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AN UNUSUAL CASE OF CYANOSIS

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ABSTRACT

A 2 months old boy with severe respiratory distress, central cyanosis and abnormal mass like structure in left hypochondrium who later on found to be a case of complex congenital heart disease with heterotaxia (asplenia syndrome)

INTRODUCTION

Heterotaxia is a clinical condition where position of heart and viscera is altered[1]. There are 3 terminology that includes all the positions: situs solitus, situs inversus totalis, situs ambiguous (heterotaxia). Classification and diagnosis of abnormal cardiac position is done in 3 segmental approach:[2]

1. Determination of atriovisceral situs
2. Determination of position of ventricles
3. Determination of position of great vessels

CASE REPORT

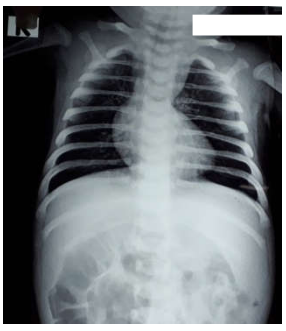
A 2 months old boy born out of non consanguineous marriage was admitted in Medical College, Kolkata with a chief complaints of frequent episodes of respiratory distress since birth. He was a term baby born out of normal vaginal delivery and antenatal history was uneventful.

At birth, the birth weight was 2.5kgs, length 53 cm, head circumference 32cm and there were no morphological anomalies. Baby was exclusively breastfed.

On examination, general condition was poor. Temperature was raised (101 deg F),Heart rate was 176/min, Respiratory Rate was 67/min with intercostal and subcostal retractions with central cyanosis, chest bilateral diffuse crepitations and occasional wheeze, cardiovascular system-1st and 2nd heart sound was heard with a grade 3 short systolic murmur best heard over left upper parasternal region with no radiation was noted. Saturation was 65% in room air improving to 70% with oxygen @ 6L/min. On gastrointestinal examination, a liver like mass was felt in left hypochondrium the consistency of which was different from spleen.

Investigations revealed haemoglobin -18.6 g/dl, total leukocyte count- 24,000/mm3, hematocrit- 53%, total platelet count -2.9 lakh/mm3, blood culture was positive for gram negative bacteria, C-reactive protein level was high- 11mg/dl. Liver function tests and renal function tests and electrolytes were normal. Peripheral blood smear shows occasional Howell jolly bodies.

Chest X-ray revealed boot shaped heart with upturned left sided apex with empty pulmonat bay and bilateral oligemic lung fields and ? bilateral hepatic shadows with absent fundic gas shadow on left side.



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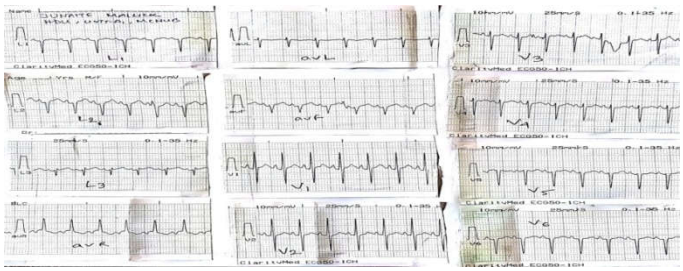
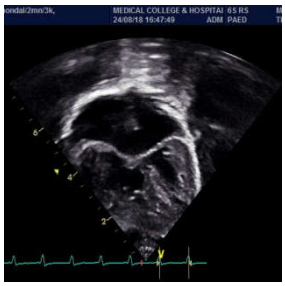
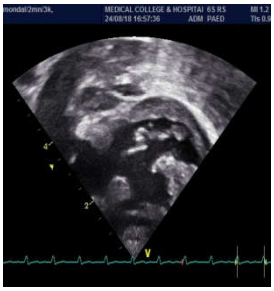
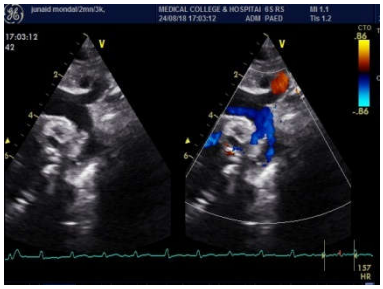


ECG showed normal p axis with superior Q axis with reversed pattern of QRS complex from V1 to V6 (normally in V1 QRS is negative and it becomes positive when proceed to V6 but here the pattern is reversed).

