Saw Tooth Cardiomyopathy with Parachute Mitral Valve – An Unusual Association

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Radiology Case. 2025 December ; 19(12):1-6 :: DOI: 10.3941/jrcr.5863

CONSENT

Informed written consent was obtained from the patient's parents for publication of clinical details and images. Identity has been protected.

CONFLICT OF INTEREST / DISCLOSURE

The authors declare no conflicts of interest and no financial disclosures.

ETHICAL STATEMENT / HUMAN AND ANIMAL RIGHTS

This study was conducted in accordance with the ethical principles of the Declaration of Helsinki. No human or animal experiments were performed beyond routine clinical care.

AUTHOR CONTRIBUTIONS

- Concept and design: DR ARATHY VIJAYAN, DR RICHA KOTHARI
- Clinical evaluation and patient management: DR VIMAL RAJ
- Cardiac imaging acquisition and interpretation: DR ARATHY VIJAYAN
- Manuscript drafting and literature review: DR ARATHY VIJAYAN, DR RICHA KOTHARI, DR VIMAL RAJ
- Critical revision of manuscript: DR RICHA KOTHARI, DR VIMAL RAJ

Key Points:

Association of Saw tooth cardiomyopathy with parachute mitral valve has not been described in previous literature.
Cardiac MRI with late gadolinium enhancement is the diagnostic standard.

ABSTRACT

Saw tooth cardiomyopathy (STC) is an extremely rare myocardial disorder, with fewer than 14 cases reported since its initial description in 2009. It is defined by distinctive myocardial projections resembling a saw tooth pattern on cardiac MRI. No consistent genetic or pathological abnormalities have been identified. Cardiac MRI with late gadolinium enhancement remains the diagnostic standard. Reported associations include apical aneurysm, patent foramen ovale, mitral valve prolapse, and myocardial bridging. We report a rare case of STC associated with a parachute mitral valve—an association not previously described—broadening the phenotypic spectrum of this under-recognized cardiomyopathy.

CASE REPORT

BACKGROUND

Saw tooth cardiomyopathy (STC) is an extremely rare cardiomyopathy, with fewer than 14 documented cases worldwide. No consistent pathological or genetic basis has been established, differentiating it from left ventricular non-compaction. Commonly reported associations include mitral valve prolapse, patent foramen ovale, and apical aneurysm. This case is significant because it describes the novel association of STC with parachute mitral valve, an anomaly not previously reported. It broadens the phenotypic spectrum of STC and emphasizes the role of advanced cardiac MRI in its diagnosis.

INTRODUCTION:

Saw tooth cardiomyopathy (STC) is a rare form of left ventricular cardiomyopathy characterized by saw-tooth like myocardial projections extending from the ventricular walls towards the ventricular cavity [1]. It is a rarely described entity, with fewer than 14 cases reported since its first mention in the literature as a variant of isolated left ventricular non-compaction [2]. The diagnosis of STC is often challenging because its clinical phenotype may closely resemble LVNC regarding symptoms, echocardiogram and ECG abnormalities. No consistent genetic associations or positive family history have been described,

even in cases where large gene panels were studied, suggesting either the need for more comprehensive genome sequencing or a non-genetic etiology. Clinical outcomes in reported cases have been largely stable, with most patients presenting with heart failure managed conservatively [3,4]. However, one patient experienced an ischemic stroke involving the left middle cerebral artery, requiring emergency thrombolysis [5]. Common associations of STC are left bundle branch block (LBBB), patent foramen ovale, myocardial bridging, left apical ventricular aneurysm and mitral valve prolapse.

Proposed criteria for diagnosing STC include saw-tooth or band-like projections originating from the interventricular septum to mid lateral segments, with apparently compact myocardium; Mild impaired left ventricular contraction function is usually clinically diagnosed with heart failure; Electrocardiogram is generally normal, or with left bundle branch block1.

We present a previously non described association of STC with parachute mitral valve.

Clinical details

An 11year male child was diagnosed with dilated cardiomyopathy at 3 months of age when he presented with excessive cry. He is a second born to non-consanguineous parents with uneventful natal history. He has achieved normal developmental milestones and is currently studying in the 4th grade with good academic performance. There is no history of recurrent fever, cough or cold. There is no history of seizures, and his hearing and vision are normal. He has been on medical therapy since diagnosis and is under close follow-up. He was doing well until 6 months back when he started experiencing chest pain and easy fatigability. On examination, his height and weight were within normal range. Legs and feet show some eczematous lesions, flat nails and brachydactyly with foetal finger pads noted. Cardiovascular and central nervous system examinations were also within normal limits. ECG (Figures 1-4) demonstrated sinus rhythm with interpolated PVCs, normal PR interval, flat P waves and LVH.

