

Isolated foetal left ventricular apical hypoplasia: would you know how to spot it?

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Brief Report

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Abstract

Isolated left ventricular apical hypoplasia is a rare cardiac malformation. This entity is easily diagnosed if its key features are recognised, but will puzzle the clinician who is not aware of its existence. It is characterised by echocardiographic findings of a spherical left ventricle, an elongated right ventricle wrapping around the deficient left ventricular apex, and abnormal papillary muscles' origin.

Case report

A 37-year-old woman underwent routine obstetric ultrasound examination at 20 weeks' gestation at our institution. She had consulted elsewhere and had been given a poor prognosis for her pregnancy, as the foetus showed an abnormally shaped left ventricle and a hyperechogenic endocardium, and endocardial fibroelastosis was suspected. As our team had previously diagnosed two patients with this malformation,¹ we were familiar with the key features of Isolated left ventricular apical hypoplasia.

Both ventricles showed a normal contractile function. Heart rhythm was normal. A specific feature (not reported in older patients) found in this foetus was a hyperechogenic endocardium restricted to the interventricular septum. This characteristic became gradually less evident as the pregnancy evolved. After discussing with the parents the suspected diagnosis and the evidence shown in the literature about possible outcomes, the pregnancy was continued, and an asymptomatic 3720 g girl was delivered at 40 weeks of gestation. Follow up showed absence of symptoms, with good weight gain and no other health issues. Close cardiological follow-up with serial echocardiographic evaluations revealed a systolic left heart function in the lower limit, with an left ventricle ejection fraction of 55%. Other echocardiographic parameters indicated a preserved left ventricular function (biphasic mitral inflow, normal inflow from left ventricle to the ascending aorta and aortic arch, and right-left flow through the foramen ovale). She is currently asymptomatic at one year of age.

Discussion

First described in 2004,² and although being a very rare heart malformation, an increasing number of case reports are being published^{3,4} in recent years in adults and children, as clinicians are getting aware of its existence.

In the adult, symptoms can range from the asymptomatic patient to patients with heart failure and rhythm anomalies, such as atrial fibrillation.^{3–5} Paediatric cases have also been reported.^{1,3–6}

Foetal reports of this malformation are exceptional, possibly in part because of lack of diffusion of the features of this rare heart malformation among obstetricians, or because it might evolve during pregnancy.

Detailed description of the anatomy of the normal foetal heart and its correlation with prenatal echocardiography can be found in the literature.⁷

In the normal heart, the ventricular apex is formed by the left ventricle (Figure 1). In contrast to the normal heart, in isolated left ventricular apical hypoplasia, the apex of the heart will not be formed by the left ventricle, but by the apex of the right ventricle (Figure 2). The foetal ultrasound will include key echocardiographic features such as an elongated right ventricle wrapping around the deficient left ventricular apex, a spherical left ventricle, and abnormal papillary muscles' origin from the left ventricular apex.⁵ We add a specific feature in the foetus, not described in post-natal life: a hyperechogenic interventricular septum (Figure 2).

Differential diagnosis should include hypoplastic left heart syndrome, endocardial fibroelastosis,⁸ and aneurysm of the left ventricle. To look for the wrapping of the right

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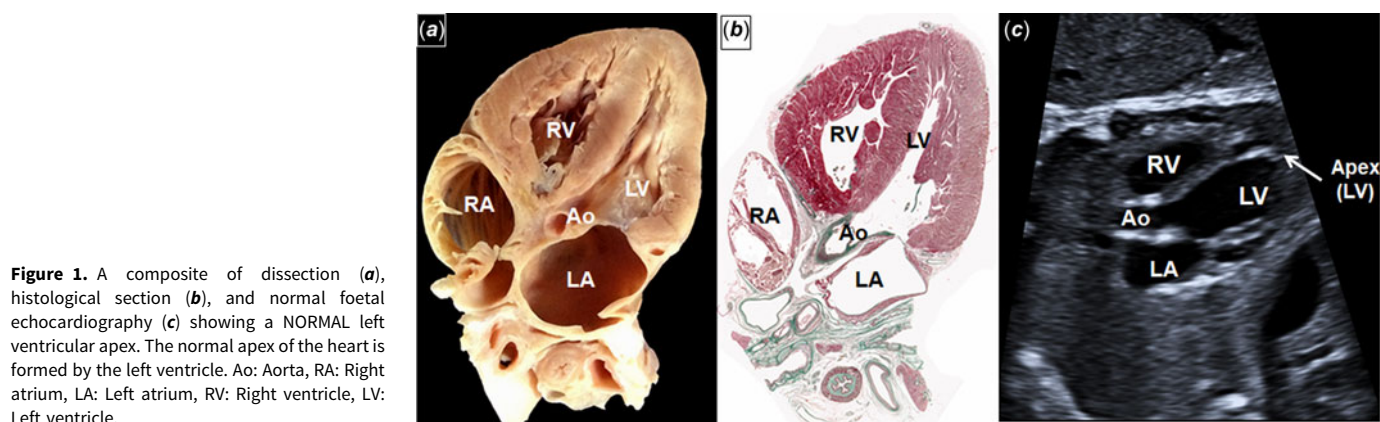


Figure 1. A composite of dissection (a), histological section (b), and normal foetal echocardiography (c) showing a NORMAL left ventricular apex. The normal apex of the heart is formed by the left ventricle. Ao: Aorta, RA: Right atrium, LA: Left atrium, RV: Right ventricle, LV: Left ventricle.

