A Pregnancy with Corrected Tetralogy of Fallot, What Should Be Done?

Riza Sufriadi¹, Mohd. Andalas¹, Novita²

¹ Obstetrics and Gynecology Department, Syiah Kuala University /Dr.Zainoel Abidin Hospital Banda Aceh, Indonesia ² Cardiology Department, Syiah Kuala University-dr. Zainoel Abidin Hospital Banda Aceh, Indonesia

Keywords: Tetralogy of Fallot, Intra Uterine Growth Restriction, Cesarean Section, Vaginal Delivery.

Abstract : The incidence of pregnancy in women with cardiovascular disease is rising. Tetralogy of Fallot (ToF) is the most common form of cyanotic congenital heart defect. Surgical results after repair of Tetralogy of Fallot have remained excellent for the last decades, the number of women with ToF reaching reproductive age following successful repair is increasing. If ToF repaired well, good cardiac function: vaginal delivery is preferred, and labor induction is safe. Cesarean delivery is limited to obstetrical indication or severe cardiac lesion. Case Report: The 32-year-old patient G5P1A3 came to the Ante Natal Care clinic of Zainoel Abidin General Hospital at 40-41 weeks' gestation. The last patient controls at the clinic at 7 months of gestation. The last ultrasound examination results postdate pregnancy, Intra Uterine Growth Restriction (IUGR), less amniotic fluid, no any sign of labor. Patient had a history of congenital heart disease known as ToF since childhood. Patient hasn't any subjective complaints about heart disease. Patient had received ToF correction in 2013 and didnot regular checkups in cardiology. Patient has done cesarean section, born female baby with birth weight 1800 grams, Apgar score 8/9.

1 INTRODUCTION

The prevalence of women with congenital heart disease increases over time. According to estimates by the European Society of Cardiology about 1600 patients with congenital heart disease in the UK enter adolescence each year, improved medical and surgical management allows more patients to reach adulthood. More advanced diagnostic tools allow for early diagnosis with better sensitivity and accuracy (Deanfield J,2003). Tetralogy of Fallot (ToF) is a congenital heart disease consisting of ventricular septal defect (VSD) subaortic membranous type, overriding aorta, infundibular pulmonary stenosis (PS) with or without PS valvular and right ventricular hypertrophy. When accompanied by ASD is called the pentalogy of fallot (Diller et al,2005). TOF events represent 10% of cases of congenital heart disease (CHD) and are the most common cause of cyanotic CHD (Rauch R,2010). The mortality rate in patients who did not undergo surgery increased gradually, ranging from 30% at age 2 years to 50% at 6 years of age. The highest mortality rate in the first year and then remain constant until the second decade. No more than 20% of TOF patients can live up to age 10 and <5-10% of patients live at the end of the second decade. Most people who survive until age 30

have congestive heart failure (CHF) (Pillutla P,2009). It is the largest left-to-left shunt, due to obstruction of blood flow to the lungs where pulmonary vascular resistance is normal. If this disorder is not corrected, a pregnancy can be achieved but maternal mortality is still high, and the fetal loss rate is over 50 percent. After surgery correction of total defects, maternal mortality is not clear beyond that of women without heart disease; the chance of offspring to get heart disease is about 5-10 percent (Anwar.B, 2004). In a study by Veldtman (2004) founded infants were small for their gestational age, 86% being born to women with unrepaired TOF. The fetus was at highest risk when the mother had a combination of obstetric risk factors and cardiac risk factors. We were able to demonstrate a negative correlation between infant birth weight and maternal unrepaired TOF status, morphologic PA abnormality, higher RV systolic pressure, and younger age at primary surgical repair. Unrepaired TOF and the presence of morphologic PA abnormality (hypoplastic or disconnected PA or ductal origin of PA) were independently associated with infant birth weight. Among infants with low birth weight, 50% of their mothers have chronic disease that unfavourably affects maternal placental blood flow. One can speculate that PA abnormalities, particularly in the presence of pulmonic

Sufriadi, R., Andalas, M. and Novita,

A Pregnancy with Corrected Tetralogy of Fallot, What Should Be Done?

