Cyanotic Congenital Heart Disease with Asplenia Syndrome with Pyogenic Meningitis

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Abstract
Asplenia is a well-known condition in which there is malposition and malformations of the abdominal and chest organs. We present a case of asplenia syndrome in an 8 month old infant who was diagnosed to have complex congenital cyanotic heart 4 months back. The infant was admitted this time with pyogenic meningitis. In this article we want to drive the attention of paediatricians regarding the importance of screening the child with complex heart disease for other associated anomalies.

Case Report
An 8 month old male child, 1st by birth order, born of nonconsanguineous marriage, Hindu by religion was brought with complaints of cough, breathlessness and fever since 1 week. There was a history of admission 4 months back for pneumonia where he was diagnosed to have complex congenital cyanotic heart disease. On admission child was febrile and physical examination revealed an underweight infant with weight of 3.8 Kg. His heart rate was 144/min, RR 64/min, B 92/58 mm Hg in supine position in right arm with grade 1 clubbing and central cyanosis and saturation in all 4 limbs being 76-78%. Examination bulging nonpulsatile anterior fontanelle suggestive of meningitis. Examination of the cardiovascular system revealed normal first and single second heart sounds and a grade 3/5 pansystolic murmur at apex transmitted best to back. The respiratory, abdominal and central nervous system examinations were unremarkable.

Investigations revealed Hb of 12.6 g/dL; TLC of 39,300/cmm (N 62%, L 32%, M 4% and E 2%); platelets 6,70,000/cmm with normal serum electrolytes, liver and renal function test. Blood culture showed no growth. CSF examination revealed proteins 121 mg%, glucose 15 mg% and total nucleated cells of 1,600 (N 85%, L 15%) with polymorphonuclear predominance.

Gram stain and ZN stain showed no organisms and CSF culture grew Pseudomonas Aeruginosa. X ray of chest showed dextrocardia with consolidation on left side without any cardiomegaly. ECG showed marked right-axis deviation of the P wave and of the QRS complex, and low voltage in the precordial leads, V1 through V6, without any rhythm disturbance. Echocardiogram revealed common AV canal defect and ventricular inversion with severely hypoplastic pulmonary artery and small PDA. In view of dysmorphism with dextrocardia with complex congenital heart disease abdominal ultrasonography was done which revealed right side isomerism with liver on both right and left side and absent spleen. To confirm the anatomy of thorax and abdomen MRI was done which confirmed the findings with evidence of hypoplastic pulmonary artery and MAPCAs.

A diagnosis of Situs ambiguous i.e. dextrocardia with complex congenital cyanotic heart disease with right sided isomerism (asplenia syndrome) with pyogenic meningitis was made. He was treated with I.V. antibiotics based on sensitivity of Pseudomonas isolated for 21 days to which patient responded dramatically. Patient was immunized with 2 doses of heptavalent pneumococcal vaccine and Hib vaccine and referred to cardiothoracic surgery for Blalock Taussig shunt.

Discussion
Situs solitus signifies the customary, or normal, asymmetrical arrangement of the visceralvascular anatomy. Situs inversus is the mirror image of situs solitus in toto.
Although this condition is embryologically related to other situs anomalies, situs inversus must be strictly differentiated, because it has significantly different pathophysiologic and clinical implications. All other conditions occur in a spectrum called situs ambiguous, which is commonly changed to the situs ambiguous.

Patients with situs ambiguous tend to be grouped with those in whom right- or left-sided structures predominate. Generalizations may be made in these groups. Patients with right-sided symmetry typically lack a spleen, whereas patients with left-sided symmetry typically have a segmented spleen or multiple splenules.

These common characteristics have led to the somewhat arbitrary classification of asplenia and polysplenia. However, heterotaxia, or situs ambiguous, occurs in a continuum. Many infants with situs ambiguous present with severe congenital cardiac anomalies.

