

# Apical hypertrophic cardiomyopathy: an uncommon cause of exertional chest pain in children

## Brief Report

**Cite this article:** Zhao Q, He L, and Liu F (2019) Apical hypertrophic cardiomyopathy: an uncommon cause of exertional chest pain in children. *Cardiology in the Young* 29: 1533–1535. doi: [10.1017/S1047951119002403](https://doi.org/10.1017/S1047951119002403)



Received: 20 July 2019  
Revised: 11 August 2019  
Accepted: 3 September 2019  
First published online: 30 October 2019

### Keywords:

Apical hypertrophic cardiomyopathy; chest pain; electrocardiogram

### Author for correspondence:

F. Liu, MD, Pediatric Heart Center, Children's Hospital of Fudan University, 399 Wan Yuan Road, Shanghai 201102, People's Republic of China. Tel: 86-21-64932800; Fax: 86-21-64931002; E-mail: [liufang@fudan.edu.cn](mailto:liufang@fudan.edu.cn)

Qu-ming Zhao , Lan He  and Fang Liu

Pediatric Heart Center, Children's Hospital of Fudan University, Shanghai, People's Republic of China

### Abstract

Apical hypertrophic cardiomyopathy is an uncommon morphologic variant of hypertrophic cardiomyopathy, which is rarely diagnosed in childhood. To date, very few cases of asymptomatic children younger than 18 years have been reported in the literature. To the best of our knowledge, this is the first case of paediatric apical hypertrophic cardiomyopathy presenting with exertional chest pain, with characteristic electrocardiographic, echocardiographic, MRI, and cardiac angiography findings.

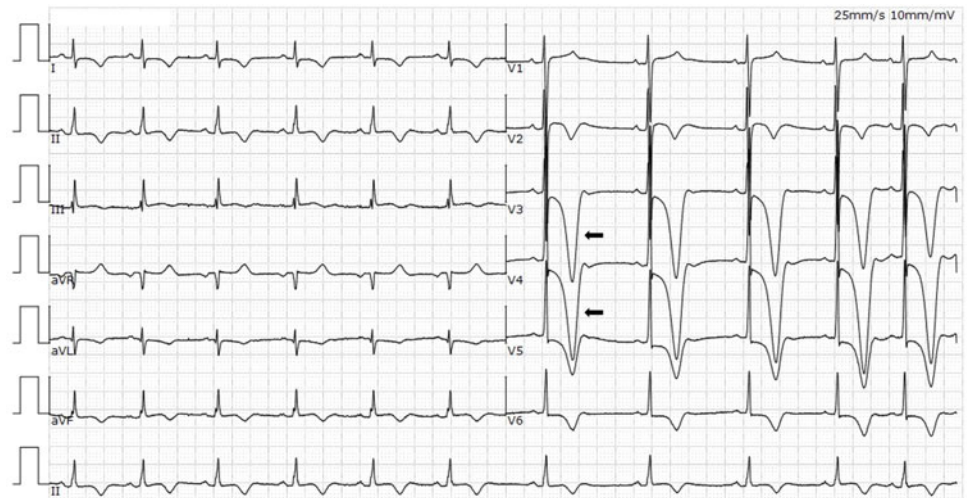
Apical hypertrophic cardiomyopathy is an uncommon morphologic variant of hypertrophic cardiomyopathy that predominantly affects the apex of the left ventricle, characterised by giant negative T waves on the electrocardiogram and spade-shaped left ventricle cavity.<sup>1</sup> Since it is typically diagnosed in midlife, the vast majority of current reports of apical hypertrophic cardiomyopathy are confined to adults,<sup>2–4</sup> with very few cases in children,<sup>5</sup> all of whom were asymptomatic and detected incidentally due to abnormal electrocardiogram findings at a routine health check-up. We present a school-age boy who presented with exertional chest pain found to have apical hypertrophic cardiomyopathy.

