Double Inlet Left Ventricle - A Case Report

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Case Report

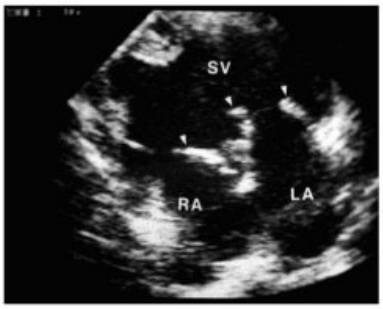
Abstract: Double inlet left ventricle is a very rare congenital anomaly which occurs in about 5 - 10 of 100,000 live births. Only the left ventricle of heart is developed properly and both atrial chambers carry blood into this ventricle. This means that oxygen-rich blood mixes with oxygen-poor blood. The mixture is then pumped into both the body and the lungs. The problem most likely occurs early in the pregnancy, when the baby's heart develops. However, the exact cause of DILV is unknown. We are presenting here a case of 2 year old child with single ventricle having left ventricular morphology diagnosed by echocardiography. Great vessels were transposed and pulmonary artery was stenosed.

Introduction

The term "single ventricle anomaly" is used to describe a group of cardiac defects that may differ quite dramatically from each other but share the common feature that only one of the two ventricles is of adequate functional size. Some of the anomalies described as single ventricle defects include: Tricuspid atresia Hypoplastic left heart syndrome Double inlet left ventricle Many of the heterotaxy defects Some variations of double outlet right ventricle Single ventricle of heart with double inlet is a rare congenital anomaly and has an incidence of about 0.05- 0.1:10,000 live births. The prenatal diagnosis of this rare birth defect has rarely been described. The diagnosis is suspected on a four-chamber view and confirmed by echocardiographic analysis. It is corrected by Fontan procedure, or Fontan/Kreutzer procedure, which is a palliative surgical procedure used in children with complex congenital heart defects. It involves diverting the venous blood from the right atrium to the pulmonary arteries without passing through the morphologic right ventricle.[1]

Case report

A two year old boy, born of nonconsanguineous marriage, fullterm normal delivery presented with bluish dislocation of skin, nose and lips. He has poor weight gain and poor feeding since 3 months of age. On examination baby was cynosed with grade two clubbing. His weight was 6 kg that was 2 standard deviation below 5%. Examination showed ejection systolic murmur and systolic thrill. 2D Eecho was suggestive of single ventricle of left ventricular morphology. Right ventricle was rudimentary which is called as outlet chamber and great vessels were transposed. Aorta was arising from outlet chamber and pulmonary artery from morphologcal left ventricle. Pulmonary artery was stenosed.



Discussion

DILV is one of several congenital heart defects known as single (or common) ventricle defects There are three types of cardiac anomalies associated with the single ventricle double inlet heart: left ventricular type(70% cases) right ventricular type with a rudimentary contralateral chamber (15-20%cases) a single ventricle of the indeterminate type. A dominant left ventricle with rudimentary right ventricular chamber is the most common form of double inlet ventricle. These forms of single ventricle can sometimes be differentiated by prenatal ultrasound.[2] Aspects of anatomy requiring special attention in the postnatal period include: the size and position of the communication between the single ventricle and the outflow chamber, the bulboventricular foramen, the anatomy and function of the atrioventricular valves, and the anatomy of the pulmonary and aortic outflow tracts. Pulmonary and aortic outflow tract obstruction may occur and impact significantly on patient prognosis and management. Subpulmonary obstruction may be due to posterior deviation of the infundibular septum, atrioventricular valve tissue or subvalvular fibrous tissue. Varying degrees of hypoplasia of the ascending aorta, aortic valve, and outflow chamber, or a restrictive bulboventricular foramen may result in aortic outflow tract obstruction. These aspects of anatomy may

be confirmed at catheterization but may be elucidated by echocardiography.

a) Associated anomalies: These include anomalies of the valves entering the cardiac chambers, including stenosis, overriding, or straddling imperforate valves. In over 90% of cases of left single ventricle, the great arteries will be transposed. Atresia of the pulmonary or aortic valves or trunks, truncus arteriosus and subaortic stenosis may also be associated.

