

Cardiac diverticulum

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Abstract

Cardiac diverticulum is a very rare congenital malformation that may be either fibrous or muscular. Fibrous diverticulum refers to aneurysm and usually appears as an isolated anomaly. Patients may be asymptomatic. Muscular diverticulum of the ventricles is characterized by a muscular appendix emerging from the left ventricular apex, rarely from the right ventricle or from both chambers. They are frequently associated with other congenital anomalies. Because of the association with major midline abdominal anomalies, cardiac muscular diverticulum is most often recognized in infants and only rarely as an incidental finding in older children or adults. Diverticula usually appear in the newborn period as a pulsating umbilical mass protruding through defects in the pericardium, diaphragm and midline abdominal wall. Natural history of the disease is severe and most patients die in the first year of life especially when associated with severe heart malformations. Surgical resection is indicated as soon as the diagnosis of congenital ventricular diverticulum is made.

Keywords

Cardiac diverticulum - fibrous diverticulum - muscular diverticulum - Cantrell syndrome.

Disease name and synonyms

- Cardiac diverticulum
- Ventricular diverticulum

European pediatric cardiac code

Reference of "Cardiac diverticulum" is 07.06.03.

Definition

Cardiac diverticulum is a congenital malformation that may be either fibrous or muscular (1).

The terms "fibrous diverticulum" or "congenital aneurysm" refer to aneurysm whose etiology is unknown. It usually appears as an isolated anomaly (1, 2).

Frequency

Cardiac diverticulum is very rare, no data on incidence or prevalence is available.

Main symptoms

Fibrous diverticulum

The cardiac wall is constituted mostly of fibrous tissue with few or no muscular fibers. Its connection with the ventricular cavity is wide. It is commonly located at the apex or the base of the left ventricle (1). Patients with fibrous diverticulum may be asymptomatic.

When located at the apex, it may calcify or form thrombi which may lead to embolic accidents.

When located at the mitral ring it may cause mitral incompetence (1).

Muscular diverticulum

Muscular diverticulum of the ventricles is a rare anomaly first described by O'Bryan in 1837. It is characterized by a muscular appendix emerging from the left ventricular apex, rarely from the right ventricle or from both chambers. The diverticular musculature has mechanical activity synchronous with the activity of the ventricles. Its connection to the ventricular chamber is usually narrow (1, 2).

Muscular diverticulum is frequently associated with other congenital anomalies including those of the abdominal wall, diaphragm, sternum, pericardium and the heart itself (usually ventricular septal defect).

The association of a ventricular diverticulum with a midline supra-umbilical abdominal wall defect, a defect of the lower part of the sternum, a deficiency of the anterior diaphragm, a defect of the diaphragmatic pericardium and a congenital heart malformation constitutes the Cantrell syndrome (3).

Because of the association with major midline abdominal anomalies, cardiac muscular diverticulum is most often recognized in infants and only rarely as an incidental finding in older children or adults. Diverticulum usually appears in the newborn period as a pulsating umbilical mass protruding through defects in the pericardium, diaphragm and midline abdominal wall. It is important to differentiate this entity from the ectopia cordis in which no cardiac silhouette is found in the thorax of the chest roentgenograms. A true diverticulum may protrude beyond the thorax but the basic cardiac position remains normal (2).

Natural history

Natural history of the disease is severe and most patients die in the first year of life especially when associated with severe heart malformations. Specific causes of death of the diverticulum are rupture, heart failure and tachyarrhythmias.

Diagnostic methods

Fibrous diverticulum

The defect may be discovered only from alterations of the cardiac silhouette on chest x-rays or through modifications of ST segment on ECG similar to non congenital ventricular aneurysms.

Muscular diverticulum

It is usually diagnosed by echocardiography.

Management

Surgical resection is indicated as soon as the diagnosis of congenital ventricular diverticulum has been made because of the risk described above (4).

References

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