### Case report

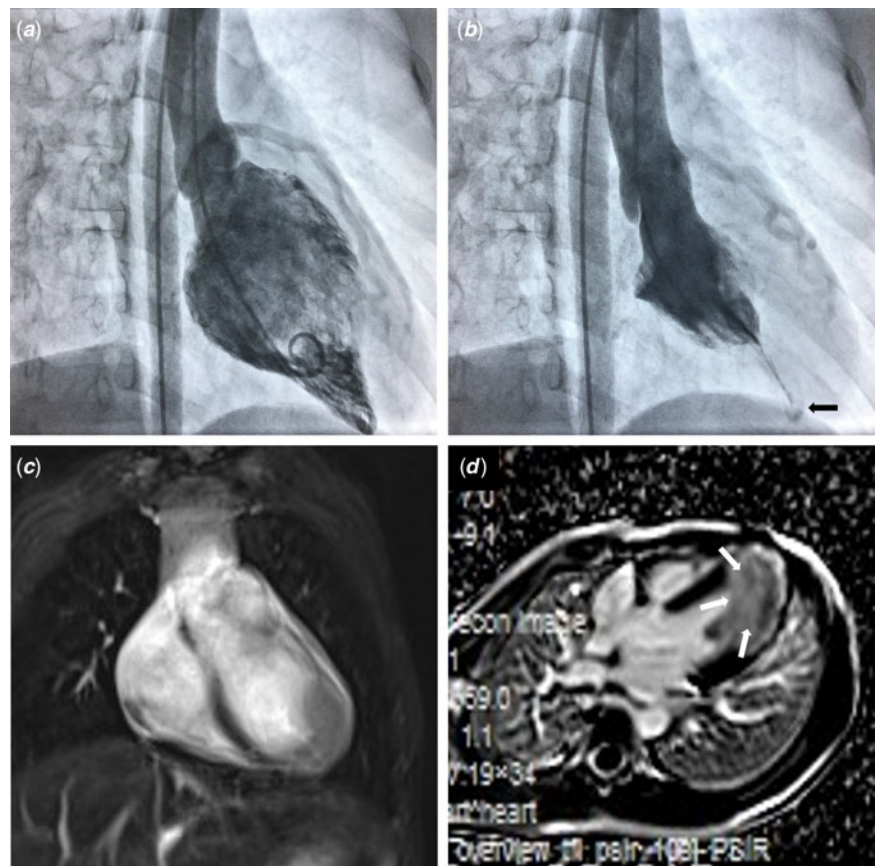
A 9-year-old boy was admitted to our hospital secondary to a 2-week history of chest pain. The chest pain localised to the left chest which was precipitated by exertion, but recently occurred more frequently and occurred with both with activity and rest. The patient denied any chest trauma, he had not experienced syncope and had no family history of cardiac disease. Physical examination was within normal limits. Laboratory data revealed a normal complete blood count, biochemical parameters, creatine kinase, brain natriuretic peptide, and cardiac troponin I. Chest radiograph was negative. Electrocardiogram revealed constant giant negative T waves with ST depression in the anterior and lateral leads (Fig 1), which was unrelated to the presence of chest pain and showed no resolution in the next few days. However, the initial echocardiography did not suggest cardiomyopathy or coronary artery abnormalities.

Cardiac angiography was performed to further evaluate cardiac causes of chest pain and abnormal electrocardiogram. Coronary angiography demonstrated enlarged epicardial left coronary artery without focal stenosis and myocardial bridging. Left ventricle angiography was suggestive of a spade-like appearance, which is characteristic of apical hypertrophic cardiomyopathy (Fig 2). MRI confirmed the findings, and large areas of delayed enhancement were consistent with apical fibrosis (Fig 2). After focusing on the myocardium of left ventricle apex, a second echocardiography revealed a progressive increase in left ventricle wall thickness towards the apex without evidence of outflow tract or midventricular obstruction. Left ventricle end-diastolic dimension at the level of the mitral valve leaflet tips was 40 mm with an ejection fraction of 68%. The end-diastolic thickness of left ventricle apical and basal free wall was 17 and 8.5 mm, respectively. The end-diastolic thickness of apical and basal interventricular septum was 12 and 8.2 mm, respectively. Continuous electrocardiogram monitoring during hospitalisation found no significant arrhythmias and a 24-hour Holter monitor only showed occasional uniform premature ventricular contractions. His parents also underwent echocardiography and electrocardiogram, both of which were normal.

Though he had not yet undergone confirmatory genetic testing, the presence of apex-predominant left ventricle hypertrophy with an ace-of-spades configuration of the left ventricle cavity, and giant negative T waves, strongly suggested apical hypertrophic cardiomyopathy as the aetiology of his chest pain. On admission day seven, our patient was discharged home on metoprolol, and he remained symptom-free at 1-month follow-up.



**Figure 1.** Electrocardiogram shows a loss of septal Q waves, high QRS voltage, and repolarisation abnormalities with deep T wave inversion, especially in leads V3 and V4 (black arrow). Such abnormalities are also evident in inferior and lateral leads.



**Figure 2.** Left ventricle angiography in the right anterior oblique view and cardiac MRI. (a) The typical spade-like appearance at end-diastole. (b) During end-systole, a small cavity in the apical portion (black arrow) was separated from the basal cavity by long cavity obliteration. (c,d) Cardiac MRI with gadolinium enhancement demonstrating focal apical hypertrophy and fibrosis (white arrows).

## Discussion

Since apical hypertrophic cardiomyopathy was first reported in Japan 40 years ago, a growing number of literatures have described its clinical features,<sup>1</sup> helping clinicians recognise that apical hypertrophic cardiomyopathy may be a commonly missed or delayed diagnosis, especially when patients present with acute chest pain and an abnormal electrocardiogram.

