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Vaginal Delivery in Maternal Cyanotic Congenital Heart Disease-Transposition of Great Arteries

Dr.Meenaakshi Karthikraj, PG student, Dr.Kavitha D'Souza, Professor & Head
Department of Obstetrics & Gynaecology, AJ Institute of Medical Sciences
Rajiv Gandhi University of Health Sciences, Mangalore, Karnataka, India.
Corresponding Author: Dr.Meenaakshi Karthikraj

Abstract: *Transposition of great arteries(TGA) is the most common cyanotic congenital heart disease(CHD). Pregnancy induces hemodynamic changes which in women with CHD, can endanger mother and child. We had two cases of TGA, one had shunt surgery pre-pregnancy, managed efficiently and delivered vaginally without any complications. Both had IUGR. Prostaglandin E_1(PGE_1) assisted labour induction for case-1 and oxytocin acceleration for case-2 done under infective endocarditis prophylaxis with cardiologist opinion. Uterine, fetal and cardiovascular monitoring done. Under epidural labour analgesia with levobupivacaine, both delivered vaginally, first by vacuum assistance. Postpartum period uneventful. We conclude that appropriate pre-pregnancy counselling, high-risk pregnancy management by multidisciplinary team at tertiary-care centre and effective analgesia, continuous monitoring and assisted second-stage of delivery can decrease the additional haemodynamic load of labour in CHD pregnancies.*

Keywords: *Cyanotic congenital heart disease(CHD), Transposition of great arteries(TGA), hemodynamic changes, pregnancy in CHD.*

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I. Introduction

Transposition of great arteries(TGA) is the most common cyanotic(5-7%) congenital heart disease(CHD)[0.4–1.3 %[1]]. Spontaneous mortality within first 2years is 90%[2]. Almost all patients who reach adulthood had prior reparative surgery. Some with large VSD & pulmonary vascular disease survive due to Eisenmenger physiology. Pregnancy induces hemodynamic changes[increase preload, decrease afterload, hypercoagulability], which in women with CHD, can endanger mother and child. We had two cases of TGA, case-1 uncorrected and case-2 had shunt operation, both managed efficiently and delivered vaginally without any complications.

Case-1: 28year primi, 32weeks gestation with oligohydramnios, had intermittent palpitations, breathlessness (NYH Association gradeII), central cyanosis, grade3/6 ejection systolic murmur(ESM), room air saturation 86%, normal renal functions, electrolytes, coagulation functions and albumin 3.5, with elevated serum lactate(19.6 mg/dl). Blood gas analysis showed pH 7.47, pCO_2 29 mmHg, pO_2 59.5 mmHg, base excess(BE) -1.4 and bicarbonate 23.9 on room air. Electrocardiogram(ECG) showed normal sinus rhythm but axis deviations. Chest skiagram(CXR) unremarkable. Echocardiogram revealed dextrocardia, levo-TGA, double-outlet right ventricle(DORV), large sub-aortic ventricular septal defect(VSD) with bi-directional shunt, severe pulmonary stenosis(PS), dilated right atrium and ventricle.

Case-2: 24year, primi, 36weeks 5days, in latent-phase of labour, undergone Blalock-Tausig shunt at 9 years, without correction of TGAs, was asymptomatic, had clubbing, central cyanosis, grade2/6 ESM, room air saturation 60%, Hb 16.5gm/dl, normal renal and coagulation profile, with elevated S.Lactate(23.9 mg/dl). Blood gas analysis showed pH 7.52, pCO_2 19.4mmHg, pO_2 69mmHg, BE -4.2 and bicarbonate 20.9 on 5 liters/min oxygen flow. Echocardiogram revealed dextro-TGA, VSD, severe PS, OS-Atrial septal defect(ASD), dilated right atrium and ventricle. Both had IUGR. After high risk consent, ProstaglandinE_1(PGE_1) assisted labour induction for case-1 and oxytocin acceleration for case-2 done under infective endocarditis prophylaxis with cardiologist opinion. Uterine and fetal monitoring done with supine-lateral tilt and continuous O_2(face mask). With labor onset, lumbar epidural catheter inserted at L_{3-4} level using air-syringe loss-of-resistance technique in sitting position, co-loading 200ml ringer lactate and radial artery catheter for hemodynamic monitoring by anaesthetist. $T_{9/10}$ level sensory blockade achieved with epidural levobupivacaine and fentanyl 80mg, 120µgm(case-1) and

32.5mg, 70µgm(case-2), respectively. Both had adequate pain relief. Repetitive trans-thoracic echocardiac evaluation done. Invasive monitoring with central venous pressures(CVP) considered only if necessary. With full cervical dilatation and vertex 'zero'station, successful vacuum-assisted vaginal delivery achieved in case-1 and vaginal in case-2 with adequate APGARscores. S.Lactate soon after delivery was higher[24.3mg/dl(case-1); 37mg/dl(case-2)]. Postpartum period uneventful.

