

**Patient with undiagnosed tetralogy of Fallot
presenting for Obstetric Surgery**

Abstract:

We herein report a rare case of a 32 years old female presenting in the Operation Theatre for an emergency dilation and evacuation. The Case was initially postponed due to changes in ECG on the operation Table for further the cardiac evaluation. She was later confirmed as a case of Tetralogy of Fallot that was never diagnosed previously. The Surgery was conducted on the next day under General Anesthesia with Rapid Sequence Induction. The patient was later referred to a cardiologist after discharge for further management of her condition.

Keywords: Dilation and Evacuation, Right Ventricular Hypertrophy, Unrepaired Tetralogy of Fallot, Ventricular Septal Defect.

Introduction:

Tetralogy of Fallot (TOF) is most commonly found cyanotic heart defect with prevalence of 5% to 6% among all congenital heart malformations.¹ Its hallmark is overriding aorta with anterior and superior infundibular septal displacement giving rise to the tetrad of ventricular septal defect (VSD), causing infundibular obstruction and leading to right ventricular hypertrophy (RVH).^{2,3} Pulmonary stenosis or regurgitation may cause Right Ventricular dysfunction and ultimately failure, progressive tricuspid valve regurgitation, atrial and ventricular arrhythmias and in few cases sudden cardiac arrest.^{4,5}

Pregnancy though a physiological but is a quite stressful condition and many hemodynamic changes occurring in the body. Parturient with TOF have increase chances of fetal loss of up to 24%⁶ and their children are more likely to have congenital heart anomalies in comparison to general population. **Complications in a patients who usually presents with a repaired Tetralogy of Fallot in pregnancy** are associated with severe pulmonary hypertension, left ventricular dysfunction and severe pulmonic regurgitation with right ventricular dysfunction.⁴⁻⁶

Cardiac and obstetric complications are more likely to occur in patients without surgical repair of TOF^{7,8,9}. The most common cardiac complications include progressive dilatation of the right ventricle leading to ventricular failure, atrial and ventricular arrhythmias, progressive aortic root dilatation, endocarditis and thromboembolism¹⁰. The common obstetric complications include the increased risk in miscarriage, premature birth, and low birth weights, arrhythmias, congestive cardiac failure, infective endocarditis, postpartum hemorrhage, paradoxical embolism and thromboembolism⁷. Among them, the pulmonary hemorrhage, brain abscess and thromboembolism have been found to be the most common causes of death¹¹.

Parturient usually face symptoms of congestive heart failure at the time of delivery otherwise pregnancy outcomes in patients with repaired TOF are found to be good. Usually elective Caesarian-Section is preferred to prevent complications and if trial of labor occurred it usually ended up in emergency Caesarian- Section because of labor complications. In parturient undergoing labor, either required invasive arterial blood pressure monitoring, continuous telemetry, or had experienced congestive heart failure that required diuresis, had obstetric or neonatal complications or anesthetic complications and sometimes even neonatal death.¹²

Several case reports¹⁴⁻¹⁹ and limited case series²⁰⁻²⁷ have been published describing obstetric, cardiac and anesthetic concerns of pregnancy and delivery but they usually comprised of women

with palliated, incompletely repaired or completely repaired TOF therefore management of pregnant females with unrepaired TOF remains challenging. Here, we are reporting a case of undiagnosed TOF in an unbooked, multiparous female presented after 16 weeks of gestation for Dilatation and Evacuation (D&E). The cause of failure for diagnosis was surprising to say the least. Upon investigation with the patient, her previous surgeries and course of pregnancies were done in a low cost rural setups that were not up to the mark with the monitoring standards. This could be one of the cause of a failure of diagnosis.

Fig 1: ECG report

Case Summary:

We report a case of D&E with Tubal ligation in a female with undiagnosed TOF. A 32-years-old, a known case of hypertension presented in the ER with a complaint of PV Bleeding. She had gestational age of 16 weeks her pelvic Ultrasound revealed single nonviable fetus. Her previous pregnancies had been uneventful and had no reported perioperative complications. She was taking Methyldopa 250mg regularly for the past 1.5 years. There was family history of hypertension (mother of patient) while no history of any congenital heart disease. She was immediately admitted and planned for an emergency D&E and Tubal Ligation under general anesthesia.