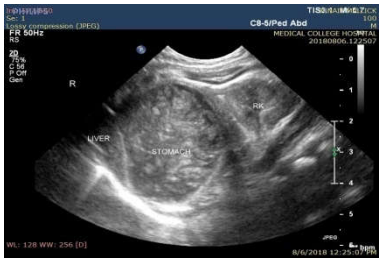
Echocardiography showed levocardia, complete balanced atrioventricular septal defect(AVSD), large(6mm) ostium primum ASD(L>R), large inlet VSD(11mm), large ostium secondum ASD(L>R), pulmonary atresia, vertical PDA(L>R), native branch pulmonary arteries were not well visualized, left arch with no coarctation, aorta from left ventricle with good ventricular function.

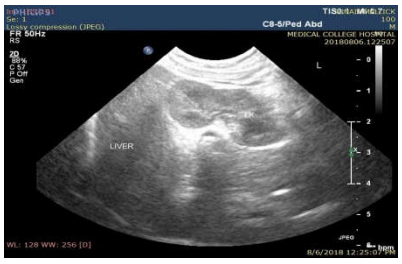




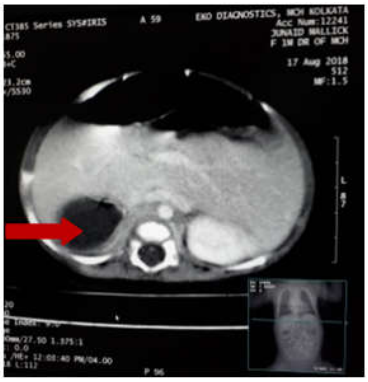


Ultrasonography (USG) of whole abdomen revealed liver extending from right hypochondrium to left hypochondrium. Intrahepatic biliary radicals (IHBR) were not dilated. Gall bladder was seen on left side, contracted. Spleen was absent.

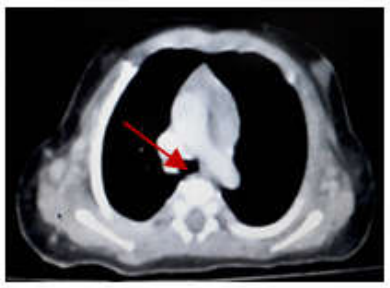




Contrast enhanced computed tomography (CECT) abdomen showed same findings that of USG abdomen with absent spleen and right sided stomach bubble.



CT angiography of thorax revealed Atresia of Main branch pulmonary artery.



DISCUSSION

The cause of this disorder is failure of differentiation into right and left sided organs during development. It is a rare condition and if not evaluated properly, may be missed. One should suspect heterotaxia if any of the followings are found[3]: 1) symmetrical midline liver, 2) discordant cardiac apex and stomach bubble, 3) biliary atresia in a neonate with congenital heart disease, 4) symmetrical main stem bronchi, 5) superior axis on ECG.[4]

CONCLUSION

Levocardia with situs inversus and dextrocardia with situs solitus are almost associated with major cardiovascular malformations and extracardiac malformations[5].

If any of the 5 above mentioned features are found, heterotaxia should be kept in mind as a differential diagnosis and must be ruled out, else may be missed.

Most common cause of death in case of asplenia syndrome is sepsis by capsulated organisms mainly pneumococcus[6].

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent. In the form the patient has given his consent for images and other clinical informations to be reported in journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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