonic PPH (post-partum hemorrhage) and 1.1 litre blood was lost. It was controlled by uterotonics and uterine massage. Four units of platelet rich plasma and one unit whole blood was transfused. In the immediate post-operative period, she maintained O_2 saturation in the range of 80%. Patient was managed in a high dependency unit with intensive monitoring (especially of oxygen saturation and hematocrit apart from vital parameters).

Although antibiotic prophylaxis was started preoperatively and continued in the post-operative period; patient became febrile from post-operative day three. Thereafter, she had a progressively falling O_2 saturation fluctuating between 45% and 75%, and also had tachycardia (pulse rate-110/minute). The minimum O_2 saturation was 45.4% on post-operative day five (pH-7.38). The patient also had repeated cyanotic spells, and the hematocrit had fallen from 52% to 38.5%. She was started on tab propranolol (20 mg thrice daily), one unit blood transfusion was given, intravenous infusion of normal saline (100 ml/hour) was continued to increase the preload, injection morphine 2 mg was given, O_2 inhalation @ 4-6 l/min was continued throughout. Patient became afebrile within 72 hours of starting higher antibiotics (according to blood culture report), and had a progressive uphill course from post-operative day seven with rising haematocrit (to 42.3%) and O_2 saturation (76%).

The baby was kept in neonatal intensive care unit in view of low birth weight and neonatal jaundice. The baby showed no clinical evidence of heart disease.

The patient was discharged along with her baby in satisfactory condition on post-operative day twelve. Prior to discharge, patient was counselled about the need for reparative procedure of the cardiac lesion and for contraception.

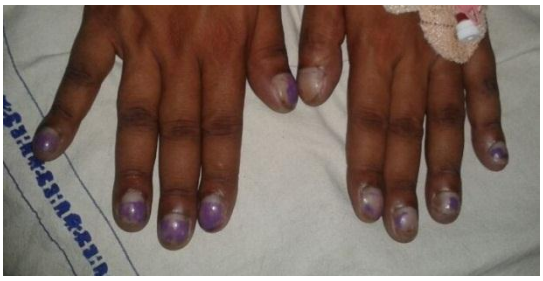

Figure 1: Clubbing of finger nails.

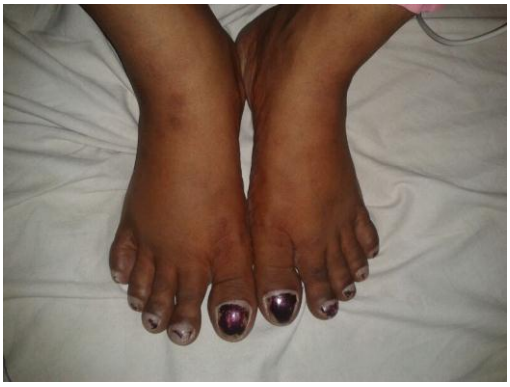

Figure 2: Clubbing of toe nails.

DISCUSSION

Uncorrected TOF in adulthood is rare, with only few patients surviving to adulthood without surgical correction. TOF with pregnancy is even rarer.

Pregnancy is associated with worsening hypoxia and cyanosis. This is explained by the increase in right to left shunt which occurs because of fall in the peripheral vascular resistance due to vasodilatation. So, there is reduction of pulmonary blood flow and increase in aortic blood flow, which further leads to reduction of arterial O_2 pressure and its saturation.[5-7] Thus, gestation increases mortality and morbidity rates of uncorrected TOF patients, especially those with history of syncope, polycythemia and right ventricular hypertrophy.[8] Risk is increased when arterial oxygen saturation levels at rest are below 85%.

This explains why our patient became more symptomatic during pregnancy. Chronic hypoxemia also explains the Fetal Growth Restriction (FGR) encountered in our patient.

Our patient also had polycythemia and thrombocytopenia. In uncorrected TOF, secondary polycythemia is result of the chronic hypoxemia which occurs due to the right to left cardiac shunt. This is a physiological mechanism for compensating the low tissue O_2 distribution. Thrombocytopenia in our patient is explained by the depression of thrombopoiesis in bone marrow, which occurs due to excessive erythropoiesis.[9] The worsening of hypoxia during pregnancy leads to further increased erythropoiesis, causing further depression of thrombopoiesis in the bone marrow. Thus, worsening of hypoxemia leads to worsening thrombocytopenia.

Uncorrected TOF is a life threatening condition for both mother and fetus. It is classified as moderate risk for pregnancy with heart disease [2A] with mortality rate reaches 5-15%.

CARPREG score is one of the most widely used risk score,[10,11] according to which the risk of cardiovascular and maternal complications in our patient was 27%.[12]

For all cyanotic congenital heart diseases, there is increment of fetal death risk (45-50%)[4,6] and increase in premature delivery (30- 50%) and FGR. With increasing maternal hypoxia, the percentage of live-born infants fall and when hypoxemia is intense enough to stimulate a rise in hematocrit above 65%, pregnancy wastage is virtually 100% .[7,13] Incidence of cardiac defects in infants of TOF patients ranges from 3 to 17%.[14]

Therefore, comprehensive management is essential.

Intensive monitoring is needed during pregnancy, labour and postpartum.

During pregnancy, the additional observations needed include blood gas analysis (especially pO_2 and saturation of O_2) and hematocrit/hemoglobin. Fetal growth and well-being needs to be monitored carefully, which is done by fetal USG/biometry, Doppler velocimetry and cardiotocography.

