CASE REPORT

A Five (5) chamber heart (Cor Triatriatum) in Infancy: A rare congenital heart defect

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ABSTRACT

Five-chambered heart is extremely rare in children. We report a case of asymptomatic five chamber heart detected in infancy. The patient is 2-day-old and managed in a special care baby unit (SCBU) for neonatal sepsis. During routine follow-up at the age of 1 month, she was found to have an asymptomatic murmur. Echocardiograph reported five-chambered heart, concluding that it is Cor triatriatum, supravalvular pulmonary stenosis and secundum atrial septal defect. The child is still been followed-up and is still asymptomatic at 7 months. Five-chambered heart, although rare, can occur even if asymptomatic.

Key words: Asymptomatic, five chamber heart, infancy

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INTRODUCTION

Heart defects are among the most common birth defects. Worldwide, congenital heart defects/diseases contribute substantially to morbidity and mortality in childhood, especially in the third-world countries where facilities for management are often deficient.[1] About 35,000 infants (1 out of every 125) are born with heart defects every year in the United States.[2] The incidence in the tropics is not expected to differ from the 8-10 per 1000 live-births documented in the developed world.[3] The defect may be so slight that the baby may be asymptomatic for many years after birth, or so severe that it is life threatening. Heart defects are the leading cause of birth defect-related deaths.[4] However; the use of echocardiography has improved description of congenital heart diseases and their early diagnosis.[5] This, has led to dramatic increases in survival of children with serious heart defects.

Cor triatriatum is a rare congenital cardiac anomaly, in which a fibromuscular membrane divides the atrium in two. It was first reported in 1868.[6] Cor triatriatum, a heart with 3 atria (triatrial heart), is a congenital anomaly in which the left atrium (cor triatriatum sinistrum) or right atrium (cor triatriatum dextrum) is divided into 2 parts by a fold of tissue, a membrane, or a fibromuscular band.[7] However, variable types of subtotal cor triatriatum are also noted, with only the right or left pulmonary veins draining into the upper chamber.[8]

Cor triatriatum represents 0.1% of all congenital cardiac malformations and may be associated with other cardiac defects in as many as 50% of cases. Examples of associated cardiac defects include atrial septal defect, persistent left superior vena cava with an unroofed coronary sinus, partial anomalous pulmonary venous connection, ventricular septal defect and more complex cardiac lesions such as tetralogy of Fallot, atrioventricular canal and double outlet right ventricle. Associated bicuspid pulmonary valve, aortic valve atresia and heterotaxy have also been described.[9]

Congenital pulmonary vein stenosis is a very rare association with cor triatriatum.[10]

The morbidity and mortality of cor triatriatum sinistrum is high in those who are symptomatic in infancy. This is due to the severely restrictive opening in the accessory membrane and the association with major cyanotic or acyanotic congenital heart lesions. Mortality may exceed 75% in untreated symptomatic infants. Significant sequel is unusual with cor triatriatum dextrum as it is not commonly associated with life-threatening symptoms or major congenital cardiac defects.

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CASE REPORT

A female baby seen on the second day of life was delivered by a 25-year-old primiparous Youth Corper. The mother attended antenatal clinic in Federal Medical Centre Owerri from the 16th week of gestation. However, during the 8th week of pregnancy, the mother had threatened abortion for which some drugs were given (injections and tablets names unknown) at a hospital in Lagos. She also had some liquid herbal preparation at 8 months of gestation; other drugs she took were essentially haematinics. She denied a history of alcohol, coffee and cigarette use. The pregnancy was carried to term. She went into spontaneous labour with an 18-h history of rupture of membrane. Delivery was spontaneous, vertex and supervised by medical personnel. She gave birth to a 3.6-kg baby girl who cried immediately after, and breastfeeding commenced. The child was transferred to the Special Care Baby Unit from the postnatal ward on account of history of fever on day 2 of life.

Examination revealed a full-term newborn, febrile (38.1©) with a respiratory rate of 100 cycles per minute, acyanotic, heart rate of 110 beats/min heart sound- I and II only no murmur heard. A diagnosis of probable neonatal sepsis was made. Investigations carried out included blood culture that did not yield any growth. However, the child completed a course of antibiotics (Ampicillin–Sulbactam and Gentacin) and, with clinical improvement, was discharged home.

