

Cor triloculare biatriatum – A case report

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ABSTRACT

Cor triloculare biatriatum or double inlet single ventricle is a congenital heart defect in which both atria are connected to a common or dominant ventricle. The present report presents an index case and describes the embryological basis and clinical aspects of this extremely rare anomaly. A three months old infant presented with extreme respiratory distress without cyanosis and repeated chest infections. The patient was diagnosed to be a case of single ventricle with both atria opening in the common ventricular chamber. The common ventricular chamber (single ventricle) was connected to a rudimentary outflow tract. The great arteries were in a position of d-transposition of great arteries. However, there was no pulmonary or aortic stenosis. A clear concept and awareness regarding this condition and its clinical manifestations is bound to facilitate timely intervention with improved success rates.

Keywords: Double inlet single ventricle, Congenital heart defect, Cardiac anomaly.

INTRODUCTION

The incidence of congenital heart defects is 70 per 10,000 live births, whereas the single ventricle heart has an incidence of about 0.05-0.1 per 10,000 live births.¹ The malformation is a complex congenital heart defect characterized by a single or common ventricular chamber that receives atrial blood flow by ways of two atrio-ventricular valves or through a single valve. Some cases have been reported previously in literature.^{2,3,4} The ventricular output may occur directly into the great vessels or partly or completely through a rudimentary outflow chamber.⁴ Single ventricle results from failure of development of two-trabecular heart components, the left ventricle from the primitive ventricle and the right ventricle from the bulbus.⁵ The patients usually present within the first month of life. As the ductus arteriosus reduces in caliber within the first few days of life, those infants with associated severe subpulmonary stenosis or aortic obstruction present with cyanosis or poor peripheral perfusion, respectively.

MATERIALS AND METHODS

A 3-month old male infant (weight 3 kg) presented in the Department of Pediatrics of Government Medical College, Amritsar, Punjab, India with severe respiratory distress and noisy respiration. The infant was apparently well for 22 days of life and was feeding well. Thereafter, he developed repeated respiratory infections. The infant was treated in a primary care centre for chest infections. The respiratory distress worsened and the baby refused to feed. The baby was referred to our hospital for further investigations and management.

The anatomy department was requested after investigations on the infant to provide an embryological basis for this rare condition to the residents and faculty of pediatrics department. Our institute has a well developed liaison between clinical and basic departments. The first and corresponding author frequently visits the pediatrics ward and outpatient departments to carry our anthropometric studies on newborns and infants. Also the anatomy faculty and residents perform ward duties in pediatric and other departments after teaching hours. Those patients who do not survive pediatric surgery their hearts are frequently sent to anatomy department for opinion and review regarding any possible congenital anomaly. The progress of this infant was meticulously followed by the authors as this is a rare condition. A comprehensive review of literature was performed and embryological aspects were ascertained in detail pertaining to the case.

RESULTS

Examination done revealed bilateral crepitations and wheeze in the chest. In addition, there was an ejection systolic murmur in the pulmonary area. The respiratory rate was 60/min and the pulse was 180/min and was regular. The chest radiograph revealed a globular cardiac shape and a thin narrow pedicle. Bronchovascular markings were moderately increased suggesting pulmonary overcirculation. The Echocardiogram was done and yielded non-specific changes. On Echocardiography, both the atria were seen opening into the common ventricle (Fig.1).

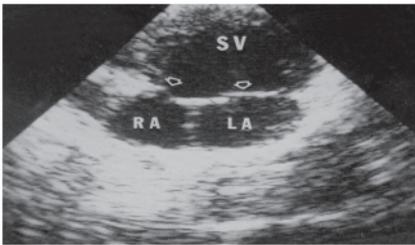


Fig1: Echocardiogram showing both the atria opening into the common ventricular chamber.RA-Right Atrium;LA-Left Atrium;SV-Single Ventricle.

The aorta was seen arising anteriorly from the rudimentary chamber and pulmonary trunk was arising from the single ventricle posteriorly. The great arteries were in a position of d-transposition of great arteries. However, there was no pulmonary or aortic stenosis. The Echo of the infant was reported as a single ventricle with d-transposition of great arteries.

The common ventricular chamber (single ventricle) was connected to a rudimentary outflow tract (Fig.2).

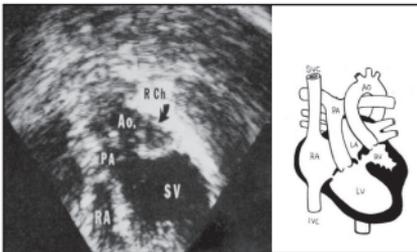


Fig.2: Echocardiogram showing both great arteries arising from single ventricular chamber.Right side of figure shows the diagrammatic representation of the Echo. LV-Left Ventricle;RV-Right Ventricle,R Ch-Right Chamber, PA-Pulmonary Artery;Ao-Aortic opening .