Vaginal birth is the preferred mode of delivery in women with TOF. Caesarean section is indicated only for obstetrical indications.[6,15-17] This is because blood loss is lesser in vaginal delivery (400-500 cc) compared with 800-1000 cc in caesarean section and use of anesthetic drugs may cause hypotension.

In our patient, caesarean was done for obstetrical indication.

If caesarean is indicated, general anesthesia is preferred to avoid hypotensive effect of regionally administered anesthetic drugs. Administration of massive loading fluids before procedure is recommended to avoid the hypotensive effect.[15-17] However, in our patient regional anesthesia was given with a successful outcome.

Infective endocarditis prophylaxis is recommended, which was given in our patient.

Approximately 15% of patients with TOF have a deletion of the short arm of chromosome 22 as the genetic cause of the disease and there is 50% probability of transmission to offspring.[18] At delivery, the infant has a risk of apnea and a senior pediatrician should be available for neonatal resuscitation.

Delicate multidisciplinary team work is essential for management of patients with uncorrected TOF and pregnancy. Without optimum intensive obstetrical and medical management, prognosis of these patients remains poor with high morbidity and mortality rates.

ACKNOWLEDGEMENTS

We would like to appreciate the medical staff of the cardiology and anesthesia department of VMMC & Safdarjung hospital, who played an invaluable role in management of this patient.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Denise van der Linde, Elizabeth E.M. Konings, Maarten A. Slager, Maarten Witsenburg, Willem A. Helbing, Johanna J.M. Takkenberg et al. Birth Prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol. 2011 Nov;58(21):2241-7.

2. Semeraro O, Scott B, Vermeersch P. Surgical correction of tetralogy of fallot in a seventy-five year old patient. Int J Cardiol. 2008;128:98-100.
3. Bailliard F, Anderson RH. Tetralogy of fallot. Orphanet J Rare Dis 2009;4:2.
4. Veldtman GR. Outcomes of pregnancy in women with tetralogy of fallot. J Am Coll Cardiol. 2004;44:174-80.
5. Sankaran VG, and Brown DW. Congenital heart disease. In: Lilly LS, eds. Pathophysiology of Heart Disease. 4th ed. Baltimore: Lippincott Williams & Wilkins; 2007: 390-393.
6. Elyakam U. Pregnancy and cardiovascular disease. In: Braunwald E, Zipes DP, Libby P, eds. Braunwald's Heart Disease: A Textbook of Cardiovascular Medicine. 6th ed. Philadelphia: W.B. Saunders; 2001: 2172-2177.
7. Cunnigham FG. Cardiovascular disease. In: Rouse D, Rainey B, Spong C, Wendel GD, eds. Williams Obstetrics. 23rd ed. New York: McGraw-Hill Companies; 2005: 963-970.
8. Presbitero P, Somerville J, Stone S et al. Pregnancy in cyanotic congenital heart disease. Outcome of mother and fetus. Circulat. 1994;89:2673-6.
9. Levine SP. Thrombocytopenia: pathophysiology and classification. In: Lee GR, et al., eds. Wintrobe's Clinical Hematology. 10th ed. Philadelphia: Lippincott Williams & Wilkins; 1999: 1579-1611.
10. Vahanian A, Baumgartner H, Bax J, Butchart E, Dion R, Filippatos G et al. Guidelines on the management of valvular heart disease: the task force on the management of valvular heart disease of the european society of cardiology. Eur Heart J. 2007;28:230-68.
11. Drenthen W, Boersma E, Balci A, Moons P, Roos-Hesselink JW, Mulder BJ et al. Predictors of pregnancy complications in women with congenital heart disease. Eur Heart J. 2010;31:2124-32.
12. Siu SC, Sermer M, Colman JM, Alvarez AN, Mercier LA, Morton BC et al. Prospective multicenter study of pregnancy outcomes in women with heart disease. Circulat. 2001;104:515-21.
13. Warnes CA. Cyanotic congenital heart disease. In: Oakley C, Warnes CA, eds. Heart Disease in Pregnancy. 2nd ed. Massachusetts: Blackwell Publishing; 2007: 43-47.
14. Uebing A, Steer PJ, Yentis SM, Gatzoulis MA. Pregnancy and congenital heart disease. Br Med J. 2006;332:401-6.
15. Oakley C. Expert consensus document on management of cardiovascular diseases during pregnancy. Eur Heart J. 2003;24:762-5.
16. Morgan GE. Maternal and fetal physiology and anesthesia. In: Morgan GE, Mikhail MS, Murray MJ, eds. Clinical Anesthesiology. 4th ed. New York: Lange Medical Books/McGraw-Hill; 2006: 876-877.
17. Morgan GE. Obstetric anesthesia. In: Morgan GE, Mikhail MS, Murray MJ, eds. Clinical Anesthesiology. 4th ed. New York: Lange Medical Books/McGraw-Hill; 2006: 902, 912.
18. Payne RM, Johnson MC, Grant JW et al. Toward a molecular understanding of congenital heart disease. Circulat. 1995;91:494-504.

DOI: 10.5455/2320-1770.ijrcog20140935
Cite this article as: Gupta K, Bajaj B, Das B. Successful outcome of pregnancy in uncorrected tetralogy of fallot. Int J Reprod Contracept Obstet Gynecol 2014;3:799-802.