Echocardiography revealed mildly reduced left ventricular function with hypokinesia of anterior septum. Mitral valve prolapse with grade II mitral regurgitation. No aortic stenosis/ coarctation of aorta/anomalous origin of left coronary artery from pulmonary artery (ALCAPA). No LV non compaction. Further evaluation with cardiac MRI was then done.

Cardiac MRI revealed early dilated LV with ejection fraction of 48%. Saw tooth appearance of myocardium was seen with crypts and bands, especially along inferior and lateral walls. On delayed enhancement imaging, subtle subendocardial enhancement was noted along the apical lateral wall and apex. No myocardial edema was seen on STIR, and there was no LV thrombus. Origin of papillary muscle along mid left ventricular lateral wall was noted with its tip attached to the mitral valve. No left ventricle apical aneurysm, mitral valve prolapse or patent foramen ovale was seen.

Patient was continued on betablocker, angiotensin receptor-neprilysin inhibitor (ARNi), aldosterone antagonist for management of heart failure. Whole exome sequencing was suggested; however, the family did not proceed with the genetic analysis due to financial and personal reasons.

DISCUSSION

Saw tooth cardiomyopathy is a rarely described entity, with only a few cases mentioned in the literature. Although initially thought of as a variant of left ventricular non compaction cardiomyopathy, genetic and pathologic analysis done so far has not found a consistent correlation.

Cardiac MRI is sufficient for making the diagnosis. This case emphasizes the clinical relevance of cardiac MRI assessment in every case of dilated cardiomyopathy, as echocardiography did not detect the myocardial bands and projections. Extended genetic analysis might or might not have provided clinically significant information for change in patient management. Commonly described associations with this entity do not include parachute mitral valve.

Summary statement

This case report presents a previously undescribed association of Saw tooth cardiomyopathy with a parachute mitral valve, expanding the phenotypic spectrum of this under-recognized cardiomyopathy.

TEACHING POINT

Saw tooth cardiomyopathy is an ultra-rare diagnosis that can remain undetected on echocardiography but is readily identified by cardiac MRI. This case highlights the necessity of comprehensive MRI evaluation in pediatric dilated cardiomyopathy to uncover subtle myocardial architectural abnormalities and coexistent anomalies such as parachute mitral valve, expanding its clinical spectrum.

QUESTIONS

Question 1: Which embryological explanation best accounts for the morphological differences between saw tooth cardiomyopathy (STC) and left ventricular non-compaction (LVNC)?

A. Arrest of normal trabecular compaction during embryogenesis

B. Aberrant myocardial fiber alignment with focal delamination of compact myocardium

C. Secondary remodeling due to chronic LV wall stress in dilated cardiomyopathy

D. Excessive endocardial fibroelastosis interfering with myocardial development

Answer: B. Aberrant myocardial fiber alignment with focal delamination of compact myocardium

Question 2: In the reported case, which ECG abnormality was documented, and what is its clinical implication?

A. Persistent left bundle branch block – risk for dyssynchrony

B. Interpolated premature ventricular complexes – predisposition to ventricular arrhythmias

C. Pre-excitation syndrome – risk of supraventricular tachycardia

D. Prolonged QT interval – potential for torsades de pointes

Answer: B. Interpolated premature ventricular complexes – predisposition to ventricular arrhythmias

Question 3: Which imaging detail confirmed the diagnosis of parachute mitral valve in this patient?

A. Leaflet elongation with bileaflet prolapse

B. Single papillary muscle originating from mid-lateral LV wall with chordal convergence

C. Commissural fusion of both valve leaflets with doming motion

D. Restricted posterior leaflet excursion due to abnormal chordal tethering

Answer: B. Single papillary muscle originating from mid-lateral LV wall with chordal convergence

Question 4: Which histopathological correlate is most likely if tissue biopsy were performed in STC patients with subendocardial LGE?

A. Transmural myocyte dropout and replacement fibrosis

B. Patchy subendocardial interstitial fibrosis with preserved midwall architecture

C. Concentric hypertrophy of myocardial fibers with nuclear atypia

D. Lymphocytic myocarditis with necrotic foci

Answer: B. Patchy subendocardial interstitial fibrosis with preserved midwall architecture

Question 5: From a prognostic perspective, which factor increases the long-term risk of adverse outcomes in STC?

A. Presence of isolated saw-tooth projections with preserved ejection fraction

B. Concomitant congenital valve anomaly such as parachute mitral valve

C. Absence of family history and negative genetic testing

D. Normal ECG without conduction abnormalities

Answer: B.

Question 6: Which imaging modality is considered the diagnostic gold standard for identifying saw tooth cardiomyopathy?