A thorough preoperative assessment was done. Apart from her hypertensive history, she had no other known medical illness. Although, she complained of dyspnea on moderate exertion. She denied any history of chest pain, palpitations, or previous episode of Hospital admission due to cardiac disturbances. There was no Cyanosis, peripheral edema, or clubbing on examination. She was a Gravid 5, Para 4 with a history of 4 Caesarian sections. The lab investigation showed an Hemoglobin of 9.2g/dl, Platelet count of 180 with the rest of the values being unremarkable.. After taking her in the operating room, standard ASA monitoring was applied. She had a Blood

pressure of 170/100mmHg, Heart rate was 140 and her oxygen saturation on room air was 92%. Her ECG on the monitor showed significant ST depressions. A 12 Lead ECG was ordered followed by an echocardiogram and full cardiology workup before proceeding for surgery. Echocardiography revealed, Mild Right Ventricular Dilation, mal aligned VSD, 20% over riding of aorta, Left ventricular Hypertrophy (LVH) and ejection fraction (EF) of 55%, PASP of 40mmHG with PV of 3.12m/s indicating Moderate pulmonary stenosis thus diagnosed as case of irregular TOF. She was started on propranolol 10 mg twice a day and planned her D & E and laparoscopic bilateral salpingectomy under general anesthesia (GA).

A review was taken from the cardiologist, who counselled the family regarding the condition and diagnosed the patient as a variant of TOF. He further advised Inj. propranolol 10mg before starting the procedure to control the heart rate. As it was an emergent procedure, her surgery was proceeded on the next morning under after obtaining a high risk written and informed consent. She was given injection metoprolol 2mg prior to induction. Inj. Augmentin 1.2g was given prophylactically. Before induction, her heart rate was 138bpm, O₂ Saturation was 96% on room air, and Her Blood Pressure was 160/90mmHG. GA was induced with RSI with Propofol 60mg and Suxamethonium 100mg. Later, Midazolam 1mg, and Nalbuphine 6mg were given with continuous ECG monitoring and 2 pints of packed cell volume transfused intraoperatively. Maintenance was done with 2% Sevoflurane. Surgery remain uneventful with a smooth recovery. On discharge, she was asked to follow up with Cardiologist as outpatient too.

Fig 2: Provisional report of Echocardiography

Discussion:

With surgical repair of TOF, women easily survive through reproductive age but without repair they rarely reach child bearing age and conceive. As hemodynamic changes during pregnancy cause extra load on heart, causing cardiac dysfunction and may result in maternal and perinatal morbidity. The common obstetric complications seen in such cases are increased risk of miscarriage, premature birth, and low birth weight, postpartum hemorrhage, paradoxical embolism, thromboembolism, congestive cardiac failure, infective endocarditis and arrhythmias.^{7,8} Pulmonary hemorrhage, brain abscess and thromboembolism found to be the most common cause of death among them.¹¹

Thus, routine cardiac examination should be performed before pregnancy to exclude possible cardiac diseases and the cardiac surgery should be performed early if required.²⁸ As in our part of world where females usually see the gynecologist first time after conception, ECG should be included in early baseline investigations to screen for any possible undiagnosed cardiac issues that can be managed earlier. For pregnant women with TOF, close monitoring should be strengthened no matter whether they have received surgical repair or not. Decision for the mode of delivery should be individualized by weighing the risks and benefits in a given clinical situation.

Conclusion:

TOF has a high-risk of cardiac and obstetric complications for pregnant women, especially in those who did not received repair surgery. Cardiac assessment should be included in routine examination at antenatal visits so that any undiagnosed issue can be screened earlier and cases may have better prognosis under timely care of multidisciplinary professionals

Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

Consent

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

Conflict of interest:

There was no conflict of interest.

References

1. Hofman J.I : "Incidence of congenital heart disease: I. postnatal incidence". *Pediatr Cardiol* 1995; 16: 103
 2. Anderson R.H, Allwork S.P, Ho S.Y, Lenox C.C and Zuberbuhler J.R : "Surgical anatomy of tetralogy of Fallot". *J Thorac Cardiovasc Surg* 1981; 81: 887.
 3. Lillehei C.W, Varco R.L, Cohen Met al. : "The first open heart corrections of tetralogy of Fallot: A 26 to 31 year follow-up of 106 patients". *Ann Surg* 1986; 204: 490.
 4. Harrison D.A, Siu S.C, Hussain F, MacLoghlin C.J, Webb G.D and Harris L : "Sustained atrial arrhythmias in adults late after repair of tetralogy of Fallot". *Am J Cardiol* 2001; 87: 584.
 5. Gatzoulis M.A, Balaji S, Webber S.Aet al. : "Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: A multicentre study". *Lancet* 2000; 356: 975.
 6. Veldtman G R, Cannolly H M, Grogan M, Ammash N M, Warnes C A. Outcomes of pregnancy in women with tetralogy of fallot. *J Am Coll Cardiol* 2004 Jul; 44(1)174–180.
 7. Kaur H, Suri V, Aggarwal N, Chopra S, Vijayvergiya R, Talwar KK. Pregnancy in patients with tetralogy of fallot: outcome and management. *World J Pediatr Congenit Heart Surg.* 2010;1(2):170–4.
 8. Partana P, Tan JK, Tan JL, Tan LK. Multiple pregnancy in a primigravida with uncorrected Pentalogy of Fallot. *BMJ Case Rep.* 2017.
 9. Parker JA, Grange C. Anaesthetic management of a parturient with uncorrected tetralogy of Fallot undergoing caesarean section. *Int J Obstet Anesth.* 2015;24(1):88–90.
 10. Sarikouch S, Boethig D, Peters B, Kropf S, Dubowy KO, Lange P, Kuehne T, Haverich A, Beerbaum P. Poorer right ventricular systolic function and exercise capacity in women after repair of tetralogy of fallot: a sex comparison of standard deviation scores based on sex-specific reference values in healthy control subjects. *Circ Cardiovasc Imaging.* 2013;6(6):924–33.
 11. Sinto R, Nasution SA. Pregnancy in a woman with uncorrected tetralogy of fallot. *Acta Med Indones.* 2009;41(2):81–6.
 12. Arendt, Katherine W. MD^{*}; Fernandes, Susan M. MHP, PA-C[§]; Khairy, Paul MD, PhD[§]; Warnes, Carole A. MD[†]; Rose, Carl H. MD[‡]; Landzberg, Michael J. MD[§]; Craigo, Paula A. MD^{*}; Hebl, James R. MD^{*} A Case Series of the Anesthetic Management of Parturients with Surgically Repaired Tetralogy of Fallot, *Anesthesia & Analgesia*: August 2011 - Volume 113 - Issue 2 - p 307-317 doi: 10.1213/ANE.0b013e31821ad83e
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13. Busky A, Grusetz M. Pregnancy and delivery following operation for tetralogy of Fallot. *Am J Obstet Gynecol* 1955;70:1143

