CONGENITALLY CORRECTED TRANSPOSITION OF GREAT ARTERIES- A CASE REPORT
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Abstract: Most cases with congenitally corrected transposition of great arteries (CC-TGA) are diagnosed in childhood because of associated cardiac abnormalities. When isolated or complicated by other mild anomalies, CC-TGA can cause no symptoms until adult life or can even go unrecognized. However, most of these patients present with heart failure with age. Our patient, a 28 yr old, multiparous lady was admitted and evaluated for progressive dyspnea, palpitations and chest pain. Patient was diagnosed to have CC-TGA with severe regurgitation of left sided (tricuspid) valve on echocardiography. Clinical findings, diagnostic methods and management are discussed briefly.

Keyword: Transposition of great arteries (TGA), congenitally corrected transposition of great arteries (CC-TGA), ECHO

INTRODUCTION:
Transposition is the most common cyanotic congenital heart lesion presenting in a neonate. The overall annual incidence is 20-30 per 100,000 live births¹. Levo -Transposition of the great arteries (L-TGA), also commonly referred to as congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital anomaly, accounting for less than 1% children with congenital heart diseases. Among these, it is equally rare to have no other associated structural cardiac anomalies².

CASE REPORT:
A 28 yr old lady, married for 10 yrs, P3L3A0 was admitted in our hospital with a history of retrosternal chest pain and difficulty in breathing of 1 week duration which progressed from NYHA class II to NYHA class IV. H/O orthopnea was present but there was no H/O paroxysmal nocturnal dyspnea or hemoptysis. H/O palpitations were present which were regular and paroxysmal. No H/O hospitalization for similar complaints in the past. No H/O DM/ HTN/CAHD/ asthma/ epilepsy/CVA/TIA. No H/O Rheumatic fever. Patient was born out of consanguineous marriage, with H/O uneventful childhood.

Patient married at the age of 18 yrs. She is P3L3A0. H/O dyspnea, palpitations and chest pain were present in antenatal and postnatal periods. Last child birth was 2 months back.

On examination (at admission), patient was moderately built and nourished, tachypnoeic, pallor was present but there was no evidence of cyanosis or clubbing. Pulse and blood pressure were normal. JVP: not elevated. Apical impulse seen and felt in left 6th ICS in anterior axillary line, hyperdynamic in character. Systolic thrill felt at the apex with soft/inaudible S1, a harsh blowing pansystolic murmur of grade 4/6, radiating to the axilla and the back, heard with diaphragm of the stethoscope, pt in left lateral position with breath held in expiration was heard. Tricuspid area S1 soft,a blowing pan-systolic murmur of grade 3/6 heard with diaphragm of stethoscope, pt leaning forward, breath held in inspiration. Pulmonary area: P2 was palpable and on auscultation a loud P2 with an ejection systolic murmur, grade 3/6 was heard with diaphragm of stethoscope, pt lying down and breath held in expiration. Aortic area: S1 S2 heard. No other pulsations/thrills/murmurs or heart sounds were found. Basal crackles and wheeze were heard in both chest fields suggestive of left sided heart failure. Patient’s haemoglobin was 9.8 mg%, other routine blood parameters were within normal limits. ECG: right axis deviation, RV hypertrophy, left atrial enlargement, poor progression of R waves from V1-V6.

Chest x ray- cardiomegaly.

Echocardiogram (ECHO) report is as follows:
- aorta arises from the morphologic right ventricle
- pulmonary artery arises from the morphologic left ventricle
- aorta is anterior and to the left of the pulmonary artery
- left sided AV valvular (tricuspid) regurgitation-moderate to severe
- right sided AV valvular (mitral) regurgitation (mild)
- IAS/IVS intact