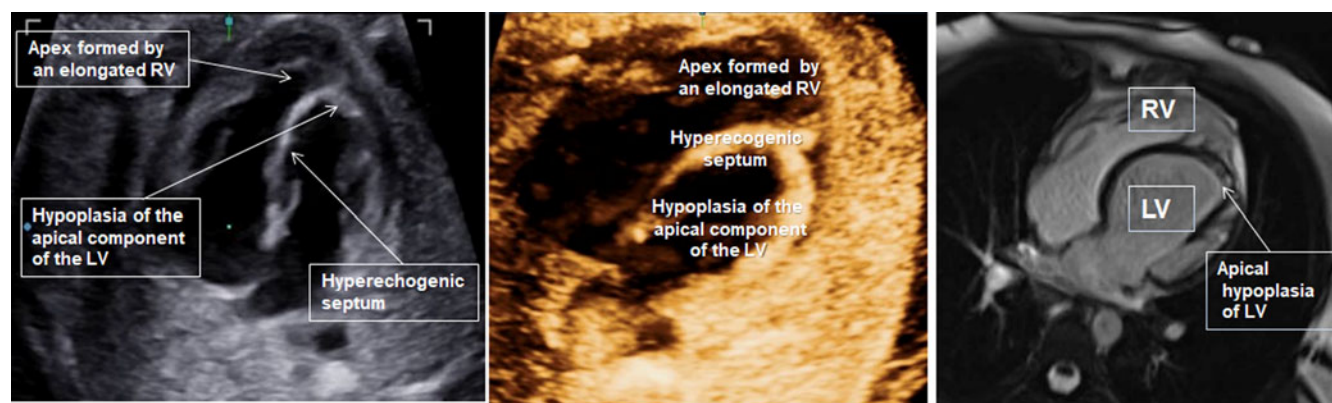


Figure 2. To compare with Figure 1, here is depicted an ABNORMAL left ventricular apex with hypoplasia of the apical component of the LV, the apex of the heart being formed by the elongated right ventricle that wraps around the left ventricle, and the feature we add to the previously described, a hyperechogenic interventricular septum as seen in the foetal scan (left). The apex of the heart in Isolated Left Ventricular Apical Hypoplasia (ILVAH) is formed by the right ventricle. These features can be enhanced by 3D Echocardiography (centre) and cardiac MRI (right).

apex around the missing left apex (Figure 2) and the hyperechogenic septum will be two valuable clues for the diagnosis in the foetus.

Information to the family needs to include the fact that it is a rare and not well-known heart malformation with an uncertain evolution, which frequently goes unnoticed, and that most cases described in the literature in young people remain asymptomatic, with symptoms such as heart failure and rhythm disturbances developing in adulthood, generally responding to medical treatment.

A prenatal consult with a paediatric cardiologist is advised, and follow up during childhood needs to be scheduled. Cardiac MRI (Figure 2) is becoming a valuable tool in order to follow up the ventricular function in these patients, and it offers additional information, such as the imaging of fatty tissue at the hypoplastic apex.⁹

The aim of this paper is to draw attention to this abnormal development of the left ventricle, as the use of foetal echocardiography for screening expands worldwide.

Many of us have learned from our teachers and mentors in medical school that “the eyes cannot see what the mind does not know.” Now we know.

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Competing interests. The authors declare none.

Ethical standard. The authors confirm that consent for submission and publication of this case report has been obtained from the patient.

Twitter. hashtag PedsCards. Suggested tweet: ILVAH: how to spot it.

References

1. Ruiz Martín P, Ortiz Garrido A, Zabala Argüelles JI, Urbano Carrillo CA. Hipoplasia aislada del ápex del ventrículo izquierdo. Isolated left ventricular apex hypoplasia. REC Cardiol Clinics 2023; 58: 64–65.
2. Fernandez-Valls M, Srichai B, Stillman AE, White RD. Isolated left ventricular apical hypoplasia: a new congenital anomaly described with cardiac tomography. Heart 2004; 90: 552–555.
3. Liao S, Jin P, Miao R, Huang X. Multimodality imaging for the diagnosis of left ventricular apical hypoplasia: a case presentation and review of the literatures. Echocardiography 2024; 41: e15793.

4. Bassareo PP, Duignan S, James A, Dunne E, McMahon CJ, Walsh KP. Isolated left ventricular apical hypoplasia: systematic review and analysis of the 37 cases reported so far. *World J Clin Cases* 2023; 11: 5494–5503.
5. Román R, Anchante H, Menacho K, Medina F. Isolated left ventricular apical hypoplasia: case report. *Eur Heart J Case Rep* 2023; 7: 1–8.
6. Marin C, Sanchez ML, Maroto E, Ossaba S, Ruiz Y, Zabala JI. MR imaging of isolated left ventricular apical hypoplasia. *Pediatr Radiol* 2007; 37: 703–705.
7. Picazo-Angelin B, Zabala-Argüelles JI, Anderson RH, Sánchez-Quintana D. Anatomy of the normal fetal heart: the basis for understanding fetal echocardiography. *Ann Pediatr Cardiol* 2018; 11: 164–173.
8. Yan H, Zhou K, Zhang Z, et al. Approach an appropriate decision on fetus with endocardial fibroelastosis in collaboration with cardiovascular profile score. A case report. *Medicine* 2016; 95: e4124.
9. Meléndez G, Muñoz L, Meave A. Hipoplasia de la porción apical del ventrículo izquierdo. *Rev Esp Cardiol* 2010; 63: 984.