*Cardiology in the Young*

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# Congenital left ventricular aneurysm of interventricular septum: prenatal diagnosis and long-term management

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**Abstract**

Congenital left ventricular aneurysm is a rare anomaly consisting of an extroflection from left ventricular walls with a wide connection. Most left ventricular aneurysms are asymptomatic and diagnosed as incidental finding, but can be associated with complications during follow-up. We present a case of congenital left ventricular aneurysm at the level of the interventricular septum with secondary remodelling of the left ventricle.

**Brief Report**

**Cite this article:** Cavalla F and Cipriani A (2022) Congenital left ventricular aneurysm of interventricular septum: prenatal diagnosis and long-term management. *Cardiology in the Young* **32**: 2036–2037. doi: 10.1017/S1047951122001032

Received: 29 December 2021
Revised: 21 February 2022
Accepted: 9 March 2022
First published online: 31 March 2022

**Keywords:**
Left ventricular aneurysm; prenatal diagnosis; CHD

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Congenital left ventricular aneurysm is a rare cardiac anomaly that consists of a thin-walled dyskinetic or akinetic structure with a wide connection to the left ventricle.[1]

Prevalence of left ventricular aneurysms is 0.04% in the general population and 0.02% in CHD.[2]

Diagnosis can be made after exclusion of coronary artery disease, local or systemic inflammation or traumatic causes as well as cardiomyopathies.[2]

Its origin could be partially explained by an embryonic anomaly, starting in the fourth week.

Gembruch and colleagues firstly described congenital left ventricular aneurysm as a prenatal finding.[3]

Clinical presentation, natural history, and management are not well defined, because of the rarity of this malformation. The improvement of echocardiography has led to an increased frequency of congenital left ventricular aneurysm prenatal detection.[4] Most left ventricular aneurysms are localised at the apex of the left ventricle (61.5%). Other locations include anterolateral (15%), lateral (8%), septal (8%), posterolateral (4%), and inferior (4%) segments of the left ventricle.[1]

Most left ventricular aneurysms are asymptomatic and diagnosed as incidental finding. In some cases, left ventricular aneurysms are associated with systemic embolisation, heart failure, valvular regurgitation, ventricular wall rupture, ventricular tachycardia, or sudden cardiac death. Diagnosis is established by imaging studies such as echocardiography, magnetic resonance, or left ventricular angiography.

Treatment must be tailored on clinical presentation and includes conservative management, surgical resection, anticoagulation after systemic embolisation, radiofrequency ablation, implantable cardioverter defibrillator, and antiarrhythmic drugs.[2]

## Case report

In our patient, fetal echocardiography at 20 weeks of gestation revealed a left ventricular aneurysm at the level of the septum as an incidental finding.

After his birth, a secondary cause of left ventricular aneurysm was excluded and the patient was diagnosed with congenital left ventricular aneurysm. His left ventricle showed a secondary remodelling with moderate dilation, but preserved ejection fraction.

In particular, the aortography showed a normal anatomy of the coronary arteries with right coronary artery dominance. Cardiomyopathies and inflammatory diseases were not suspected. Myocardium appeared normal apart from the aneurismatic region. Additionally, mother and baby were both tested negative for viral infections (Parvovirus B19 and others).

Our patient had a normal psychomotor development and regular height and weight growth.

He was asymptomatic with good exercise capacity and without the need of therapies.

He was prescribed aspirin and started his follow-up visits twice a year until 7 years old, then once a year. Between 12 and 15 years old, our patient was checked up twice a year, as regards his linear and ponderal growth.

The boy was evaluated with echocardiogram (biannually), with 24-hour Holter monitoring (annually) and with cardiac magnetic resonance (every 3 years).

He also performed an exercise stress test depending on his physical activity.

At the age of 15 years old, he was globally re-evaluated.

The ECG showed sinus rhythm and left axis deviation.