DOI: 10.5220/0008792102190222

In Proceedings of the 2nd Syiah Kuala International Conference on Medicine and Health Sciences (SKIC-MHS 2018), pages 219-222 ISBN: 978-989-758-438-1

Copyright © 2020 by SCITEPRESS - Science and Technology Publications, Lda. All rights reserved

regurgitation, may adversely affect augmentation of maternal cardiac output at rest or during exercise. This may result in depressed placental blood flow and, subsequently, intrauterine growth retardation (Veldtman GR, 2004.). About 10% of women with a history of ToF correction have short-term cardiovascular complications during pregnancy. Supraventricular and ventricular arrhythmias are well-known long-term sequelae of intracardiac repair of ToF. The use of cardiac medication before pregnancy strongly predicted cardiovascular events. The incidence of SGA is higher than in the general population. The premature birth rate also was higher. However, the long-term effects of pregnancy are poorly researched Balci (2011) about 10% of women with a history of ToF correction have short-term cardiovascular complications during pregnancy. Supraventricular and ventricular arrhythmias are well-known long-term sequelae of intracardiac repair of ToF. The use of cardiac medication before pregnancy strongly predicted cardiovascular events. The incidence of SGA is higher than in the general population. The premature birth rate also was higher. However, the long-term effects of pregnancy are poorly researched (John Lynn Jefferies, 2016).

2 CASE REPORT

The 32-year-old pregnant women with G5P1A3 came to the Antenatal Care clinic of Zainoel Abidin Hospital at 40-41 weeks' gestation. The last patient controls at the clinic at 7 months of gestation, last ultrasound examination results in polyclinic were said postdate pregnancy, Intra Uterine Growth Restriction (IUGR), less amniotic fluid. The patient does not sign of labor or Premature rupture of membranes. Fetal movement is felt active. The history of flour albous is recognized, but not itchy and odorless. Fever during pregnancy is refused. Micturition and defecation within normal limits. Patients had a history of congenital heart disease known as ToF. Patients had received TOF correction in 2013. Menstrual history within normal limits. Patients had abortion 3 times (1, 2 and 3 pregnancies), 2-year-old child with BBL 2,500 grams, born with Cesarean section due to transverse lie presentation.

Examination of vital signs within normal limits with normal nutritional status. Blood pressure was 120/80, pulls 80 beat/ min and no murmur or gallop. Laboratory examination within normal limits. The Echocardiography results: mild atrial regurgitation, ejection fraction 45%, septal dyskinetic: corresponded to Chronic Heart Failure, in general good clinical condition, and tolerance of cesarean section with moderate risk. Obstetric findings: no dilatation of cervix with head presentation, uterus was not contractions, fundal height 25 cm. Ultrasound examination obtained a single live fetus head percentage with gestational age 40-41 weeks, estimated fetal weight 1813 gram, intra uterine growth restriction accompanied by oligohydramnios. Cardiotocography finding was

reassuring with baseline 145 beat/minute, good variability and no decelerations. Patient has done cesarean section, born female baby with birth weight 1800 gr, Apgar score 8/9.

3 DISCUSSIONS

The prevalence of women with congenital heart disease increases over time. According to estimates by the European Society of Cardiology about 1600 patients with congenital heart disease in the UK enter adolescence each year, improved medical and surgical management allows more patients to reach adulthood. More advanced diagnostic tools allow for early diagnosis with better sensitivity and accuracy (Deanfield J,2003).