II. Discussion

In TGA, there is AV-concordance with ventriculoarterial-discordance. In d-TGA, aorta arises rightward, anterior to pulmonary artery, from systemic right-ventricle(RV). Associated Lesions with d-TGA are anomalies of coronary ostia, VSD(45%), LVOT obstruction(25%) and Coarctation of aorta(5%). Sequelae are right(systemic) ventricular dysfunction, tricuspid valve regurgitation, subpulmonary stenosis and rhythm disorders. In our series, case-1 had the third variety and case-2 first variety(Fig.1). In case-1, aorta was anterior and left of pulmonary artery(PA). This variety constitutes only 7% of TGA types, where blood streaming specificity to a particular artery is favoured. In both patients, both arteries arise from RV. Presence of large VSD channelizes oxygenated blood towards RV/Aortic opening from LV. This flow is further complemented by a severely stenosed pulmonary valve for similar redirection of deoxygenated blood to PA. During delivery, cardiac output increases upto 80%, precipitous ventricular failure may result. Systemic vascular resistance decreases during pregnancy, but will increase dramatically at delivery. Delivery also results in sudden increase in venous return. Our patients had bi-directional shunt flow with mixing and this was worse in case-2. Shunt, decider of oxygenation adequacy. Clubbing and cyanosis, probably due to PS induced diminished PA blood flow. Pain, acidosis and hypoxemia worsen preexisting low pulmonary flow status and epidural analgesia becomes vital. S.Lactate levels detect extent of tissue hypoxemia. Higher postpartum lactate could be an indicator of hypoxemia during delivery, partly offset by pain relief. Risks of pregnancy to mother include cardiac risks[endocarditis, congestive heart failure[CHF](4.8%), embolism, dysrhythmias(4.7%), anoxia], obstetric events[PPH(8-29%), PIH(5.5-13%), preeclampsia(3.2-10%), preterm delivery(16-65%)] and to fetus[miscarriage, premature birth(12%), small-for-gestational age(14%), intra-uterine growth retardation[IUGR], mortality(4%)]. Impaired uteroplacental blood flow, explains increased foetal complication rate. Inheritance risk is 1.0–1.8%. Risk enhancers are pre-eclampsia(30%risk of CCF) or multiple pregnancies. Prophylactic low-dose aspirin after 12^{th}week is advised. Assisted reproductive technologies carry risks(fluid retention, hypercoagulability), that may endanger pregnant woman with CHD.Management involves risk-assessment, risk-evaluation, counselling and multidisciplinary, step-wise, management(Fig.2)[3]. Pre-pregnancy assessment and counselling is the best. WHO classification of maternal pregnancy risk is the most reliable method of risk assessment. If assessment reveals very high-risk, termination of pregnancy, early in pregnancy, in tertiary centre is advised. Both our cases were of WHO Risk category 3. Cardiac assessment includes, detailed history, electrocardiogram, echocardiography and additional testing. Aim of submaximal exercise testing, safe after 12weeks, is to achieve 80% of predicted heart rate. As chronotropic incompetence is common with CHD, cardio-pulmonary exercise testing with measuring gas-exchange, aiming for a respiratory exchange ratio of 1.0 may be an alternative to standard bicycle or treadmill exercise testing. Cardiac MRI safe after first trimester. Cardiac medications, angiotensin converting-enzyme inhibitors, angiotensin or aldosterone-antagonists, contraindicated during pregnancy. For women with low or intermediate risk, cardiac follow-up advised towards end of first trimester, ~20weeks and at ~28–32weeks of gestation. Those at high risk, need monthly or bimonthly followup. Predictors of maternal cardiovascular and offspring complications during pregnancy include Cardiac Disease in Pregnancy(CARPREG) and ZAHARA risk scores[4]. Both our cases had CARPREG maternal risk score of 1point=27%; ZAHARA maternal risk score of 1point=7.5% and 0.75=33.3% fetal risk.Antenatal monitoring includes foetal echocardiography ~18-21weeks and doppler velocimetry. Antibiotic prophylaxis given for Infective endocarditis, β-blockers for arrhythmias and anti-coagulants for thromboembolism. CHF peaks around late second-trimester/peri- & early postpartum. Serial measurements of natriuretic peptides(Pro-BNP/BNP) may help in risk stratification. Normal levels at 20weeks provide good negative predictive value. Treatment is by bed rest, supplemental oxygen, fluid balance; Inotropes, diuretics(↓pulm. congestion); β-blockers, Hydralazine & nitrates (↓afterload) and thromboembolic prophylaxis.

