CCJ
Cardiovascular Cardiometabolic Journal (2020) 1, 18-25
https://e-journal.unair.ac.id/CCJ

Case Report

Management of Pregnancy in 31-Year-Old Woman with Persistent Atrial Flutter After Cone Reconstruction for Ebstein's Anomaly

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ARTICLE INFO

Article history:
Submitted 31st January 2020
Reviewed 18th February 2020
Accepted 16th March 2020
Available online March 2020

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Keywords:
Atrial Flutter
Ebstein Anomaly
Cone reconstruction

ABSTRACT

There is a growing number of mothers suffering innate heart disease which could potentially risk their gestation period. Ebstein's anomaly refers to a rare innate disease in the heart which accounts for less than 1% of most cases of innate heart disease, and only 5% of the patients surviving beyond 50 years of age. Cone reconstruction (CR) is an option for the repairment of tricuspid valve in patients with Ebstein anomaly. However, persistent arrhytmia, such as AFL, is possible to occur after CR. This case report aims to opt suitable pregnancy management of patients with persistent atrial flutter (AFL) after Cone reconstruction for Ebstein's Anomaly. We report a 31 year-old woman who consulted for pregnancy planning due to history of Ebstein's Anomaly, and had undergone successful cone reconstruction. No recorded history of AFL is reported. Physical examination finding was tricuspid regurgitation murmur without signs and symptoms of right ventricular failure. Electrocardiography showed counter-clockwise typical AFL. Echocardiography finding demonstrated post Cone reconstruction (CR), mild tricuspid regurgitation, and EF of 76%.

Introduction

The statistical data of mothers suffering from innate heart disease during their gestation period is rising, because more than 90% grew with it when they reached adulthood. Most common problems during gestation and delivery are bleeding, arrhythmias, failure of the heart, and infrequently maternal death [6]. Ebstein's anomaly refers to a rare innate disease in the heart which accounts for less than 1% of most cases of innate heart disease, and only 5% of the patients surviving beyond 50 years of age [7,8]. It is characterized by the attachment of septal and posterior leaflet to the myocardium underlying it with the dislocation of the functional tricuspid annulus downward, resulting in the dilation of atrialized portion of right ventricle (RV) and true tricuspid annulus [9]. Several methods have been tried for the surgical repairment of the patients suffering the anomaly Ebstein.

The managing preparations mainly focused on the repairment of the monocusp of the tricuspid valve with discerning plication of the atrialized right ventricle. Cone reconstruction was an option to repair tricuspid valve in Ebstein anomaly patients. A leaflet tissue of 360 degrees and made to surround the real annulus is the result of this strategy [10]. Actually, after conducting CR, the risk of arrhythmia for young patients with EA is very small [11]. However, despite the low risk arryhthmia, a persistent arrhytmia, such as AFL could still occur after CR. Nevertheless, both AFL and pregnancy could influence to thromboembolic problems [7]. Pregnancy, especially after first stability, raised the risk of thromboembolism up to 1.5 times [2]. It is presumed that AFL patients have a similar risk of thromboembolism to those with atrial fibrillation [3]. This case report focuses on elaborating suitable pregnancy management of persistent atrial flutter patients after Cone reconstruction for Ebstein's Anomaly.

Case Presentation

31 years old woman consulted for pregnancy planning due to the history of Ebstein's anomaly post her CR. The patient was asymptomatic and was clinically stable with blood pressure of 100/70mmHg, heart rate of 60 bpm, respiratory rate of 18 tpm, and BMI of 25.11. Physical examination findings were TR murmur, without signs and symptoms RV failure. Electrocardiography showed counterclockwise typical atrial flutter. Electrocardiography showed counter-clockwise typical AFL (Figure 1). Chest X-Ray demonstrated normal aortic and pulmonal segent, cardiac waist (+), downward cardiac apex, cardiomegaly with CTR 69% (Figure 2). Echocardiography finding showed post Cone reconstruction, mild tricuspid regurgitation, and EF of 76%. Her laboratory tests showed no remarkable finding. Her routine medications were bisoprolol 1x 2.5 and Aspirin 1x80mg.