14. Meyer E, Tulskey A, Sigmann P, Silber E. Pregnancy in the presence of tetralogy of Fallot. Observations on two patients. *Am J Cardiol* 1964;14:874
15. Piccaro M, Rastelli D, Zecchi P, Moneta E. Tetralogy of Fallot and pregnancy. *Minerva Ginecol* 1982;34:547–57
16. Ralstin JH, Dunn M. Pregnancies after surgical correction of tetralogy of Fallot. *JAMA* 1976;235:2627–8
17. Malbranche-Aupecle MH, Mavel A, Jahier J, Kamp A, Feldman JP. Pregnancy in a patient with an isotopic pacemaker placed for surgical atrioventricular block following treatment of tetralogy of Fallot. Apropos of a case. Review of the literature. *Rev Fr Gynecol Obstet* 1985;80:105–7
18. Larsen-Disney P, Price D, Meredith I. Undiagnosed maternal Fallot tetralogy presenting in pregnancy. *ANZJOG* 1992;32:169–71
19. Vaclavinkova V, Machado L. Delivery in a multipara with unoperated Fallot's tetralogy. *Int J Gynaecol Obstet* 1994;44:165–6
20. Singh H, Bolton PJ, Oakley CM. Pregnancy after surgical correction of tetralogy of Fallot. *BMJ* 1982;285:168
21. Nissenkorn A, Friedman S, Schonfeld A, Ovadia J. Fetomaternal outcome in pregnancies after total correction of the tetralogy of Fallot. *Int Surg* 1984;69:125–8
22. Siu SC, Sermer M, Colman JM, Alvarez AN, Mercier LA, Morton BC, Kells CM, Bergin ML, Kiess MC, Marcotte F, Taylor DA, Gordon EP, Spears JC, Tam JW, Amankwah KS, Smallhorn JF, Farine D, Sorensen S. Prospective multicenter study of pregnancy outcomes in women with heart disease. *Circulation* 2001;104:515–21
23. Veldtman GR, Connolly HM, Grogan M, Ammash NM, Warnes CA. Outcomes of pregnancy in women with tetralogy of Fallot. *J Am Coll Cardiol* 2004;44:174–80
24. Khairy P, Ouyang DW, Fernandes SM, Lee-Parritz A, Economy KE, Landzberg MJ. Pregnancy outcomes in women with congenital heart disease. *Circulation* 2006;113:517–24
25. Meijer JM, Pieper PG, Drenthen W, Voors AA, Roos-Hesselink JW, van Dijk APJ, Mulder BJM, Ebels T, van Veldhuisen DJ. Pregnancy, fertility, and recurrence risk in corrected tetralogy of Fallot. *Heart* 2005;91:801–5
26. Pedersen LM, Pedersen TAL, Ravn HB, Hjortdal VE. Outcomes of pregnancy in women with tetralogy of Fallot. *Cardiol Young* 2008;18:423–9
27. Gelson E, Gatzoulis M, Steer PJ, Lupton M, Johnson M. Tetralogy of Fallot: maternal and neonatal outcomes. *BJOG* 2008;115:398–402
28. Wang, K., Xin, J., Wang, X. *et al.* Pregnancy outcomes among 31 patients with tetralogy of Fallot, a retrospective study. *BMC Pregnancy Childbirth* 19, 486 (2019). <https://doi.org/10.1186/s12884-019-2630-y>