Misoprostol or low-dose continuous infusion/small repeated bolus Oxytocin can be used for labour induction. Labour is monitored with left lateral posture, CVP/Intravenous line + air filter(bubble trap) to prevent paradoxical emboli(air/thromboemboli), antibiotic prophylaxis and continuous O_2. Effective analgesia and assisted second stage can effectively decrease additional haemodynamic load of labour. Adequate analgesia balance systemic & pulmonary vascular resistances, minimize catecholamines release by pain, preventing increase in pulmonary vascular resistance. Decrease in systemic vascular resistance(SVR) and/or venous return avoided(as right-to-left shunt may be increased) by using intravenous fluids appropriately and avoiding aortocaval compression.Maintenance of SVR, intravascular volume, venous return and prevention of aortocaval compression taken care. Saline-filled syringe preferred over air-filled(loss-of-resistance technique used, to avoid paradoxical

air-embolism). LiDCOplus(lithium indicator dilution calibration system) monitor(minimally invasive continuous data provider) and continuous telemetry monitoring preferred owing to high incidence of arrhythmias. If urgent caesarean delivery planned, it is best in cardiac operating room with immediate availability of cardiopulmonary bypass.Vaginal delivery, with early epidural analgesia is the preferred mode of delivery. Average blood loss during vaginal delivery(500mL) counteracts the impact of auto-transfusion from contracting uterus. Low-dose continuous infusion/small repeated bolus Oxytocin or Prostaglandin-F analogues(contraindicated with pulmonary hypertension, right ventricular failure) can be used to control postpartum bleeding.

III. Conclusion

It is essential for appropriate pre-pregnancy counselling(pregnancy deteriorates functional class), high-risk pregnancy management of CHD mothers by multidisciplinary team(obstetrician, cardiologist, anaesthetist, neonatologist) at tertiary-care centre.

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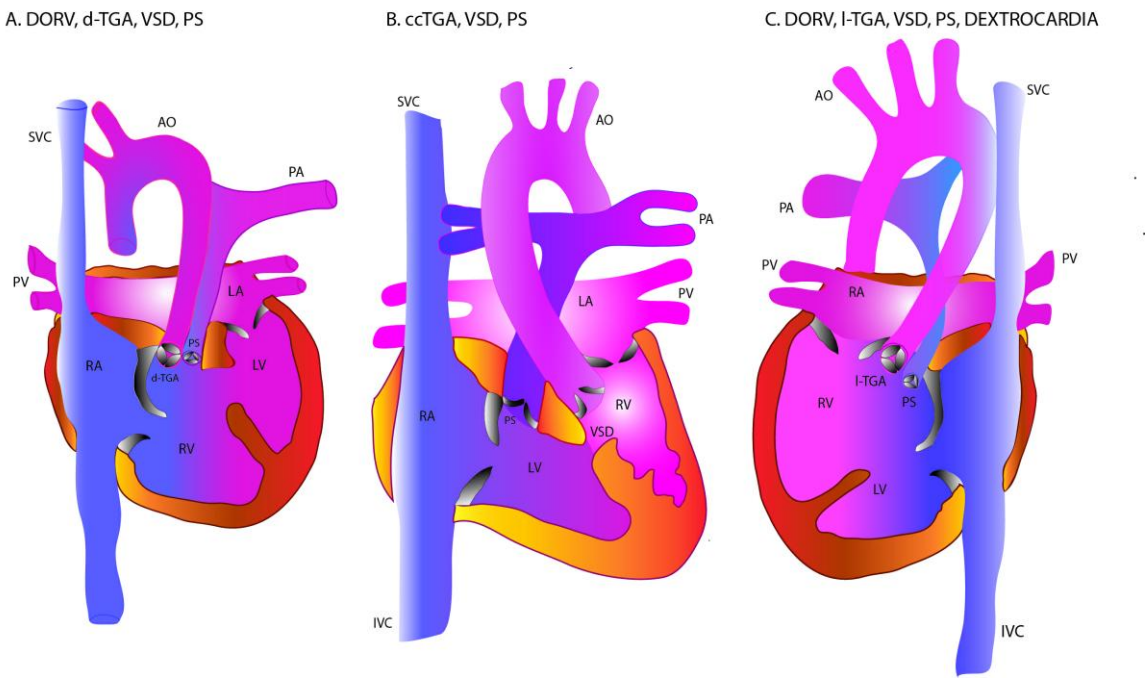