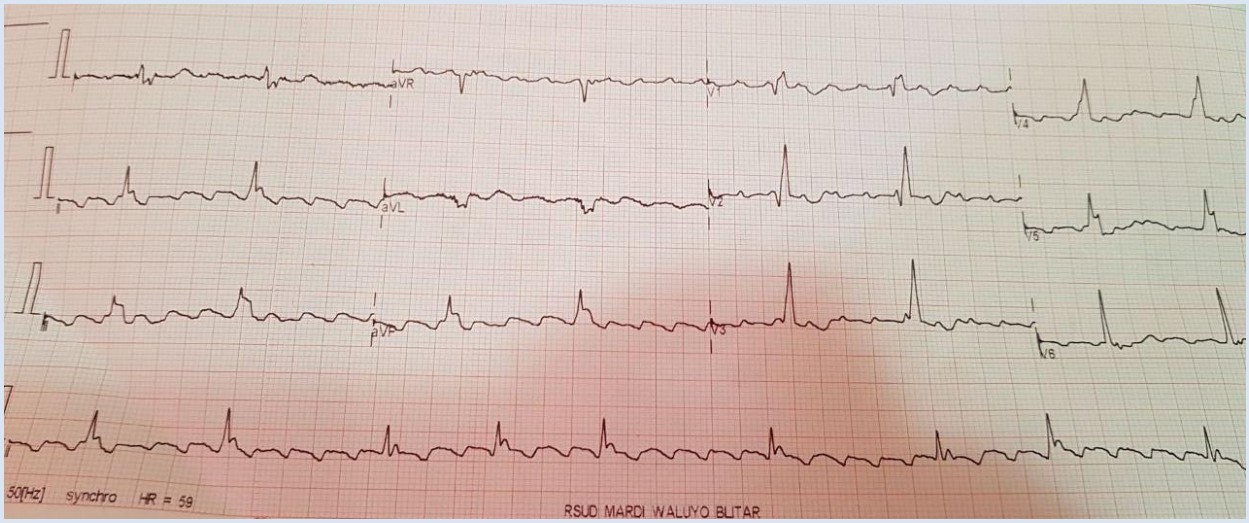

Figure 1. AFL type 1 patterns on ECG

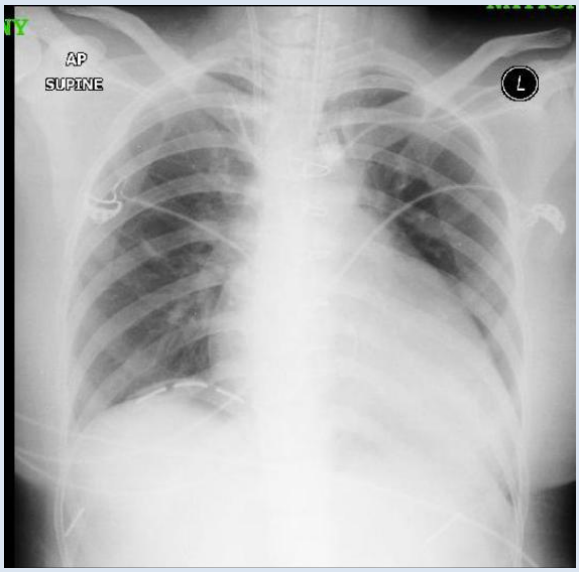

Figure 2. Chest X-Ray post Cone Reconstruction

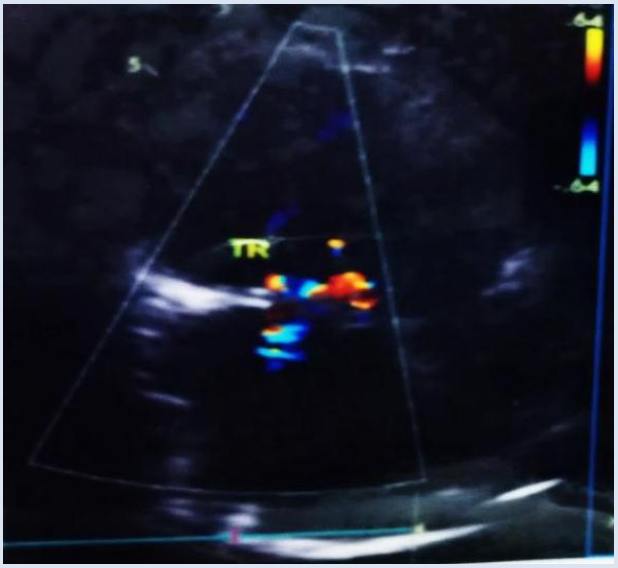

Figure 3. Echocargiography post CR

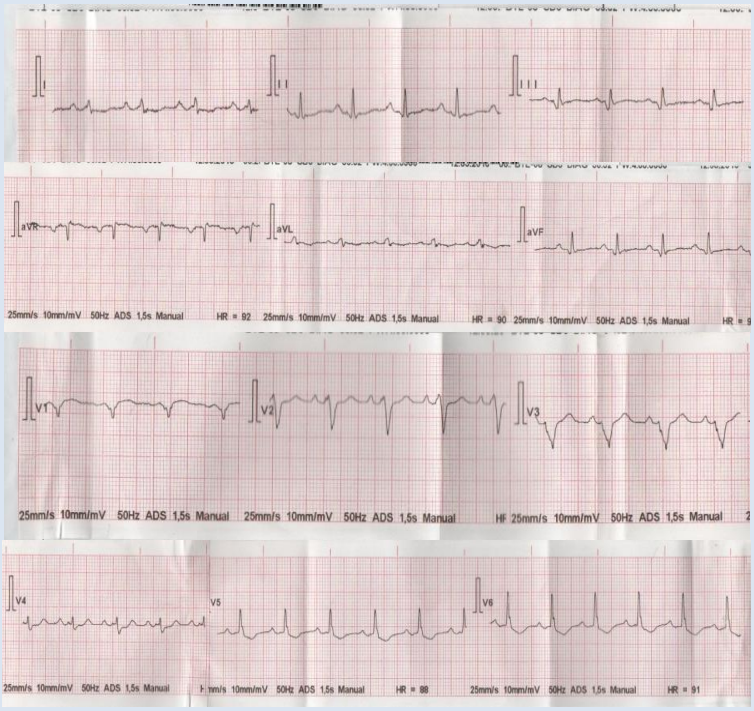

Figure 4. Sinus Rhythm Pattern on ECG before Cone Reconstruction

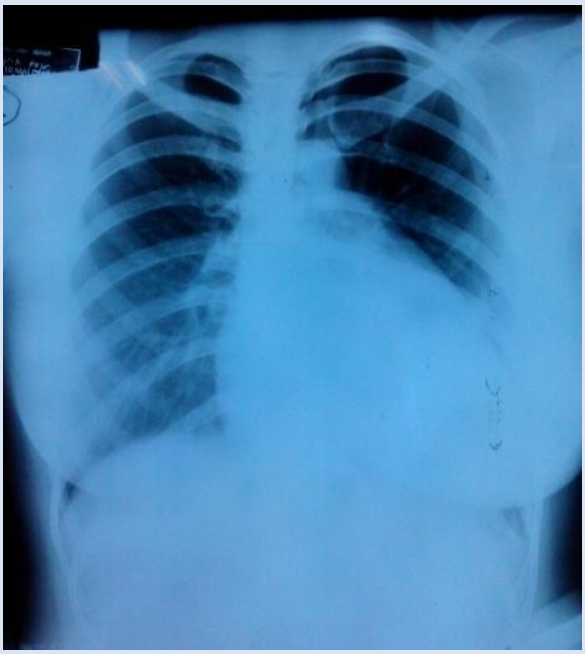

Figure 5. Chest X-Ray before Cone Reconstruction

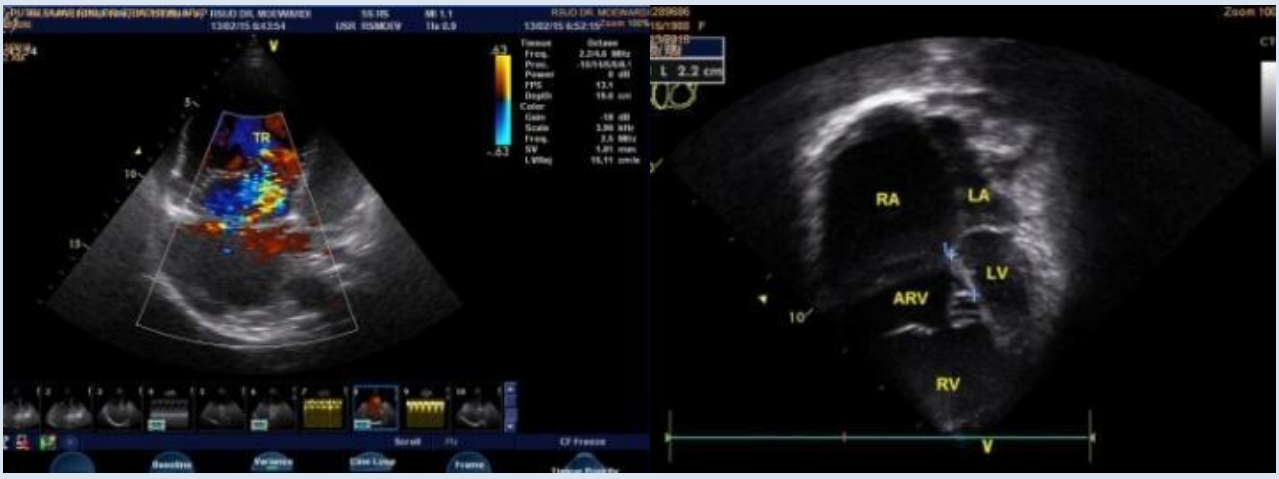

Figure 6. Echocardiography of Ebstein's Anomaly before CR

Past medical history showed that patient was diagnosed with ebstein's anomaly without other concomitant congenital heart disease, no recorded history of atrial flutter. Her ECG before CR was sinus rhythm with incomplete LBBB (Figure 4). Her previous chest X-ray showed cardiomegaly with

rounded cardiac apex, with CTR of 56%, and the absent of cardiac waist (Figure 5). Patient undergone successful cone procedure with mild residual TR. Echo finding demonstrated the typical features of Ebstein anomaly. Severe TR was observed with displaced insertion of posterior leaflet far toward RV apex, without coaptation. It showed RA and RV dilatation, D shaped LV, and normal LV with EF of 64% (Figure 6).

Discussion

