

SINGLE VENTRICLE WITH DEXTROCARDIA IN AN ADOLESCENT FEMALE

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Abstract: An adolescent girl with progressively increasing generalized cyanosis, failure to thrive progressive weakness since childhood presented with overt clubbing fingers with no family history. On investigation, it was proved to be a case of dextrocardia with double outlet (single) ventricle.

INTRODUCTION

Congenital Heart disease (CHDs) in children continues to be a major health problem world wide with the incidence in different studies varying from 1-17.5/1000 live births and 10% of spontaneously aborted fetuses. The available data on CHDs in India shows an incidence of 1-5 / 1000 live births. An angiocardigraphic study of fourteen cases of dextrocardia projected with the importance of sequential chamber localization and segmental approach to the description of the anatomy of the heart was discussed¹. Congenital cardiac anomalies seen with dextrocardia included pulmonary stenosis, pulmonary atresia, single ventricle². In one series³, fifty children with established congenital heart disease (CHD) were surveyed for the immune profile ventricular septal defect (VSD) was the commonest lesion (56%) followed by Tetralogy of Fallot (TGA, 16%) atrial septal defect (ASD; 8%) patent ducts arteriosus (PDA, 4%) transposition of great arteries (TGA; 4%) aortic stenosis (AS; 4%) and pulmonic stenosis (PS), tricuspid atresia (TA), single ventricle with pulmonic stenosis (SV with PS) and dextrocardia with ToF(2% each).

Single ventricle combined with dextrocardia is one of the rarest congenital anomaly in clinical practice. It is also known as Double Inlet Ventricle or Univentricular heart. Dextrocardia occurs in 1 per 7000 to 1 per 10,000 population. Nearly 90 to 95 percent of these individuals have a normal heart. When Dextrocardia is associated with abnormal or ambiguous visceral arrangements, the heart is usually abnormal and malformations including Univentricular heart are noted.⁴

CASE REPORT

An adolescent girl of 16 years age, presented with complaints of generalized blackish discoloration of skin and eyeballs that were present in infancy and increased its intensity during childhood. Her fingers started swelling, progressively associated with increasing generalized weakness since childhood. She left school earlier and spent most of time in bed with complete inability to participate in household activities. She had difficulty in walking with frequent fatigue and breathlessness on exertion; she had recurrent bouts of low grade fever for which she was managed at local hospitals. She started losing her feminine look at puberty

with sparse axillary and pubic hairs along with smaller breast development compared with peers. There was no history of menarche or regular periods except occasional trickling of blood per vagina along with severe pain in loin that made her bedridden. She had an uneventful home delivery with nil immunization history till date. None of her siblings had any major medical or surgical problems. There was no history suggestive of pulmonary or cardiac diseases in her paternal and maternal tree. Her mother did not suffer from any disease during pregnancy and had no ante-natal, intra natal and post natal cares. She did not practice any form of temporary contraceptive methods and had never taken any form of medication during pregnancy.

Clinical Findings: Clinically there was marked cyanotic violaceous black tongue, deep blue sclera, thin built, of short stature with mesomorphic features. Her weight was 24 kgs. With height of 123 cm. and BMI=15.9 She had clubbing with blackish discoloration on twenty nails. On examination of cardio-vascular system a right para sternal lift was palpable; a systolic thrill was noted; there was bruit over precordium. On auscultation 1st heart sound was normal; the 2nd heart sound was single and loud. The ejection systolic murmur grade III was found on auscultation. On examination of respiratory system there was breathing difficulty with a respiratory rate of 45 per minute to breath sounds were vesicular. Pulse oxymetry showed 67% oxygen saturation. Secondary sexual characters were present except there was no adolescent change in voice, adolescent increase in width of girth of pelvis and buttock. There was better growth of auxiliary or pubic hair (Tanner stage 3), in comparison of less well developed breast (Tanner stage 2) in comparison to her age.

Investigations: Hemoglobin level was 10.2 gm/dl with few Reticulocytes and nucleated red cells and microcytes. Complete blood count: normal : ESR 34 mm 1st hr. Roentgenographic examination of chest (figure 1) confirmed Dextrocardia without cardiomegaly. A bulge on upper girth border of cardiac silhouette in the postero-anterior projection suggested a rudimentary outflow chamber. Pulmonary vasculature was diminished (oligemic) and suggested pulmonary outflow obstruction. Electrocardiogram findings (figures 2) revealed left axis deviation with spiked P waves in VR, VF, V1, V2 and bifid in II, III, V3-V6 A negative P wave in lead I indicated reversed atrial arrangement confirming Dextrocardia (atrial situs inversus) QRS complexes were characteristically larger in right chest leads and progressively smaller from V4-V6 in left sided leads. There was 'W' pattern of QRS complex in Lead II and V2 and T waves were deeper in I,