At follow-up at the 2nd week of life, the child weighed 3.5 kg, heart rate was 130 beat/min and a grade 3 pansystolic murmur was heard, following which a chest radiograph and electrocardiograph (ECG) was requested.

At 1 month of life, a review of ECG showed:
heart rate: 128 beats/min,
RR: 468 mS,
P: 70 mS,
PR: 94 mS,
QRS: 54 mS,
QT: 290 mS,
QTc: 426 mS,
Axis P-39©,
QRS-147© and

T-18, which suggested supraventricular arrhythmia, abnormal right superior axis deviation.

Chest radiograph showed perihilar mottling on both the lung fields. Heart and thoracic cavity were within normal limits. An impression of bronchopneumonia was made by the Consultant Radiologist.

Murmur was still grade 3/6, pansystolic maximal at the left-lower sternal edge.

Echocardiograph was subsequently requested and the result showed that a 2D echocardiogram was done [Figure 1]with the following results:

Situs solitus
Separate left atrium with dividing membranous running in the direction of long axis of left atrium close to the lateral wall.

No evidence of flow across the septum. Mitral valve draining the larger, more medial chamber, normal atrio-ventriculo-arterial connection. Normal relationship of aortic root to the pulmonary trunk.

Supravalvular pulmonary membrane causing stenosis noted. High velocity turbulent flow in the main artery noted (PA Vmax = 335 cm/s PG = 45 mm).

Secundum atrial septal defect about 1.2 mm noted with left to right shunt.

Normal wall motion and thickness, good global left ventricular contractility.

Normal pericadium.
Conclusion
Cor Triatrietum.
Supravalvular pulmonary stenosis.
Secundum atrial septal defect [Figure 1]

The child is presently being followed-up regularly. During her last visit, at 7 months of age, she weighed 9.6 kg, length was 73 cm, occipito-frontal circumference was 47 cm and mid-upper-arm circumference was 15 cm (all within normal for age).

Cardiovascular examination revealed a child with heart rate of 120 beats/min, apex beat located at the 4th left intercostals space, with a grade 3/6 systolic murmur heard loudest at the upper left sternal edge. The respiratory rate was 42 cycles/min and no organs palpably enlarged on digestive system examination.

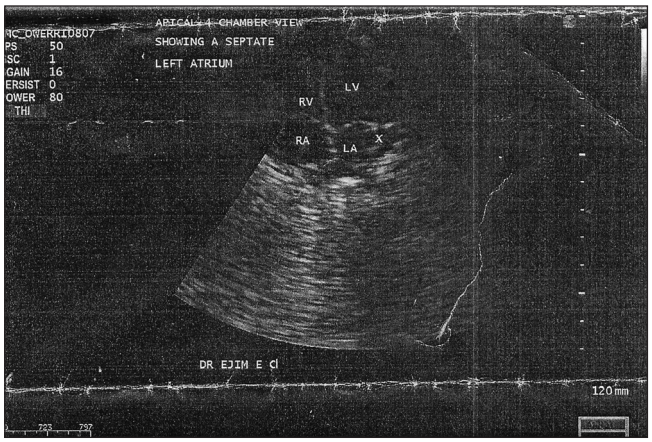

Figure 1: Echocardiogram showing the distal chamber (X) and the left atrium; the right atrium (RA), left ventricle (LV), and right ventricle (RV)

DISCUSSION

This was the first case of a five-chambered heart seen and managed in the Federal Medical Centre Owerri. The case demonstrates the need for comprehensive evaluation of patients and provision of cardiac investigative facilities (X-ray, ECG and Echocardiograph machines) in health facilities. In our patient, it was an incidental finding at a routine follow-up clinic. Subsequent investigative evaluations led to the confirmation of the diagnosis of a five-chambered heart in a child who has remained asymptomatic. The dearth of facilities in our hospital is obvious here, where the patient had to be referred to the University of Nigeria Teaching Hospital Enugu for echocardiograph.

Clinical manifestations are often delayed due to the presence of a large opening; and include an asymptomatic murmur. Finding is mostly incidental on routine cardiac imaging; it therefore highlights the need for exhaustive evaluation and management because such conditions, although rare, can still be found here.

In conclusion, we reported a case of an asymptomatic congenital-heart defect A five-chambered heart-cor triatriatum, an extremely rare condition.

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