Nowadays, over than 60th year since the first successful intracardiac repair of tetralogy of Fallot (ToF). The repair largely consists of ventricular septal defect closure and the relief of variable forms of right ventricular (RV) obstruction, and usually resulting in free pulmonary insufficiency (PI). Early in the experience with TOF repair, attention was on quantity of life (palliative). Palliative shunts were widely used to permit repair at a safer, older age. The dividend from a full relief of obstruction included excellent function for decades for patients formerly suffering from morbid or lethal disease (Bichell, 2014). This case report explained about the women 32 years old had a history of congenital heart disease known as ToF. Patients had received ToF correction in 2013. Echocardiography results: mild atrial regurgitation, ejection fraction 45%, septal dyskinetic, corresponded to Chronic Heart Failure, in general good clinical condition, and tolerance of cesarean section with moderate risk. This patient has been successful repaired of tetralogy of fallot and good functional status was classified in a NYHA classes I and II. Improved survival of patients with congenital heart disease now means that there are more adults than children with repaired congenital disease in developed countries. Medium-term survival of repaired tetralogy of Fallot (rToF) has been documented to be good, with 20-year survival rates at or above 90% (Dennis M, 2017).

Women with corrected ToF have risk of cardiovascular and obstetric events during pregnancy. Most events are well treatable. The incidence of offspring events is markedly increased. Cardiovascular and offspring outcomes are strongly related with the use of cardiac medication before pregnancy. Surgical status before pregnancy also appears to predict pregnancy outcome. This patient had successful repaired of tetralogy of fallot and good functional status was classified in as women in NYHA classes I and II. Improved survival of patients with congenital heart disease now means that there are more adults than children with repaired congenital disease in developed countries. Medium-term survival of repaired tetralogy of Fallot (rToF) has been documented to be good, with 20-year survival rates at or above 90% (Dennis M, 2017).

The 32-year-old patient G5P1A3 came to the clinic at 40-41 weeks' gestation. The last patient controls at the clinic at 7 months of gestation. Patient with bad history of antenatal care, has been abortion 3 times (1, 2 and 3 pregnancies). Higher prevalence of miscarriage relevant with research by Balci at al: from 157 pregnancies in 74 of these patients with post corrected ToF, 19% ended in a miscarriage and 2,5% in an elective abortion (Balci,2011). Its similar correlation with research by Pedersen at al; that outcomes of pregnancy, and fertility, in a series of women who underwent surgery for tetralogy of fallot, prevalence of spontaneous abortion is 15% (Pedersen LM, 2008 Aug;18).

During normal pregnancy and delivery, there are dramatic alterations in cardiovascular physiology. Systemic vascular resistance falls, blood volume increases, cardiac output increases secondary to increased heart rate and stroke volume, and a physiological left ventricular hypertrophy occurs. Pregnancy in unrepaired TOF carries a major risk of maternal complications, including heart failure, arrhythmia and endocarditis, which can give rise to fetal problems including miscarriage and preterm labour. The risk is particularly high when the average systemic oxygen saturation falls below 85%. In repaired tetralogy of Fallot (rTOF), the risk of pregnancy is dependent on the degree of residual haemodynamic impairment. When a good repair has been achieved, pregnancy is usually well tolerated in the absence of pregnancy complications such as pre-eclampsia. However, in women with residual shunts, right ventricular outflow obstruction and/or right ventricular dysfunction, the increased overload volume of pregnancy can lead to heart failure and arrhythmias. (Veldtman GR, 2004.). We not founded adverse of cardiac events in this patient such as arrythmia.

Ultrasound examination obtained a single live fetus head percentage with gestational age 40-41 weeks, estimated fetal weight 1813 gram, intrauterine growth oligohydramnios. restriction accompanied by Cardiotocography finding was baseline 145, good variability and no decelerations. In pregnancies with cardiovascular events, significantly smaller for gestational age children were born. In this case, ultrasound examination results Intra Uterine Growth Restriction, low infant birth weight was related to the maternal state of women who had not undergone reparative surgery or to morphologic pulmonary artery abnormalities. In a research infant who were small for gestational age, 71% were born to women with untreated TOF (Child JS, 2004). The incidence of SGA (19%) is also higher than in the general population although it is lower than the 35% recently mentioned by Gelson et al. The use of maternal cardiac medication before pregnancy was the most important predictor of offspring outcome. Maternal hemodynamic abnormalities as well as direct effects of maternal cardiovascular medication may undermine placental blood flow and induce placental insufficiency with subsequent intrauterine growth restriction resulting in children born SGA as well as in premature birth. The strong association between maternal cardiovascular events and SGA points in this direction. Palliative surgery before correction

appears to influence offspring outcome negatively. Some neonatological outcomes were high mortality percentage, partially due to prematurely born babies. It was also notices that new-borns were born with low body weight for their age which was closely related to frequency of negative cardiovascular outcome during pregnancy which can lead to hemodynamic changes and placenta insufficiency as a result (Balci,2011).