Women with congenital heart disease require careful monitoring before, during, and after pregnancy. In pregnancy, hemodynamic and hormonal alteration could disturb the arrhythmias and the prothrombic state that contribute to thromboembolic problems [1]. Haemodynamic changes in the course of pregnancy include plasma volume raise, heart rate, and decrease of aortic pressure and systemic vascular resistance. Plasma volume increase in the peak of 32 weeks of gestation, it counts higher by 40 to 50 percent than the pre-gestation period. Rate of heart also reaches its peak at 32 weeks of pregnancy; almost higher by 20% than in the pre-gestation period [12]. Variability of heart rate was oppressed during pregnancy, this impaired autonomic nerve activity and the overload volume of the heart and operational wound could count for the presence of tachyarrhythmia during pregnancy [14]. Pulmonary artery pressure stays the same during pregnancy, because the increased volume of pulmonary blood will decrease pulmonary vascular resistance. In the first trimester, there would be rapid rise of arterial pressure; louder pulse of the jugular venous with heartbeat that splits secondly. In the late pregnancy, hypotension could be encountered because of the inferior vena cava compression by the enlarged uterus.

Hypercoagulable state also happened in pregnancy. Factors of anticoagulant such as coagulation factor I, V, VII, VIII, X and XII; von Willebrand factor; and plasma fibrinogen are activated. Fibrinolytic inhibitors are also activated, such as plasminogen activator inhibitor (PAI-1 and PAI-2) [6]. Because of this hypercoagulability, patients with arryhthmia require thourough care. AFL increased the risk of thromboembolism and may make the heart function worse [1,13]. Presumably, AFL patients have a similar risk of thromboembolism to those with atrial fibrillation [13], while risk of ischemic stroke only significantly increased at CHA2DS2-VASc score of >4,[5,15] or in CHADS2 score of more than 2 [15]. Thrombophylaxis is needed for high risk pregnant woman, but careful consideration on the medication choice should be made. Oral anticoagulant use during pregnancy could increase the risk of bleeding in the time of delivery, also fetal deaths which could be resulted from sudden miscarriage. Heparin was the preferred option since it does not cross the placenta and relatively safe [1]. Vitamin K antagonists are still tolerable for most cases (without history of congenital heart disease) from second trimester untill 1month before expected delivery [13].

