A case report of dextrocardia with incomplete visceral heterotaxy

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Abstract:
Heterotaxy means different arrangement, these grouped into bilateral right sidedness or right isomerism and bilateral left sidedness or left isomerism. Normally there is asymmetric arrangement of thoracic, cardiac and abdominal organs. When there is symmetry asplenia or polysplenia syndromes results with complex congenital heart disease. Asplenia has bad prognosis in early neonatal period. The morbidity and mortality of polysplenia is associated with complexity of cardiac lesion. This case is a five year old boy presented with recurrent respiratory tract infection and clinically dextrocardia with evidence of left to right shunt. Investigation revealed atrial situs inversus with AV, VA concordance with systemic and pulmonary venous anomalies and left pulmonary vein stenosis, bronchial situs ambiguous with right side IVC and aorta without life threatening cardiac and extracardiac anomalies.

Keyword: Heterotaxy, Asplenia, Polysplenia, Ambigous, Dextrocardia

INTRODUCTION:
Heterotaxy is a Greek word which means heteros = other, different and taxis = arrangement1.
The ambiguous means visceroatrial situs uncertain or indeterminate because of disordered arrangement of abdominal and thoracic organs\(^2,3\). Normally there is right and left bronchi with different morphology and right sided liver, left side spleen and stomach and so on. When there are changes in this normal arrangement cardiac malposition or heterotaxy occurs.

**Case description:**
Five years old boy was presented with h/o recurrent respiratory tract infection since one year of age, no h/o cyanotic spells, squatting episodes and heart failure. There was no significant prenatal, natal and postnatal history. On examination of the patient weight 15 kgs, height 100cm, MAC (mid arm circumference) 12 cms, no cyanosis, no pallor, spO2 95% all 4 limbs, there were no markers of CHD, right precordial bulge, grade I right PSH and palpation and percussion confirmed dextrocardia. Auscultation revealed 2\(^{nd}\) sound normally split, P2 loud, grade II ESM at 2\(^{nd}\) RICS, grade III PSM at RLSB and there was no added sounds.

**LABORATORY INVESTIGATIONS:**
Blood smear didn’t show any abnormality.

**CXR:**
Dextrocardia, the right hemidiaphragm lower than left hemidiaphragm (figure 2), bilateral hyparterial bronchus and CTR 55%, bronchial left sidedness (figure 3) figure 2

**figure 3**

**ECHOCARDIOGRAPHY:**
Abdominal situs ambiguous by presence IVC and AORTA on right side (figure 4), IVC is posterolateral to AORTA on same side, Atrial situs inversus and dextrocardia (figure 5), L looping of ventricle, Atrioventricular concordance, Venticuloarterial concordance, Bilateral SVC present RSVC draining into coronary sinus, LSVC draining into RA, PAPVC , both right pulmonary veins drainage into RA (figure 6), left pulmonary venous drainage into LA.

**figure 1**

**Electrocardiogram:** P wave, T wave in lead I is inverted and biphasic QRS, P wave, T wave in lead AVR is positiveand biphasic QRS, Reverse progression of R wave in V1-V6, Consistant with atrial situsinvesus with dextrocardia (figure 1)
USG ABDOMEN:
Right liver with six hepatic veins, Gall bladder is right side, IVC and AORTA is right side of spine, AOTRA is anteromedial to IVC, Supra hepatic segment of IVC takes course anterior to aorta, travelling posterior and parallel to morphological right atrium before draining into morphology right atrium, Supra hepatic portion of IVC is draining into RA,

Last three hepatic veins directly drain into (RA figure 7), Spleen noted in left side- multilobulated.

BARIUM MEAL STUDY:
No evidence of gut malrotation (figure 8,9)

CARDIC CATHETERISATION STUDY:
It revealed supra renal IVC split into two, one continues as azygous drains into RSVC, other one runs on the left side suprahepatic segment became left sided and drains into morphologic RA (figure 10, 11). RSVC drains into coronary sinus into RA
LSVC drains into morphological RA (figure 13) which is inverted.

