University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2019, Vol. 5(1)

CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES COMPLICATED PREGNANCY (ccTGA) - A CASE REPORT

PUNITHAVATHI J JAYARAJ

Department of Obstetrics and Gynaecology,MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

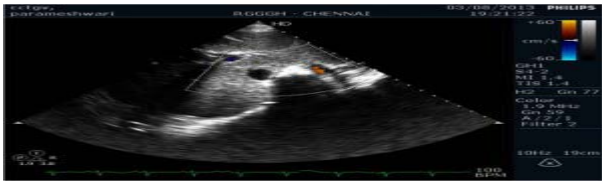
Abstract : ccTGA is a rare condition, accounts for 0.5-1 percent of all congenital heart diseases, characterised by both atrioventricular and ventriculoarterial discordance. Here we are presenting a case of ccTGA complicated pregnancy delivered at IOG for its rarity.

Keyword :ccTGA, pregnancy

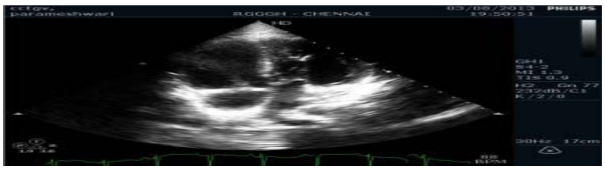
CASE SUMMARY:

18 yrs old, primigravida with LMP- 3/ 11/12 and EDD- 10/8/13 with 37 wks+3 days of gestation, referred from private hospital as a case of maternal congenital heart disease with anemia and IUGR. Since patient is not affordable for planned cesarean section she was referrd to our hospital. She was admitted with complaints of abdominal pain on & off for 1 week, breathlessness on exertion and palpitation. she was able to perceive fetal movements well. Her menstrual cycles were regular, marital history- married since 10 months with second degree consanguinity.past history- she was diagnosed as having heart disease (ccTGA with DORV,VSD,PS) at 15 yrs of age with complaints of dyspnoea and palpitation on exertion, she was on antifailure drugs, No family history of congenital heart disease.

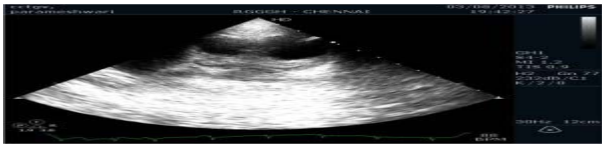
On examination, she was comfortable at rest, afebrile , not dyspnoic and tacypnoic, ANAEMIC +, No pedal edema, No clubbing and cyanosis. Her vitals were stable, JVP not elevated, CVS- S1S2 + with single loud S2, precordial thrill+, PANSYSTOLIC MURMUR in apical area & EJECTION SYSTOLIC MURMUR in pulmonary area+, RS- normal vesicular breath sounds+ ,no added sounds. Per abdomen:uterus 32 weeks, not acting, head unengaged, FH good. Per vaginal examination: cervix uneffaced, os 1 CM dilated, membrane+ , head at -3 station, pelvis adequate, no show and draining per vaginum. Her hemoglobin- 8.4 gm%, PCV - 25%, platelets-1.2 lakhs, RFT,LFT and electrolytes were normal.She was treated with 1 unit of Packed cell. cardiology opinion was obtained- advised to continue antifailure drugs, IE prophylaxis, to avoid hypotension during delivery.she kept under observation in ICU. She went into spontaneous labour on 19/7/13, under double setup with epidural analgesia and dopamine support, she delivered a alive boy baby of 1.7 kg with 8/10 and 9/10 apgar on 20/7/13 at 3.30 am by outlet forceps. PPIUCD was inserted, dopamine support weaned after 24 hrs, antifailure drugs continued, one more unit of packed cell transfused postnatally, post natal period was uneventful.Cardiology opinion reviewed on 14 th postnatal day: ECG showed sinus rhythm, RAD, absent septal Q wave in V5,V6 & AVL leads. ECHO showed situs solitus, levo cardia,AV discordance with VA discordance, large non constrictive VSD, severe valvular pulmonary stenosis, normal biventricular function, trivial tricuspid regurgitation.



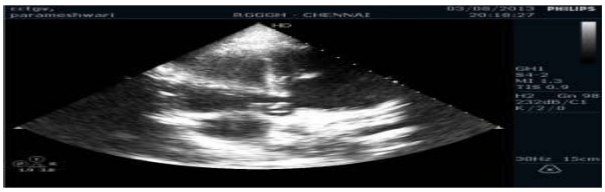
ECHO SHOWING SITUS SOLITUS



ECHO SHOWING ATRIOVENTRICULAR DISCORDANCE (RIGHT ATRIUM CONNECTED TO THE LEFT VENTRICLE AND LEFT ATRIUM CONNECTED TO THE RIGHT VENTRICLE with moderator band)



ECHO SHOWING AORTA AND PULMONARY ARTERY POSITIONED SIDE BY SIDE



ECHO SHOWING LARGE NON CONSTRICTIVE VSD

Advised to continue T.Digoxin o.25 mg 1/2 od, T.Lasix 40mg 1od, review after 3 months. ECHO done to the baby was normal. Both mother and baby discharged well.