A) Cardiac CT

B) Echocardiography

C) Cardiac MRI with late gadolinium enhancement

D) PET scan

Answer: C

Question 7: Which genetic finding has been consistently associated with saw tooth cardiomyopathy?

A) TTN truncation

B) MYH7 mutation

C) No consistent genetic abnormality

D) LMNA mutation

ANSWER - C

Question 8: Which statement is true regarding outcomes in saw tooth cardiomyopathy?

A) Outcomes are stable with conservative management

B) Always associated with genetic mutations

C) All patients present with arrhythmias

D) Most require heart transplantation

ANSWER – A

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FIGURES

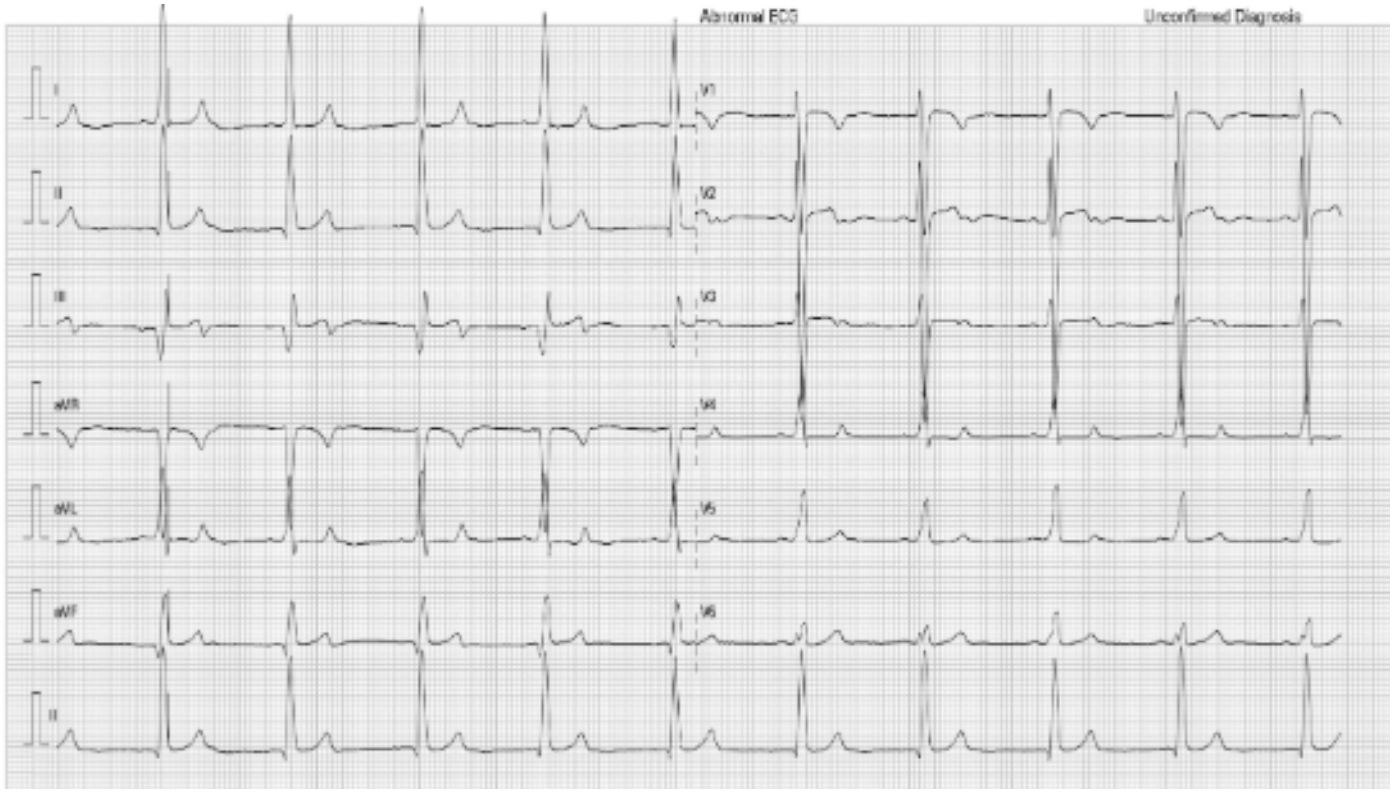


Figure 1: ECG of the child showing sinus rhythm with interpolated PVCs, normal PR interval and flat p waves