**CMR STUDY:**
Right side aorta and IVC, IVC drains into m right atrium, Revealed one normal size spleen with an accessory lobe and a small second spleen (figure 14), Mirror image dextrocardia, Atrial situs inversus and AV, VA concordance, Atrial and ventricular morphology (figure 15), bilateral SVC (figure 16), Left side pulmonary veins drain into morphologic RA due to malposition of septum primum and right pulmonary veins drain into morphologic LA, Left pulmonary vein at hilum has 40-50% stenosis (figure 17).
DISCUSSION:
This patient is a partial or incomplete heterotaxy with mirror image dextrocardia which is very rare. Which is neither asplenia nor polysplenia syndrome though he has multiple spleen. Visceral heterotaxy occurs in 0.8% cases of congenital heart disease\(^4\). Dextrocardia incidence is 1.6% among CHD. Situs inversus with dextrocardia has incidence of 1 in 8000 live births\(^5\). The mirror-image dextrocardia with situs inversus has 90-95% normal heart. Heterotaxy syndrome grouped into bilateral right sidedness (right isomerism, asplenia) and bilateral left sidedness (left isomerism, polysplenia)\(^1,2\). In about 15% of cases splenic tissue does not coincide with type of isomerism. Incidence of CHD in heterotaxy syndrome is very high 50-100%. Asplenia syndrome usually associated with mesocardia, though dextrocardia with associated cardiac anomalies like anomalous systemic venous drainage, endocardial cushion defects, common AV canal, common ventricle, transposition of great arteries, severe pulmonary stenosis or atresia and anomalous pulmonary venous connections can occur. Visceral heterotaxy with asplenia exhibits the highest incidence of bilateral SVCs with a completely unroofed coronary sinus, its about 64%\(^6\). Polysplenia syndrome usually associated with levocardia or dextrocardia and rarely mesocardia. The associated cardiac anomalies specific to polysplenia is interrupted IVC with azygous communication to SVC and also anomalous systemic and pulmonary venous drainage, ASD, VSD, DORV and left sided obstructive lesions. Bilateral SVC in polysplenia although rare compared to asplenia, its about 13%\(^6\). Bilateral suprahepatic IVC was present in 28% of the asplenia group and in 6% of the polysplenia. In a few cases a right-sided IVC may become left-sided at the level of the liver and at its suprahepatic segment. Rarely bronchial morphology is not consistent with atrial morphology\(^7\). But heterotaxy is not always complete, many times findings are variable\(^8\). There is segmental approach for prospective two-dimensional echocardiographic study of complex heart disease, these segments are 1. Systemic and pulmonary veins 2. Atrial situs 3.
AV connection 4. Ventricles and infundibulum 5. VA connection 6. Great arteries and ductus arteriosus. Echocardiography plays major role in diagnosis of cardiac malpositions but other investigations also play major role in identifying cardiac and extra cardiac lesions. Cardiac lesions associated with heterotaxy syndrome are complex and prognosis is bad to worse. In our case abdominal situs is normal with multiple spleen, bronchial situs ambiguous (bilateral left bronchi) with mirror image dextrocardia. The bronchial left sidedness is not consistent with bilateral left atrium, this patient had normal left and right atrial appendages. And both aorta and IVC on right side. Inferior vena cava split into two and one drains into M. RA and other part continuous as azygous continuation and drains into RSVC. Both systemic (bilateral SVC) and pulmonary (PAPVC) venous connection anomalies are present in our case. Right SVC which drains into coronary sinus and persistent left SVC drains into M RA. CMR identified 40-50% stenosis of left pulmonary vein near hilum. Left pulmonary veins drain into morphologic RA due to malposition of septum primum. Our case is unique because dextrocardia with heterotaxy syndrome without neonatal life threatening cardiac and extra cardiac anomalies which usually present in heterotaxy syndromes. To completely define various anomalies associated with heterotaxy syndrome other modalities of investigations like CT and CMR plays an important role. Surgical management of PAPVC by excision of atrial septum and creation of new atrial septum is needed.

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