Diagnosis

Prenatal: a single ventricular chamber on a four-chamber view of the heart with two atrioventricular valves. Postnatal: Ultrasound examination confirms the presence of a single ventricular chamber as well as the anatomy of the atrioventricular valves and the ventriculoarterial connections. Examination of ventricular outflow and aortic arch anatomy are critical because of associated anomalies. The anomaly can be detected on the four-chamber view of the heart: the ventricular septum is absent, and the two atria empty into a single ventricular chamber.

Embryology

In normal development of the heart, the left ventricle trabecular component is formed from the inlet segment of the primitive ventricle of the primary heart tube, while the right ventricle trabecular component forms from the outlet segment of the bulbus. A failure of development of these trabecular components results in a single ventricle.

Most of the congenital heart defects are sporadic. The major genetic cause for congenital heart defects includes the following:

(a) chromosomal disorders and single gene disorders constituting 8%

(b) 2% of environmental teratogens and

(c) 90% multifactorial disorders.A multifactorial means both genetic and environmental factors interact, to interfere with the development of the heart.

Increased incidence of CHDs has been noted with intrauterine viral infections, maternal drug and alcohol consumption during first trimester of pregnancy and pregnancy-induced systemic maternal disease.Various chromosomal disorders associated with this anomaly are Trisomy 21 (Down syndrome, Trisomy 18 (Edwards Syndrome), Trisomy 13 (Patau syndrome), 45 X (Turner Syndrome), Tetrasomy 22q (cat eye syndrome), Tetrasomy 12q (pallister killian syndrome), Fragile -X Syndrome, Deletion 22Q11.2 syndrome, Wolf-hirschhornsyndrome, 1q21 Microdeletions:

Microdeletion on chromosome 1q21.1, Noonan syndrome,Ellis- van Creveld and Kabuki syndrome which is characterized by distinct facial anomalies, variable degrees of mental retardation, CHDs and skeletal malformation. The CHDs include ASD, VSD, TOF, PDA, TGA, aortic Coarctation, single ventricle with common atrium and right bundle branch block[3]Management: a) If the diagnosis is made before 24 weeks, the option of termination can be offered to the parent. Otherwise, after regular monitoring to detect signs of fetal hydrops, birth is recommended in a tertiary care center. b) Postnatal management Patients with only one functioning ventricle can usually expect either a heart transplant or a series of palliative surgeries or sometimes both. The Fontan procedure, or Fontan/Kreutzer procedure, is a palliative surgical procedure used in children with complex congenital heart defects. It involves diverting the venous blood from the right atrium to the pulmonary arteries without passing through the morphologic right ventricle. The Fontan is usually done as a two staged repair.

1. The first stage, also called a Bidirectional Glenn procedure or Hemi-Fontan involves redirecting oxygen-poor blood from the top of the body to the lungs. It is done in 4-6 months of age The superior vena cava (SVC), which carries blood returning from the upper body, is disconnected from the heart and instead redirected into the pulmonary arteries. The inferior vena cava (IVC), which carries blood returning from the lower body, continues to connect to the heart. At this point, patients are no longer in that delicate balance, and the single ventricle is doing much less work. They usually can grow adequately, and are less fragile. However, they still have marked hypoxia (because of the IVC blood that is not fed into the lungs to be oxygenated). Therefore most patients are referred for another surgery.

2. The second stage, also called Fontan completion, involves redirecting the blood from the IVC (inferior vena cava) to the lungs as well. This surgery is usually performed when the child is 18 months - 3 years old. After this final step, the baby is no longer cyanotic.At this point, the oxygen-poor blood from upper and lower body flows through the lungs without being pumped (driven only by the pressure that builds up in the veins). This corrects the hypoxia, and leaves the single ventricle responsible only for supplying blood to the body.[4]

References

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