Given the rarity of apical hypertrophic cardiomyopathy diagnosed in childhood and the fact that apical imaging is not an advantage of echocardiography, a paediatric cardiologist would

first consider anomalous coronary artery origin or coronary ischemia when the patients presented with chest pain and an abnormal electrocardiogram but no indication of cardiomyopathy at the initial echocardiography.<sup>6</sup> After a careful review of literature, we found only four cases of apical hypertrophic cardiomyopathy in children younger than 18 years who were asymptomatic,<sup>5,7</sup> and no paediatric cases were reported in other apical hypertrophic cardiomyopathy studies.<sup>2-4</sup> Therefore, to the best of our knowledge, this is the first reported case of apical hypertrophic cardiomyopathy presenting with chest pain in children.

The trademark electrocardiogram finding in apical hypertrophic cardiomyopathy is giant negative T waves ( $\geq 10$  mm) in the left precordial leads associated with high QRS voltage, which were found more commonly in Japanese patients (64%) as compared with the United States of America (30%).<sup>1</sup> Myocardial ischemia can also be manifested as ST-T abnormalities, but ST-segment depression and T wave inversion are generally concordant to low-voltage QRS complex.<sup>8</sup> Though Sakamoto summarised 10 years of clinical data and found no giant negative T in children younger than 15 years,<sup>9</sup> we now know that this is not the case, and that characteristic electrocardiogram findings are sometimes more sensitive than echocardiography for diagnosing apical hypertrophic cardiomyopathy. MRI may facilitate a more comprehensive evaluation of hypertrophic segments and myocardial fibrosis.

Similar to classic hypertrophic cardiomyopathy, current studies have demonstrated a familial association with apical hypertrophic cardiomyopathy including an autosomal dominant inheritance pattern. Several hundred mutations have been identified, scattered among at least 27 hypertrophic cardiomyopathy susceptibility genes encoding various sarcomeric and non-sarcomeric proteins. Generally, the apical hypertrophic cardiomyopathy was most commonly associated with mutations in MYBPC3, MYH7, ACTC1, and TPM1.<sup>1</sup>

We hope that this case will enhance paediatrician's awareness of the characteristic electrocardiogram and imaging findings of apical hypertrophic cardiomyopathy and help in the early identification of this rare cause of paediatric chest pain.

**Acknowledgements.** The authors thank their colleagues in radiology department for the exquisite cardiac MRI images.

**Financial Support.** Key Developing Discipline of Shanghai Municipal Commission of Health and Family Planning (Pediatrics) 2016ZB0101.

**Conflict 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 and with the Helsinki Declaration of 1975, as revised in 2008, and were approved by the Ethics Committee of the Children's Hospital of Fudan University.

## References

1. Jan MF, Todaro MC, Oreto L, Tajik AJ. Apical hypertrophic cardiomyopathy: present status. *Int J Cardiol* 2016; 222: 745–759.
2. Zhai SS, Fan CM, An SY, et al. Clinical outcomes of myocardial bridging versus no myocardial bridging in patients with apical hypertrophic cardiomyopathy. *Cardiology* 2018; 139: 161–168.
3. Klarich KW, Attenhofer Jost CH, Binder J, et al. Risk of death in long-term follow-up of patients with apical hypertrophic cardiomyopathy. *Am J Cardiol* 2013; 111: 1784–1791.
4. Moro E, D'Angelo G, Nicolosi GL, Mimo R, Zanuttini D. Long-term evaluation of patients with apical hypertrophic cardiomyopathy. Correlation between quantitative echocardiographic assessment of apical hypertrophy and clinical-electrocardiographic findings. *Eur Heart J* 1995; 16: 210–217.
5. Miyamoto T, Horigome H, Kawano S, Sumazaki R. Apical hypertrophic cardiomyopathy in childhood: a long-term follow-up report of two cases. *Pediatr Cardiol* 2009; 30: 343–346.
6. Friedman KG, Alexander ME. Chest pain and syncope in children: a practical approach to the diagnosis of cardiac disease. *J Pediatr* 2013; 163: 896–901; e891–893.
7. Chaowu Y, Shihua Z, Jian L, Li L, Wei F. Cardiovascular magnetic resonance characteristics in children with hypertrophic cardiomyopathy. *Circ Heart Fail* 2013; 6: 1013–1020.
8. Hanna EB, Glancy DL. ST-segment depression and T-wave inversion: classification, differential diagnosis, and caveats. *Cleve Clin J Med* 2011; 78: 404–414.
9. Sakamoto T, Amano K, Hada Y, et al. Asymmetric apical hypertrophy: ten years experience. *Postgrad Med J* 1986; 62: 567–570.