The patient was further referred to a higher medical centre for surgery.

DISCUSSION

Cor triloculare biatriatum is also known as single ventricle or double inlet ventricle or a univentricular heart. There are four morphological types of single ventricle mentioned in literature.^{5,6} The most common variety is when the trabecular pattern of the main chamber resembles a left ventricle as in the present case. When the shape and trabecular pattern resembles a right ventricle the chamber is called a single ventricle of right ventricle (RV) type. The third variety is an undifferentiated single ventricle (single ventricle

resembling left or right ventricle).The fourth type is when the characteristics resemble both the ventricles with complete absence of ventricular septum.

The blood passes from atria into the single ventricle through the atrio-ventricular (AV) valve.⁷ There are five possibilities for the AV valve to connect with the single ventricle.^{7,8} There may be two patent AV valves connecting to single ventricle (such a scenario is called double inlet left ventricle as in present case/ double inlet right ventricle).There may be a scenario of atresia of right/left AV valve or there may be a common AV valve. In some cases there may be a straddling of the left or right AV valve in the rudimentary chamber.

There is complete mixing of oxygenated and deoxygenated blood in the single ventricle. The blood then passes through outlet chamber into the pulmonary trunk and aorta. In a single ventricle of left ventricle type (seen in 80% of single ventricle cases) the outlet chamber is considered to represent right ventricle chamber without an inlet portion. The single ventricle is separated from an infundibular outlet chamber by a bulbo-ventricular sulcus. The opening is called bulbo-ventricular foramen and ventricular septal defect. The rudimentary outlet chamber associated with the LV type of single ventricle is identified at the base of the heart on the right side (normally related infundibular chamber). This chamber may give rise to both great arteries and on occasion no great artery. Most commonly a single vessel, usually the aorta arises from the outlet chamber. Even when both great vessels originate from the outlet chamber the aorta tends to arise anteriorly. Thus in a single ventricle, the great arteries are virtually always transposed. If the outlet chamber is on the right side aorta will be in a position of d-transposition of great arteries (d- TGA / d malposition). If the outlet chamber is on the left, aorta will be in a position of l- transposition of great arteries or l - malposition. The single ventricle is usually accompanied by defective septation of truncus arteriosus leading to malposition of the great arteries.⁹

The morphogenesis of a single ventricle has been discussed by authors and an ontogenic explanation presented based on the study of normal human embryos.^{10,11} A partial arrest in the primitive widening of the AV canal has been suggested. Even after the heart tube finishes looping and folding, the atrioventricular canal provides a direct pathway only between the future atria and the future left ventricle. The superior end of the presumptive right ventricle (bulbus cordis), but not the presumptive left ventricle is initially continuous with conus cordis and truncus arteriosus that will eventually give rise to both the aortic and pulmonary outflow tracts. Extensive remodeling of the heart aligns the right and

left atrioventricular canals with their respective atria and ventricles and the left and right ventricles with their future outflow tracts.

Any defect in the complex process of extensive repositioning and remodeling of the heart tube and the complex process of septation of ventricles and truncus arteriosus of the heart tube will result in incorrect alignment of the right and left atrio-ventricular valves and the outflow tracts with large ventricular septal defect or complete absence of a ventricular septum resulting in single ventricle of various types.¹²

The etiology of most of the cardiac abnormalities appears to be multifactorial i.e they stem from the interaction of environmental or outside influences (teratogens) with a poorly defined constellation of the individual's own genetic determinants. The various factors which may lead to disturbed morphogenesis of heart are perturbations in the haemodynamics of heart, abnormal programmed cell death or abnormal cardiac neural cell crests. All these factors may be under the control of cardiac specific regulatory genes and experiments on transgenic mouse models have revealed that over expression, partial function and completely lacking function (knock outs) of cardiac specific genes may result in various types of morphogenic abnormalities of heart.^{12,13}

The diagnosis rests mainly on Echocardiography. The echocardiogram shows presence of a single ventricle as well as anatomy of atrio-ventricular valves and ventricular arterial connections. The opening of great arteries may be associated with stenosis or atresia of either pulmonary (more common) or aortic valve. The definitive treatment in live born infants is surgical however a prenatal diagnosis done before 24 weeks warrants the termination of pregnancy. Clinical manifestations relate to the presence or absence of pulmonary stenosis. Babies born with single ventricle and severe pulmonary stenosis will have cyanosis where as other infants with wide open pulmonary circulation will develop congestive heart failure secondary to pulmonary over-circulation.¹⁴

The present index case presented at three months of age with repeated chest infections, extreme respiratory distress and minimal cyanosis. A clear concept and awareness regarding this condition and its clinical manifestations is bound to facilitate timely intervention with improved success rates.

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