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An Unexpected Case of Isolated Left Ventricular Apical Hypoplasia in an Asymptomatic Patient

Rachel Anne Xuereb[1], Sara Xuereb[2], Tiziana Felice[2], Mariosa Xuereb[2], Alex Borg[2]

1. Department of Medicine, Basic Specialist Trainee, Mater Dei Hospital, Msida, Malta
2. Department of Cardiology, Mater Dei Hospital, Msida, Malta

Corresponding author:
Rachel Anne Xuereb
Email: rachel.xuereb94@gmail.com
Mater Dei Hospital, Triq Dun Karm, Msida, Malta, MSD 2090.

Highlights

Isolated left ventricular (LV) apical hypoplasia is a rare type of congenital heart disease. It is often asymptomatic in childhood but may lead to complications later on in life. The proposed mechanism is inadequate LV dilatation during development of the primitive ventricle, resulting in a spherical LV. To our knowledge, we describe for the first time a case of isolated LV apical hypoplasia in an asymptomatic patient, diagnosed on investigation of an abnormal 12-lead resting electrocardiogram (ECG).

Keywords: Isolated left ventricular apical hypoplasia; Cardiac MRI; Congenital; Rare; Asymptomatic

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LV apical hypoplasia is characterised by the presence of the following findings: a spherical LV with rightward bulging of the interventricular septum, deficiency of the myocardium within the LV apex with adipose tissue infiltrating the apex, papillary muscle originating from the anterior apex and an elongated right ventricle (RV) wrapping around the deficient LV apex [1], [2]. 25 cases have been reported in the literature so far. We report the first case of an asymptomatic patient with isolated LV apical hypoplasia, diagnosed on Cardiac Magnetic Resonance Imaging (MRI).

Case report

A 22 year old Caucasian male competitive rugby player presented for a routine preoperative assessment prior to an orthopaedic procedure. He denied any shortness of breath, dizzy spells, syncopal events, chest pain or palpitations and was previously healthy with no significant family history. Clinical examination was unremarkable. However, a 12-lead resting ECG showed bifascicular block with right axis deviation and a right bundle brunch block pattern. A 7-day ambulatory ECG (Holter) monitor and exercise stress test were normal.

Transthoracic echocardiography revealed an elongated RV, wrapping around a spherically shaped LV, with the apex formed by the RV, Figure 1. Cardiac MRI showed a spherical LV cavity and a prominent fat pad at the LV apex, partially infiltrating the thinned apical myocardium, Figure 2A, B, C. Late enhancement pulse sequences post-gadolinium injection showed no specific high signal, Figure 2D. LV cavity volumes, myocardial mass and ejection fraction were normal. A conservative approach to management is being followed with yearly Holter monitoring to assess for arrhythmias and cardiac MRI to assess ejection fraction and size of both ventricles.

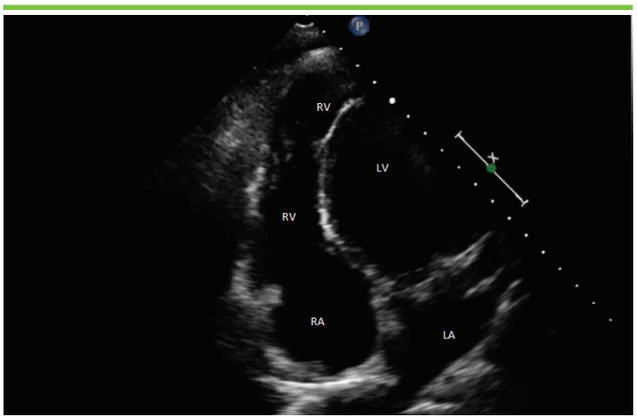

Figure 1.
Transthoracic echocardiogram apical four-chamber view. Elongated RV wrapping around spherically shaped LV. Apex is formed by the RV. RA: right atrium, RV: right ventricle, LA: left atrium, LV: left ventricle

Discussion

Isolated LV apical hypoplasia was first described in 2004 [3]. Most cases have been described among Caucasian populations. To date 25 cases have been reported in the literature, with a varied age range and presentation, summarised in Table 1

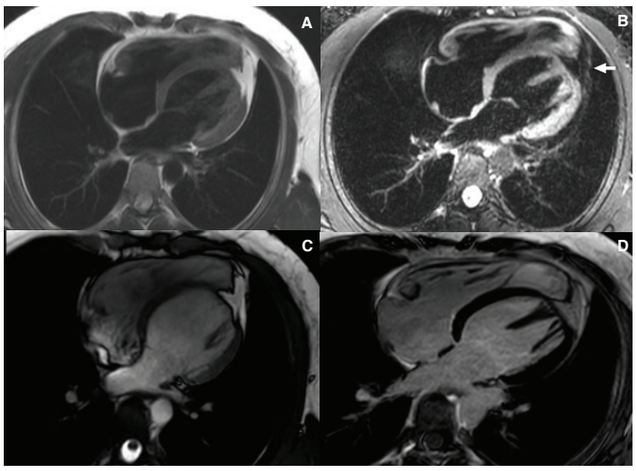

Figure 2.
(A) T2 weighted turbospin echo pulse sequence, (B) STIR (short tau inversion recovery) pulse sequence with incorporated fat suppression, (C) SSFP (steady-state free precession) cine imaging, all in four-chamber view. Note prominent layer of fat overlying, and infiltrating into the thinned LV apex (arrowhead). (D) Late enhancement acquisition post-gadolinium injection. There is no high signal suggest of macroscopic fibrosis.

Table 1: Summary of literature review

	Total
Age	3 months to 66 years Mean 29 years
Gender	10 male, 11 female 4 unspecified
Presentation	9 atypical chest discomfort 6 murmur 3 dyspnoea 2 arrhythmias 1 prenatal imaging 1 unspecified
Outcome	3 no complications 2 controlled medically 1 atrial fibrillation 1 sudden cardiac death 18 unspecified
Associations	1 patent ductus arteriosus 1 infundibular pulmonary stenosis and aortic stenosis 1 LV diverticulum 1 right ventricular tract outflow obstruction 1 mutation of lamin A/C gene

[1-10]. It is often asymptomatic in childhood, however it may cause pulmonary oedema, pulmonary hypertension, atrial and ventricular tachyarrhythmias in adults.

Symptomatic patients may need treatment for heart failure, cardiac arrhythmias and even cardiac transplantation. Differential diagnosis of isolated LV apical hypoplasia include arrhythmogenic cardiomyopathy which is characterised by fibrofatty replacement of the LV myocardium, hypoplastic left heart syndrome, LV non-compaction cardiomyopathy and congenital LV aneurysm [10].

Conclusions

Isolated LV apical hypoplasia is a unique, presumably congenital, cardiac anomaly that is important to recognise, with potentially serious complications. Cardiac MRI is useful for diagnosis and follow up.

Declarations of Interest

The authors declare no conflicts of interest.

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