Vitamin K antagonists, that are associated with warfarin embryopathy, cross the placenta in the first trimester state, the increasing risk of CNS abnormalities, fetal and neonatal bleeding near the delivery time, and losing gestation,. LMWH is recommended rather than fondaparinux in the time of gestation. There was no LMWH or UFH that was detected in the breast milk in discernible quantity or crosses the placenta [2]. Hypodermic LMWH in a maintained dose could be used in the course of the gestation's first trimester and the last month. Dabigatran which was said to be one of the new antagonists of oral thrombin, should not be used because a high dose of fetotoxicity was detected. High risk patients with atrial fibrillation seemed to be

lest effective to both single or dual antiplatelet therapy (clopidogrel and acetylsalicylic acid) than warfarin [15].

Beside the increasing risk of thromboembolic in pregnant woman and especially with overt AFL, AFL itself during pregnancy results in some serious undesirable events. Studies showed that there is a raise in maternal mortality and low birth weight during pregnancy in AFL patients [1,4]. The increase of maternal mortality found in 12% of pregnancies complicated with AFL. Thus, it is proven that the association between serious results in pregnant women suffering structural heart disease and atrial arrhythmias.[1] Lapses of suffered tachycardia, in particular the atrial flutter, are intolerable and could inflict fetal hypoperfusion in structural heart disease [15].

Ebstein anomaly tends to be related to tricuspid ejection, a RV that functions abnormally and a dilated right atrium. Patients may also have multiple accessory pathways. When there are RV size and systolic function that are reasonable, pregnancy is tolerated, as it is related to the absence of significant cyanosis or arrhythmias [14]. As the majority antiarrhythmic medication could be regarded as harmful to the fetus, starting AFL medical treatment shows complicated choice in its practice [2]. Beta-blockers cross the placenta and are associated with intra-uterine growth retardation, neonatal respiratory depression, bradycardia, and hypoglycemia, particularly for the exposure in 12-24 weeks of gestation [13].

Calcium channel blocker may also be administered to treat atrial arrhythmias, however, since oral Verapamil is an FDA class C, intrevenous Verapamil is more likely to be avoided due to the hypotensive effect. Diltiazem was associated with teratogenicity. Digoxin is an FDA class C medication, while Amiodarone is opposed because it influenced the function of fetal thyroid in the period of gestation [1]. The hemodynamic changes in gestation time influence the pharmacokinetics of the antiarrhythmic drugs, for example, a higher dose of digoxin is required for the adequate serum level during pregnancy, compared for those used out of pregnancy [4]. Despite the weakness of each antiarrhythmic drugs, their use should be considered if weigh the benefit. The guideline for the management of cardiovascular diseases in gestation time from ESC in 2011 suggested Beta-blocker for the first choice of rate control in arrhythmia during pregnancy. Strenuous exercise is also recommended because it is more effective than Digoxin. Prophylactic medications of antiarrhythmic (propafenone, flecainide, and sotalol) are considerable in the case of severe symptoms despite the rate-controlling of the drugs. Those drugs should be reconsidered in the AFL with the presence of abnormal heart structure.[15]

In patients with persistent tachyarrhythmia, including AFL, the high possibility of recurrent arrhythmias in gestation period leads to the need of performing a pre-pregnancy ablation [6,15,16]. It is known that AFL mechanism involves a critical isthmus of slow conduction restricted by anatomic structures. The CTI is involved in the re-entry circuit in typical AFL, and the implemented RF energy to this isthmus has a chance to restore. Radiofrequency ablation proved to have high success rate to fight AFL, which is known to have been one of the atrial tachyarrhythmias. This results is noted with some smaller studies presenting 91% higher success with 1 procedure [1,7]. The Canadian Heart Rhythm Society and European Heart Rhythm Association elaborated that in both symptomatic and recurrent AFL, it is considerable to perform catheter ablation given the high success and relatively low complication rates for CTI dependent

AFL. Catheter ablation for the management of CTI-dependent AFL had an significant success of 97% and a recurrence probability of 10% over the 14 months, while catheter ablation for non-CTI dependent AFL technically has higher difficulty with a wide percentage from 73 to 100 percent in the success reports [3].