Fig. 1. Types of transposition of great vessels; Type A similar to Case 2, Type C similar to Case 1

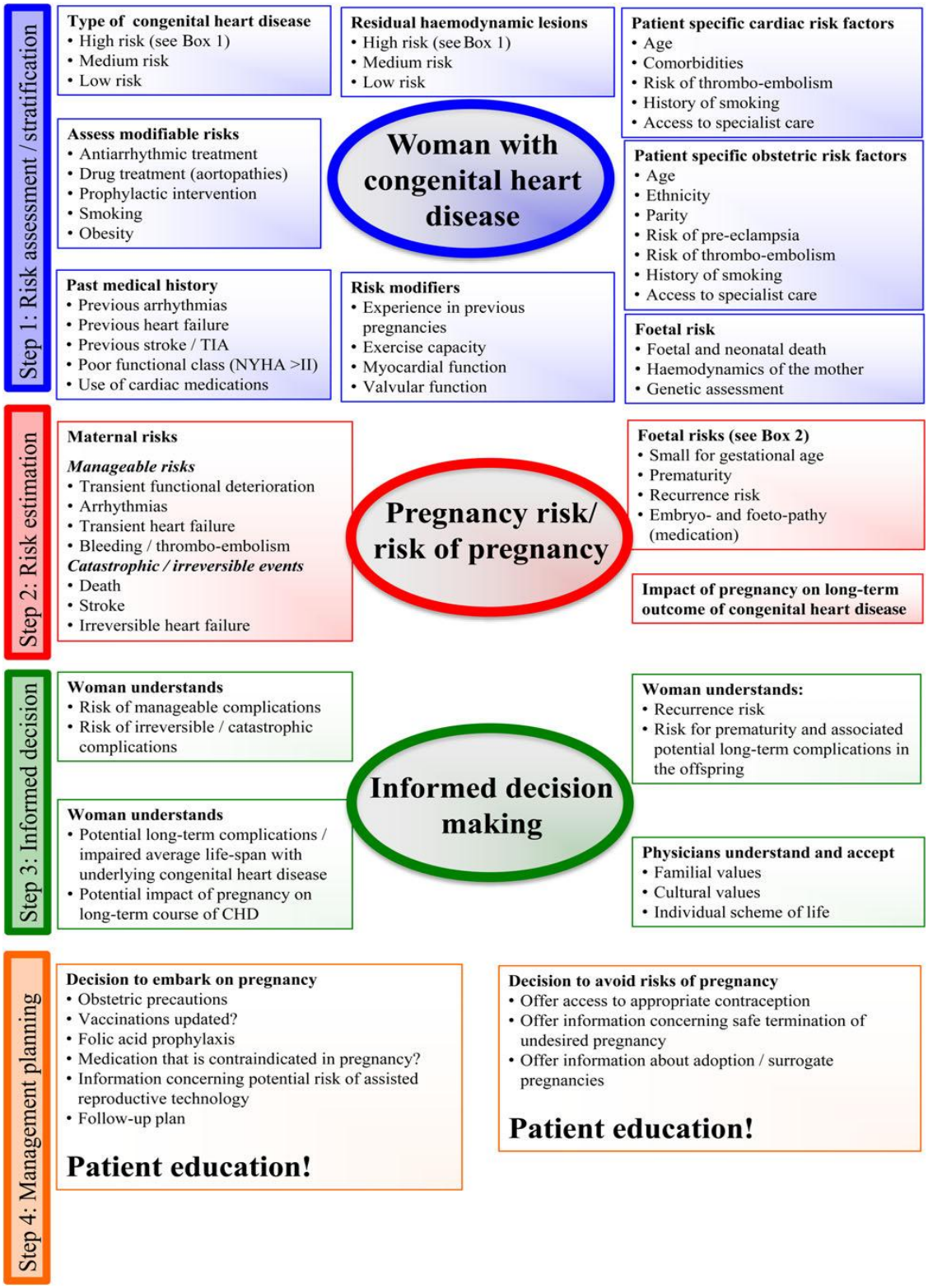

Fig. 2. Multi-disciplinary, multi-step risk assessment, counselling, and management planning[3]