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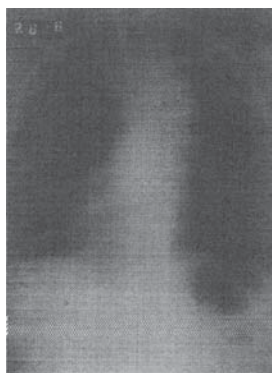


Fig.1. Chest X-Ray findings

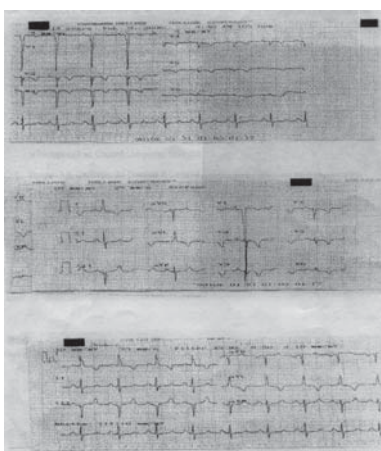


Fig.2. Electrocardiogram findings

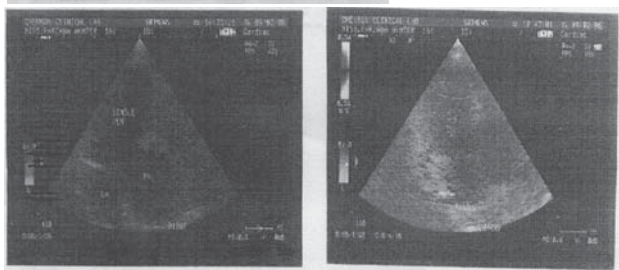


Fig.3. Echocardiography in 2D: M-Mode and Colour Doppler Study

VR, VI, all precordial leads, taller than normal in others. Echocardiography in 2D; M mode and colour doppler study (figures 3) revealed the absence of ventricular septum. The features matched with Double inlet left ventricular variety of Double inlet ventricle or single ventricle. There was a main ventricle with morphology of a normal left ventricle and two inlet valves and the right ventricle was represented by a small rudimentary outflow chamber. Aorta was arising from the rudimentary chamber and the pulmonary artery from the main chamber. Pulmonary stenosis was present.

DISCUSSION

Single ventricle combined with Dextrocardia is one of the rarest finding in clinical practice so that in standard undergraduate medical text books this findings have not been mentioned. In the developing countries, due to poor antenatal, intra natal and post natal management congenital heart disease are often missed. Asymptomatic and uncomplicated cases often go unnoticed. Double inlet (single) ventricle can account for up to 2-3% of all congenital heart disease⁵. Caregivers of the patient had been moving from pillar to post since childhood for her illness. Nobody told her the bitter truth that at the present state of development of medical science some form of palliative cardiothoracic operation is necessary for her. These costly operations will definitely improve the overall prognosis and prolongation of life by operation on pulmonic stenosis and shunt operation. But in the developing world skilled man power for this type of maneuver is still fewer than requirement. Moreover, social institutions are rarely available to the common people.

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LITERATURE REVIEW

Adult Nephritic Syndrome: Non-Specific Strategies for Treatment

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Irrespective of aetiology, the nephritic syndrome presents a range of potentially serious complications. These include thrombo-embolism infection and hyperlipidaemia. Despite the prevalence of the nephrotic state among renal patients, there has been little prospective analysis of the therapeutic approach to these potentially life-threatening events even though their pathogenesis has been examined in some detail. Most of these complications are more prevent once the albumin concentration falls below 20 g/L and it is recognized that restoration of serum albumin significantly diminishes their frequency. However, this may be difficult to achieve, especially in adults. The problems of thrombo-embolism and infection are of immediate concern but, in persistent cases, the additional issues of hyperlipidaemia and loss of bone density also require consideration for therapy. Thus, in addition to specific attempts to reduce proteinuria, it is recommended that high risk nephritic patients receive anticoagulation, pneumococcal vaccination and lipid lowering therapy. Strategies for the preservation of bone density should also be considered, particularly in patients who receive high dose corticosteroids. Among a range of non-specific treatments for proteinuria, angiotensin-converting enzyme inhibitors appear best in terms of efficiency and safety. Prospective trials are required to clarify the longitudinal impact of these generic strategies on the protection of the persistently nephritic patients.