It is proved that radiofrequency catheter ablation of right sided atrial tachyarrhythmia is associated with high acute rate and significant long term recurrences, and repeated radiofrequency catheter ablation of recurrent atrial tachyarrhythmia was effective in most of the patients. However, the arrhythmogenic substance in patients with history of previous surgery proved to be complicated, and aiming for these arrhythmias needs 3D mapping that is detailed to raise the success rate of the ablation. [17]

Conclusion

Considering the efficacy and weighing the benefits, radiofrequency ablation with detailed 3D mapping prior to pregnancy is the most beneficial and safest choice for this 31 year-old woman with persistent typical AFL with a history Cone reconstruction due to Ebstein Anomaly

Acknowledgement

The working of this paper has no conflict of interest.

References

1. Niwa K. 2008. Adult Congenital Heart Disease with Pregnancy. Korean Circ J, 48 (4) : 251.

2. Attenhofer J C, Connolly H, Dearani J, Edwards W, Danielson G. 2007. Ebstein's Anomaly. Circulation, 115; (2):277-285

3. Kron I, Roeser M. 2017 Management of Ebstein's Anomaly. Ann Cardiothorac Surg, 6; (3): 266-269.

4. Malhotra A, Agrawal V, Patel K, Shah M, Sharma K, Sharma P, et a.l 2018. Ebstein's Anomaly: "The One and a Half Ventricle Heart". Braz J Cardiovasc Surg, 33; (4).

5. Anderson H, Dearani J, Said S, Norris M, Pundi K, Miller A, et al. 2013. Cone Reconstruction in Children with Ebstein Anomaly: The Mayo Clinic Experience. Congenit Heart Dis, 9; (3): 266-271.

6. Wackel P, Cannon B, Dearani J, Sessions K, Holst K, Johnson J, et al. 2018. Arrhythmia after cone repair for Ebstein anomaly: The Mayo Clinic experience in 143 young patients. Congenit Heart Dis, 13; (1): 26-30.

7. Silversides, Candice K., Spears, Danna A. 2015. Atrial Fibrillation and Atrial Flutter in Pregnant Women With Heart Disease. JACC, 1; (4): 293-295

8. Ian AG . 2012. Thrombosis in pregnancy: updates in diagnosis and management. Hematology Am Soc Hematol Educ Program, 1: 203-207. doi: 10.1182/asheducation-2012.1.203

9. Benedict MG, Jian C, Kathryn L, Serge B, Adrian B, Kristina HH, et al. 2017. Europace 00, 1–6. doi:10.1093/ europace/euw392

10. Poppas A, Shroff SG, Korcarz CE, et al. 1997. Serial assessment of the cardiovascular system in normal pregnancy. Circulation, 95: 2407-15

11. Niwa K, Tateno S, Akagi T, et al. 2007. Arrhythmia and reduced heart rate variability during pregnancy in women with congenital heart disease and previous reparative surgery.

Int J Cardiol, 122: 143-8.

12. Camm A, Kirchhof P, Lip G, Schotten U, Savelieva I, Ernst S, et al. 2010. Guidelines for the management of atrial fibrillation: The Task Force for the Management of Atrial Fibrillation of the European Society of Cardiology (ESC). Eur Heart J, 31; (19): 2369-2429.

13. Yu-Sheng L, Yung-Lung C, Tien-Hsing C, Ming-Shyan L, Chi-Hung L, Teng-Yao Y, et al. 2018. Comparison of Clinical Outcomes Among Patients With Atrial Fibrillation or Atrial Flutter Stratified by CHA2DS2-VASc Score JAMA Netw Open, 1(4):1-14. doi:10.1001/jamanetworkopen.2018.0941

14. Regitz-Zagrosek V, Blomstrom LC, Borghi C, Cifkova R, Ferreira R, Foidart J, et al. 2011. ESC Guidelines on the management of cardiovascular diseases during pregnancy: The Task Force on the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC). Eur Heart J, 32; (24):3147-3197.

15. Amar MS, Ebru E, Iris MH, Jassim AS, Titia PE, Mark RJ, et al. 2015. Atrial Fibrillation or Flutter During Pregnancy in Patients With Structural Heart Disease. JACC: Clin Electrophysiol, 1; (4): 284-292.

16. Cordina R, McGuire M. 2010. Maternal cardiac arrhythmias during pregnancy and lactation. Obstetric Medicine, 3; (1):8-16

17. Patel N, Deshmukh A, Pau D, Goyal V, Patel S, Patel N, et a.l 2016. Contemporary utilization and safety outcomes of catheter ablation of atrial flutter in the United States: Analysis of 89,638 procedures. Heart Rhythm, 13; (6):1317-1325.