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Most affected infants present with cyanosis within the first 2 or 3 days, and many require resuscitation. Infants may develop breathlessness and heart failure over the first 3-6 weeks as pulmonary blood flow increases. A chest X-ray may appear normal or may demonstrate the classic ‘egg on a string’- so-called “Pistol double run” sign - parallel lying aorta and the pulmonary artery; the morphological left and right ventricles as the right sided morphologic LV depolarises that the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle. This form is incompatible with life unless associated with another shunt lesion. Transposition is the most common cyanotic congenital heart lesion (CHD) presenting in a neonate. The overall annual incidence is 20-30 per 100,000 live births. It is more common in males than females, with a ratio of about 3:1. Maternal factors associated with an increased risk include rubella or other viral illness during pregnancy, alcoholism, maternal age over 40 and diabetes. Most affected infants present with cyanosis within the first 2 or 3 days, and many require resuscitation. Infants may develop breathlessness and heart failure over the first 3-6 weeks as pulmonary blood flow increases. A chest X-ray may appear normal or may demonstrate the classic ‘egg on a string’ appearance (heart is slightly enlarged and appears like an egg lying on its side, narrow vascular pedicle because aorta and pulmonary artery lie one in front of the other appears like a string). Echocardiogram images are diagnostic of transposition and associated anatomic lesions. In the parasternal long axis view, a so-called ‘Pistol double run’ sign - parallel lying aorta and the pulmonary trunk is seen. Palliative treatment with prostaglandin E1 infusion to maintain ductal patency and balloon atrial septostomy are usually required soon after birth. Surgical management includes Cardiac catheterisation and balloon atrial septostomy to increase the atrial level shunt and improve mixing. Definitive corrective procedure is the arterial switch operation (Mustard and Senning), which has been found to have similar mortality and less morbidity than atrial repairs. On the other hand, Levo-Transposition of the great arteries (L-TGA), also commonly referred to as congenitally corrected transposition of the great arteries (CC-TGA) is a rare congenital anomaly -1% children with CHD.

In ccTGA, these relationships are preserved but reversed. The AV valves and the anatomic coronary arteries follow their respective ventricles. Left sided coronary resembles the anatomic right coronary artery, courses in the AV groove and gives rise to infundibular and marginal branches. Right sided coronary resembles the anatomic left coronary artery, branches into the anterior descending and the circumflex arteries. Conduction system too follows the respective ventricles as the right sided morphologic LV depolarises first. Isolated ccTGA is the exception. Associated anomalies described are: VSD (70%), subvalvular pulmonary outflow tract obstruction (40%), abnormalities of left sided, systemic tricuspid valve- 90% dysplastic or Ebstein- like tricuspid valve.

Patients may remain asymptomatic till adulthood due to preservation of physiological blood flow. Failure of systemic, morphological right ventricle with varying degrees of systemic AV valve (tricuspid) insufficiency is the norm. On physical examination a Loud A2 due to anterior and leftward aorta and features of Tricuspid insufficiency with systemic ventricular failure are found. Chest Xray maybe normal or reflect the presence of associated lesions. Dextrocardia is observed in 25% - Typical ECG shows left axis deviation. AV node conduction abnormalities may manifest with time and progress to complete heart block. Echocardiogram is of diagnostic value.

Management: Systemic AV valve and ventricle may show signs of failure by 30 yrs of age that necessitate initiation of anticoagulative measures with diuretics and afterload reducing agents such as ACE- inhibitors and beta blockers. The risk of development of conduction abnormalities is cumulative over time. Therefore, periodic Holter monitoring is warranted. Permanent pacemaker placement maybe needed. Associated lesions like pulmonary stenosis or atresia, severe tricuspid valve regurgitation or VSD may need medical management. But more often surgical intervention may be necessary. Pregnancy and CC-TGV: Most women with ccTGA reach childbearing age. Appropriate evaluation, assessment and recommendations need to be made when these patients wish to pursue pregnancy.
Women with New York Heart Association functional class III or class IV symptoms, significant systemic ventricular dysfunction (ejection fraction <40%) or significant systemic atrioventricular valve regurgitation should be counseled against pregnancy. Care must be taken to avoid rapid volume shifts during pregnancy and parturition. The second stage of labor should be facilitated to decrease the excessive hemodynamic stress on the patient. Endocarditis prophylaxis should be administered to high-risk patients.

REFERENCES:
5. Classic Imaging Signs of Congenital Cardiovascular Abnormalities: Emma C. Ferguson, MD, Rajesh Krishnamurthy, MD, Sandra A. A. Oldham, MD, FACR. Department of Diagnostic and Interventional Imaging, Section of Thoracic Imaging, University of Texas Medical School at Houston.
7. Transposition of the Great Arteries Treatment & Management. Author: John R Charpie, MD, PhD; Chief Editor: Stuart Berger, MD [Medline]