Relief from pain and apprehension is important. Although intravenous analgesics provide satisfactory pain relief for some women, continuous epidural analgesia is recommended for most the major problem with conduction analgesia is maternal hypotension is especially dangerous in women with intracardiac shunts in whom ow may be reversed. Blood passes from right to left within the heart or aorta and thereby bypasses the lungs. Hypotension can also be life-threatening if there is pulmonary arterial hypertension or aortic stenosis because ventricular output is dependent on adequate preload. In women with these conditions, narcotic conduction analgesia or general anesthesia may be preferable. (Cunningham, 2018)

For vaginal delivery in women with only mild cardiovascular compromise, epidural analgesia given with intravenous sedation often succes. is has been shown to minimize intrapartum cardiac output uctuations and allows forceps or vacuum-assisted delivery. Subarachnoid blockade is not generally recommended in women with significant heart disease. For cesarean delivery, epidural analgesia is preferred by most clinicians with caveats for its use with pulmonary arterial hypertension. Finally, general endotracheal anaesthesia with thiopental, succinylcholine, nitrous oxide, and at least 30-percent oxygen has also proved satisfactory (Cunningham, 2018).

Heart rate, stroke volume, cardiac output, and mean arterial pressure increase further during labor and in the immediate postpartum period and should be monitored closely. Fluid intake and output and pulse oximetry readings should also be carefully reviewed. Lateral positioning and adequate pain control can reduce maternal tachycardia and increase cardiac output. There is no consensus on intrapartum invasive hemodynamic monitoring, but women with New York Heart Association class III or IV disease may be candidates. Operative assistance with the second stage of labor is recommended to decrease maternal cardiac work. The immediate postpartum period is especially critical for the patient with cardiovascular disease. Blood loss must be minimized, and blood pressure maintained, but congestive failure from fluid overload must also be avoided (John, T Queeman, Catherine Y.Spong, Charles J.Lockwood, 2015).

Based on Simpson review (2012) recommends cesarean delivery for women with the following: (1) dilated aortic root >4 cm or aortic aneurysm; (2) acute severe congestive heart failure; (3) recent myocardial infarction; (4) severe symptomatic aortic stenosis; (5) warfarin administration within 2 weeks of delivery; and (6) need for emergency valve replacement immediately after delivery. Although we agree with most of these, we have some caveats (Cunningham, 2018). The indication of Caesarean section in this patient by Obstetric indication.

4 CONCLUSSIONS

Vaginal delivery is preferred, and labor induction is usually safe with collaborating care by multiple specialist. Caesarean delivery is limited to obstetrical indications, and considerations are given for the specific cardiac lesion or inavailability of teamwork and general support facilities.