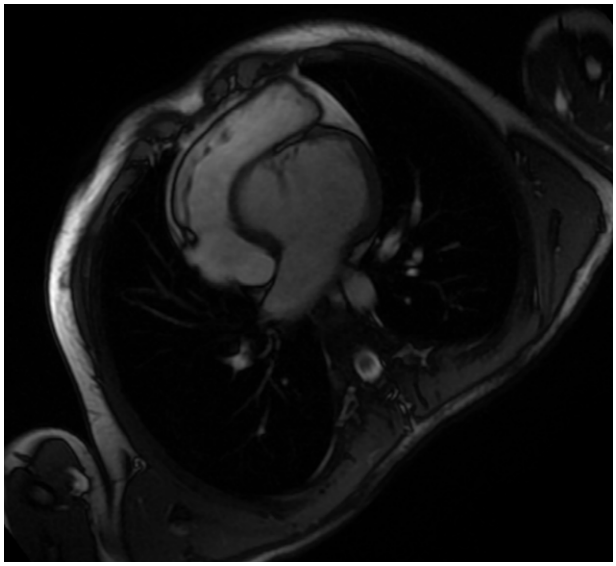

**Figure 1.** CMR image of left ventricular aneurysm. Diastolic phase.

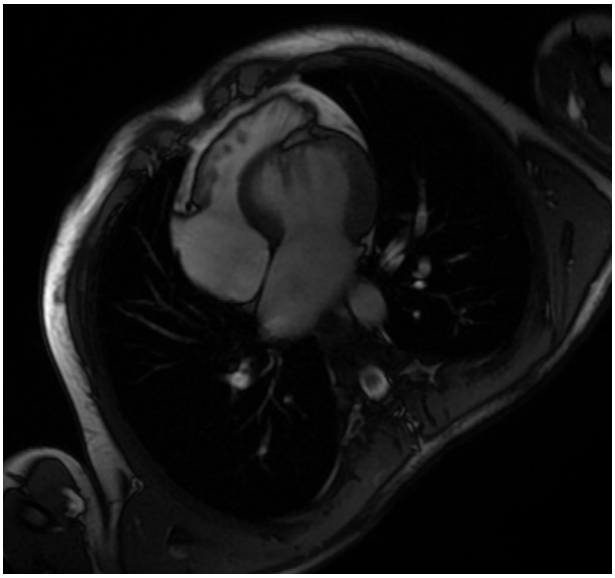

**Figure 2.** CMR image of left ventricular aneurysm. Systolic phase.

A wide septal aneurysm (5.5× 3 cm), at the level of its medial region, was clearly seen at echocardiography. The wall of the left ventricular aneurysm was thin, with reduced contractility. No thrombus was noted inside the left ventricular aneurysm. Left ventricle was therefore dilated with a diastolic diameter of 61 mm. Apart from the left ventricular aneurysm, the other regions of the left ventricle were hyperkynetic with normal wall thickness. The estimated ejection fraction was 50%. The diastolic function was mildly reduced. Other echographic parameters were normal; in particular, mitral and tricuspid valve morphology and function was normal.

He performed an exercise stress test, with normal findings.

A 24-hour Holter monitoring excluded the presence of arrhythmias.

He underwent a left ventricle angiography. The wide septal aneurysm determined a re-shape of the left ventricle with a prevalence of the transversal diameter compared to the longitudinal one. There was reduction of contractility at the level of left ventricular aneurysm. Global systolic left ventricular function appeared good, and diastolic function was mildly impaired (figure S1, S2). Left ventricular protodiastolic pressure was 0 mmHg, and telediastolic pressure was 20 mmHg. The good performance of the mitral valve was confirmed. Pulmonary artery pressures were normal, as well as the end-diastolic right ventricular function.

The aortography confirmed the normal origin and course of the coronary arteries. The left descending artery presented a reduction of its distal calibre, but with good extension to periphery.

To complete the evaluation, our young patient performed a cardiac magnetic resonance: the left ventricle appeared globose with preserved ejection fraction and without evidence of delayed myocardial enhancement (Figs 1 and 2).

## Conclusions

Congenital left ventricular aneurysms are rare cardiac anomalies that originate during the fetal period. They are often asymptomatic, and their management of left ventricular aneurysms depends on clinical presentation, symptoms, and complications.

In our case report, our young patient was totally asymptomatic. We decided on conservative management and follow-up.

Exams showed mild left ventricular diastolic dysfunction, most likely due to secondary left ventricular remodelling. The left ventricular aneurysm homogeneously increased its dimension, likewise the growth of our young patient, without changes in its shape.

Long-term follow-up is mandatory, because the dilation of left ventricle could progressively increase, leading to worsening of systolic and diastolic dysfunction until heart failure.

**Supplementary material.** To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951122001032

**Acknowledgements.** None.

**Financial support.** This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

**Conflicts of interest.** None.

**Ethical standards.** The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation (Ethical guidelines for biomedical research on human participants 2006, India) and with the Helsinki Declaration of 1975, as revised in 2008, and have been approved by the institutional committees.

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