In situs ambiguous, there are estimates that 1.44 infants per 10,000 are affected. This number is based on patients with congenital heart disease and is likely to be an underestimation, although it may include most of these patients. Although most cases of heterotaxia are sporadic, many cases are familial, and some are X linked. Thus, the abnormality is more common in males than in females. Severe and complex cardiac abnormalities are likely to be apparent at birth or soon afterward. In humans, familial situs ambiguous has been related to both autosomal and X-linked inheritance patterns, although most cases arise sporadically. Situs inversus and situs ambiguous have been described within the same family trees. A submicroscopic deletion in Xq26 and a deletion at 18p have been associated with familial situs ambiguous. Further, balanced and unbalanced autosomal translocations have also been described in sporadic cases of situs ambiguous. Finally, environmental factors, including exposure to retinoic acid and maternal diabetes, have been implicated in laterality defects among the offspring of affected parents.

Situs ambiguous, or heterotaxia, is associated with other conditions of major clinical relevance, such as intestinal malrotation, biliary atresia, splenic abnormalities and consequent immunologic derangements, faulty gastric suspension mechanisms, displacement of abdominal viscera, and aberrant vascular structures and vascular connections. Each of these abnormalities is derived from an embryologic
inability to determine laterality and establish the complex solitus asymmetry, whereas symmetrical structures remain unaffected. The heart is most often affected, and it is the organ that most frequently leads to clinically detectable abnormalities. Table 1 compares the salient features of right and left isomerisms.

Management consists of appropriate treatment of the congenital heart disease, preventing infections in asplenia syndrome with use of vaccine against capsulated organisms and timely surgical referral.

The major causes of mortality and morbidity in the heterotaxy syndromes are undoubtedly the cardiac malformations that typically occur in these conditions. These are based on the inability of the complex asymmetrical connections to develop correctly, and they predictably consist of an ambiguous and single atrium, a single ventricle, and conotruncal anomalies such as truncus arteriosus and transposition of the great vessels.

Vascular malconnections that are associated with high mortality and morbidity rates include total anomalous pulmonary venous connections (TAPVCs). Visceral abnormalities with clinically notable adverse consequences include biliary atresia and the absence of the spleen. Malrotation may become clinically evident if obstruction develops secondary to the presence of Ladd bands or if a midgut volvulus supervenes. Intrinsic duodenal obstruction, such as that secondary to duodenal diaphragm, may also occur.

References

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| **Right sided isomerism** *(Asplenia syndrome)*  
Almost all have transposition of the great arteries, severe pulmonic stenosis or atresia, and AV canal with large ventricular septal defect or single ventricle; two-thirds have anomalous pulmonary venous connection to vena cava or portal system.  
Severe cyanosis from birth on; symmetric liver and bronchi; malrotation of bowel common; Heinz or Howell-Jolly bodies or pitted red cells on peripheral blood smear; 80% have asplenia. |
| **Left sided isomerism** *(Polysplenia syndrome)*  
Most commonly have left-to-right shunts through atrial septal defects, ventricular septal defects, endocardial cushion defects, or double-outlet right ventricle; may also have left-sided obstructive lesions like aortic stenosis or coarctation; transposition and pulmonic stenosis are unusual; pulmonary veins may connect to either or both atria; two-thirds have interruption of the inferior vena cava.  
Cyanosis usually mild or absent; congestive heart failure common; malrotation of bowel common; symmetric liver, but less so than in right atrial isomerism; superior P axis in ECG common; occasional biliary atresia; usually have polysplenia. |

WHY MULTIPLE SEXUAL PARTNERS?
Understanding why people have multiple partnerships is key to efforts to change behaviour, with the realization that behaviours range from polygamy itself, to longer-term quasi-polygamy women are trapped by economic necessity and male domination.
Interestingly, both women and men prominently cite dissatisfaction with their primary partnerships, sexually and otherwise. Such relationship dissatisfaction is ascribed to lack of communication and romance, partner’s lack of skill in lovemaking monotony domestic discord, and desire for variety in partners and sexual practices.
Women, relationship with older men seem particularly often to be related to luxury good and status. Alcohol clearly facilitates risky sex.
What is the best way to address multiple sexual partnerships? What is needed are high-quality multilevel (mass media, community, clinical setting, individual) approaches to reinforce behavioural change on the basis of sound intensive research with the audience.
Mass-media approaches encompass billboards, printed advertisements, and television and radio, including call-in shows.
Yet in concert with promotion of male circumcision and use of condoms, especially for high-risk sex, it appears we are finally embarking on the right road to prevent hyperepidemic HIV transmission.