Extended Abstract
Volume 4:2, 2020

Research and Reports in Medical Sciences

Open Access

Euro Heart Congress 2020: Cor Triatriatum Sinister and Patent Ductus Arteriosus in a 23 year-old Filipino

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Abstract

Cor triatriatum is one of the rarest cardiac disease or condition that involves the left atrium. It happens and represents only 0.1-0.4% of all congenital heart diseases. It can be illustrated or denoted as one of the atrium gets divided into two chambers by a fibromuscular membrane and in general appear on the left atrium. Cor triatriatum is considered as a result because of embryologic failure of the common pulmonary vein to become subsume into the left atrium during the fifth week of embryonic development.1 The insuffucient incorporation of the common pulmonary vein results into a obstructive membrane at junction of the common pulmonary vein and the left atrium. As a result it creates two separate chambers within the left atrium; the proximal chamber consists of pulmonary veins and their convergence, whereas the distal chamber includes the left atrium and left atrial appendage . Cor triatriatum is an unusual membrane, or split, present in the left atrium above mitral valve, resulting in varying degrees of obstruction to flow into the left ventricle. It is a rare lesion, accounting for only 0.1% of CHD defects87.

The pathophysiology is essentially identical to that of mitral stenosis, and only echocardiography often differentiates the two conditions. Surgical repair is undertaken when the diagnosis is made. Mild forms of cor triatriatum might be undetected for months and might also take years in patients whose only real symptoms are recurrent respiratory infections and wheezing. CTS was first found and noted in the 1800s by Andral and Church and named “cor triatriatum” by Borst in 1905; it occurs in 0.1% of clinically diagnosed cases of congenital heart disease (CHD) and 0.4% of CHD autopsy cases. This discrepancy reflects the fact that most cases involve a nonobstructive membrane with questionable clinical relevance. The prevalence in the general population is likely to be less than 0.004%.1-3 Fewer than 350 cases have been reported since 1968. There may be a slight male predominance.Cor triatrium sinistrum is more common. In this defect there is a proximal chamber which receives the pulmonic veins and a distal chamber located more anteriorly where it empties into mitral valve. The membrane which separates the atrium into parts varies significantly in size and shape. It may appear similar to a diaphragm or be funnel-shaped, bandlike, entirely intact (imperforate) or contain one or more openings (fenestrations) ranging from small, restrictive-type to large and widely open.

In the pediatric population, this anomaly may be associated with major congenital cardiac lesions such as tetralogy of Fal lot, double outlet right ventricle, coarctation of the aorta, partial anomalous pulmonary venous connection, tenacious left superior vena cava with unroofed coronary sinus, ventricular septal defect, atrioventricular septal defect, and common atrioventricular canal. Rarely, asplenia or polysplenia will be reported in these patients. In an adult, cor triatriatum is frequently an isolated finding. Cor triatriatum dextrum is very rare and results from the complete persistence of a right sinus valve of the embryonic heart. The membrane divides the right atrium into a proximal (upper) and a distal (lower) chamber. The upper chamber receives the venous blood from both vena cavae and the lower chamber is in contact with the tricuspid valve and the right atrial appendageThe natural history of this defect depends on the size of the communicating orifice between the upper and lower atrial chambers. If the communicating orifice is small, the patient is critically ill and may succumb at a young age (usually during infancy) to congestive heart failure and pulmonary edema. If the connection is big, patients may present in childhood or young adulthood with a clinical picture similar to that of mitral stenosis. Cor triatriatum may also be an peripheral finding when it is non obstructive. Diagnosis by ECHO, possibly followed by MRI.
The ductus arteriosus is a usual fetal artery connecting the main body artery and the main lung artery (pulmonary artery). The ductus allows blood to deflect away from the lungs before birth.

Every baby is born with ductus arteriosus. After birth, the opening is not needed and it usually narrows and closes within the first few days.

Sometimes, the ductus doesn't close after birth. Failure of ductus to close is common in premature infants but not common in full-term babies. In most children, the cause of PDA is unknown. Some babies can have other heart defects along with the PDA. Normally the heart's left side pumps blood only to the body, and the right side pumps blood only to the lungs. In a baby with PDA, excess blood gets pumped from the body artery into the lung (pulmonary) arteries. If the PDA is large, the excess blood being pumped into the lung arteries makes the heart and lungs work hard and the lungs becomes congested.

If PDA is small, it will not cause any symptoms because the heart and lungs don't have to work hard. The only unusual finding may be distinctive type of murmur. Whereas if the PDA

is large, the child may breathe faster and harder than normal. As a result high blood pressure may occur. Over time this may cause permanent damage to the lung blood vessels.If the PDA (ductus) is small, it doesn't make the heart and lungs work harder. Surgery and other treatments may not be needed. One good thing about Small PDAs is they often close on their own within the first few months of life.

Normally the heart's left side only pumps blood to the body, and the right side only pumps blood to the lungs. Extra blood gets pumped from the body artery (aorta) into the lung (pulmonary) arteries in a person who is suffering with PDA. If the PDA is larger than usual, the extra amount of blood which is being pumped abundously into the lung arteries makes the heart and lungs work harder and there is a chance that lungs can become congested.

Surgery may be the best treatment option for patients suffering with PDA. The surgeon doesn't have the need to open the heart to fix or cure the PDA. An incision is made in the left side of the chest, between the ribs. The best option to perform repair on PDA is to closing it by tying it with suture (thread-like material) or by permanently placing a small metal clip around the PDA to squeeze it closed. Occasionally in the adult, a surgical patch is used. If there's no other heart defect, this restores the circulation to normal.

We report the case of a 23-year old Filipino female with orofacial cleft, presenting with symptoms of decompensated heart failure. Clinical examination revealed the presence of diastolic murmur, 5/6, with accentuated pulmonary component of the second heart sound. Cardiomegaly was seen on chest x-ray and right ventricular hypertrophy on electrocardiogram. On performing Transthoracic echocardiogram (TTE), it shows Cor triatrium sinistrum; dilated left atrium divided by a membrane into a proximal and distal chamber; dilated right atrium; dilated right ventricle with right ventricular hypertrophy; dilated main pulmonary artery; and severe pulmonic regurgitation with severe pulmonary hypertension.

The TTE perfomed should be able to provide accurate and sufficient characterization of cardiac anatomy for optimal diagnosis but because of the rarity of the disease, it can be missed. A transesophageal echocardiography (TEE) provides excellent resolution and offers invaluable assistance to surgical intervention. TEE was done upon follow up to rule out associated congenital abnormalities which only showed patent ductus arteriosus.

Surgery provides a satisfactory early and long term survival. Therefore the key is the accurate evaluation of cor triatriatum. Our report highlights early diagnosis and the utility of transesophageal echocardiography in the diagnosis of such congenital cardiac abnormalities and associated lesions.

This work is partly presented at 8th World Heart Congress on May 11-12, 2020