DISCUSSION:

EMBRYOLOGY(MOOORE ET AL 2008): Normal development of ventricular situs occurs by twisting of the primordial heart tube to the right (d looping) in 5th week of gestation. Great arteries develops from the common trunk at the top of the fetal heart, common trunk consists of bulbouscordis and truncus arteriosis. tissue growth and blood flow through this results in spiral septation, which creats two arteries. ccTGA occurs due to abnormal looping of primordial heart tube to the left(l TGA) instead of right, resulting in both Atrioventricular and Ventriculoarterial discordance and abnormal positioning of great arteries. it means that right atrium connects to morphologic left ventricle, pulmonary artery exists from morphologic left ventricle(pulmonic ventricle), atrioventricular valve has two leaflets(mitral valve) and Left atrium connects to morphologic right ventricle, Aorta exists from morphologic right ventricle (systemic ventricle), atrioventricullar valve has three leaflets (tricuspid valve). Aorta and pulmonary artery positioned either side by side OR aorta may be anterior and to the left.(WARNES 2009, ALONSO- GON-ZALEZ 2010) Isolated ccTGA occurs only in 5 % of cases, 95% it is associated with other anomalies like, Ebstein like anomaly of tricuspid valve (90%), VSD (70%), Pulmonary valvular or subvalvular stenosis(40%), complete heart block (2%), Dextrocardia (2%), abdominal situs solitus also can occur.(THORNE ET AL 2009)

CLINICAL FEATURES: Mostly they are asymptomatic, symptoms like dyspnoea on exertion, syncope due to atrial arrythmias and complete heart block, heart failure due to systemic right ventricular dysfunction and tricuspid regurgitation occurs in 3rd - 4th decade of life.(ALONSOGONZLEZ 2010)

DIAGNOSIS: Prenatal diagnosis is possible with USG features like parallel course of greatarteries, dextrocardia, abnormal insertion of paillary muscle and abnormal AV valve.(SHIMA ET AL
2009), Chest X ray- strightened upper left cardiac border, dextrocardia. ECG- Septal Q wave in lead V4& V1(QR pattern), absent Q wave in V5& V6 (rS pattern). ECHO- situs solitus, L looped ventricles, L transposed great arteries, Right ventricle can be identified by COARSE TRABECULATIONS & MODERATOR BAND and Left ventricle by SMOOTH WALLED ENDOCARDIUM & FUNNEL SHAPED APEX. Exercise testing done to assess overall cardipoulmonary function, Cardiac MRI can also be done.

MANAGEMENT: Medical treatment for CHF, Surgical treatment includes classic or physiological repair and anatomical repair

AMERICAN COLLEGE OF CARDIOLOGY AND AMERICAN HEART ASSOCIATION TASK FORCE PRACTICE GUIDELINES 2008 RECOMMENDATION FOR ccTGA COMPLICATED PREGNANCY(level C evidence): scheduled cardiology evaluation & follow up before and during pregnancy, close monitoring by multidisciplinary health care team includes obstetrician,cardiologist and obstetric anaesthesiologist, IE prophylaxis at the time of rupture of membranes for vaginal delivery. NYHA CLASS III & IV, EF <40% & Significant systemic AV valve regurgitation shoud be counseled against pregnancy. Risk of congenital heart disease in the children of a mother with ccTGA is 6 %, Recurrence risk of d-TGA for siblings of ccTGA-2.6% (PIACENTINI ET AL 2005). **ANTICIPATION OF PROBLEMS DURING PREGNANCY** (**WARNES ET AL 2006**): Dysfunction of systemic right ventricle, Increased risk of heart failure with tricuspid valve regurgitation, atrial arrythmias, complete heart block.**M. Connoly**, **Martha grogan**, **Carol M and warnes et al 1999** studied the pregnancy outcome in 22 pregnancies with ccTGA, they found that 88% vaginal deliveries, 12% cesarean section amongwhich 50% for obstetric indications, 50% for perceived maternal cardiovascular risk. None of thewomen in this series reported infertility, No therapeutic abortions, 4 women had isolated ccTGA, 18with associated anomalies, NO PREGNANCY RELATED MATERNAL DEATHS, No one developed CHF in late pregnancy. In our case also pregnancy and delivery was tolerated well without any complications.

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