REFERENCES

- Agarwal N, Gupta M, Singh N, et al. Successful Management of Pregnancy in Uncorrected Tetralogy of Fallot with Pulmonary Atresia. Journal of Obstetrics and Gynaecology of India. 2015;65(6):417– 9, n.d.
- Al-Aqeedi RF, Alnabti A, Al-Ani F, et al. Successful Delivery by a Cesarean Section in a Parturient with Severe Dilated Cardiomyopathy an Implantable Cardioverter Defibrillator and a Repaired Tetralogy of Fallot. Heart Views. 2011 Jan-Mar;12(1):26–31, n.d.
- Anwar, B. Kehamilan dan Penyakit Jantung. Universitas Sumatera Utara. 2004, n.d.
- Balci A, Drenthen W, Mulder BJ, et al. Pregnancy in women with corrected tetralogy of Fallot: occurence and predictors of adverse events. Am Heart J. 2011 Feb;161(2):307–13, n.d.
- Balci, A. e. a., 2011. Pregnancy in women with corrected tetralogy of Fallot: Occurrence and predictors of adverse events. *American Heart Journal*, *Volume*, *Issue*, 161(2), pp. 307 - 313.
- Bichell, D. P., 2014. Fourth Decade After Repair of Tetralogy of Fallot. *ahajournals.org*, 130(Circulation), pp. 1931-1932.
- Cassater D, et al. The effects of pregnancy on right *ventricular* remodeling in women with repaired tetralogy of Fallot. Int J Cardiol 2013;168:1847–1852, n.d.
- Cauldwell, M. et al. Effect of Pregnancy on Ventricular Aortic Dimensions in Repaired TOF. JAHA. 2017 10(16):1-16, n.d.
- Child and JS, a. a., 2004. Fallot's Tetralogy and Pregnancy;Prognostication and Prophesy. *Journal of the American College of Cardiology*, 44,(1), pp. 181-3.
- Chiu SN, Wu MH, Su MJ, et al. Coexisting mutations/polymorphisms of the long QT syndrome genes in patients with repaired Tetralogy of Fallot are associated with the risks of life-threatening events. Hum Genet. 2012 Aug. 131(8):1295-304, n.d.
- Cunningham, F. G., 2018. *Williams obstetrics*. New York: McGraw-Hill.
- Deanfield J, Thaulow E, Warnes C, Webb G, Kolbel F, Hoffman A, et al. Management of grownup congenital heart disease. Eur Heart J. 2003; 24:1035–84, n.d.
- Dennis M, M. B. K. I. e. a., 2017. Adults with repaired tetralogy: low mortality but high morbidity up to middle age. Open Heart, 4:(000564), p. e000564..

- Diller GP, Kempny A, Liodakis E, et al. Left ventricular longitudinal function predicts life-threatening ventricular arrhythmia and death in adults with repaired tetralogy of fallot. Circulation. 2012 May 22. 125(20):2440-6, n.d.
- John Lynn Jefferies, 2016. Mode of Delivery and Pregnancy Outcome in Women with Congenital Heart Disease. *PLoS ONE*, p. 11(12).
- John, T Queeman, Catherine Y.Spong, Charles J.Lockwood, 2015. Protocol for high risk Pregnancies: evidence based approach, sixth edition. oxford UK: Wiley Blackwell.
- Meah VL, Cockcroft JR, Backx K, Shave R, Stohr EJ. Cardiac output and related haemodynamics during pregnancy: a series of meta-analyses. Heart. 2016;102:518–526, n.d.
- Meijer JM, P. P. D. W. e. a., 2005. Pregnancy, fertility, and recurrence risk in corrected tetralogy of Fallot. *heart*, 91(6), pp. 801-805.
- Pedersen LM, 2008 Aug;18. Outcomes of pregnancy in women with tetralogy of Fallot. *Cardiol Young.*, 4(Epub 2008 Jun 18), pp. 423-9.
- Pillutla P, Shetty KD, Foster E. Mortality associated with adult congenital heart disease: Trends in the US population from 1979 to 2005. Am Heart J. 2009 Nov. 158(5):874-9, n.d.
- Rauch R, Hofbeck M, Zweier C, et al. Comprehensive genotype-phenotype analysis in 230 patients with tetralogy of Fallot. J Med Genet. 2010 May. 47(5):321-31, n.d.
- Sudoyo AW, Setiyohadi B, Alwi I, Simadibrata M, Setiati S (Ed). Buku Ajar Ilmu Penyakit Dalam. Ed. 6, Jakarta : InternaPublishing, 2015, n.d.
- Veldtman GR, 2004.. Outcomes of pregnancy in women with tetralogy of Fallot. *J Am Coll Cardio*, Volume 44, p. 174–80.