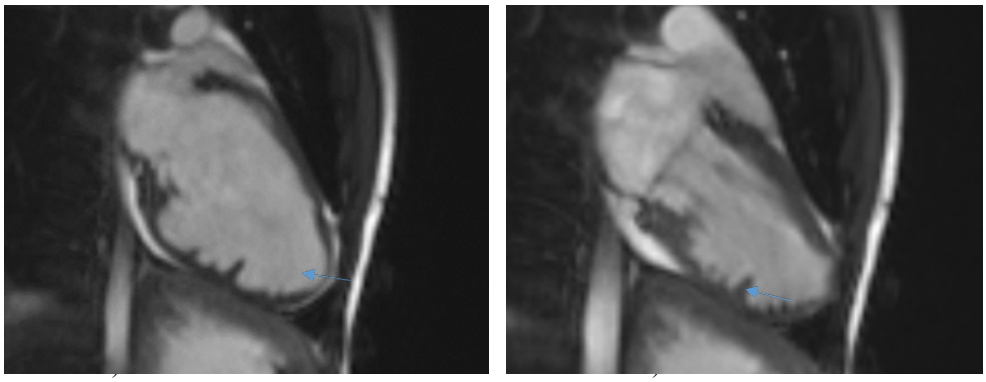
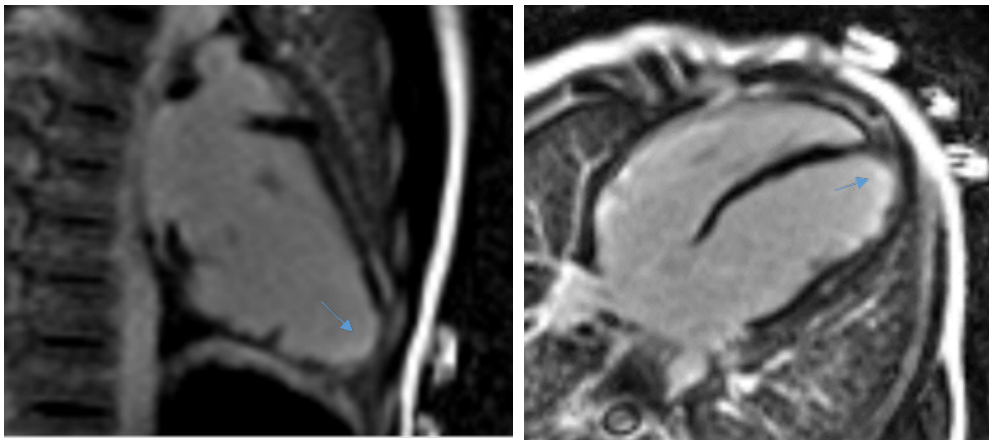
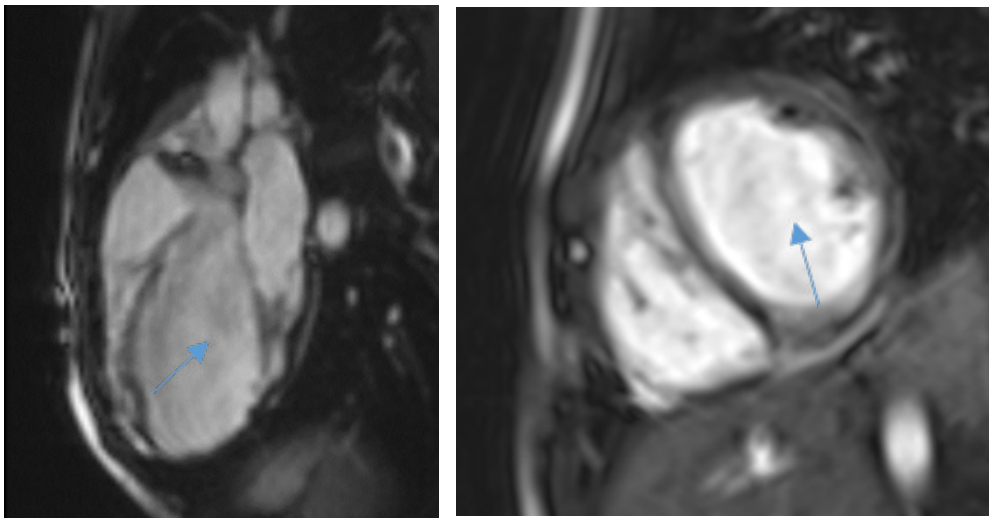


Figure 2: 2 chamber view of heart End diastole (i) and end-systole (ii) images showing saw tooth projections (blue arrows)



i) ii)

Figure 3: LGE images in 2 CH (i) and 4CH (ii) views showing subtle subendocardial enhancement along apical lateral wall and apex (blue arrows).



i) ii)

Figure 4: Cine images in 3 CH (i) and short axis (ii) planes showing parachute mitral valve (blue arrows)

KEYWORDS

Saw tooth cardiomyopathy; Parachute mitral valve; Cardiac MRI; Pediatric cardiomyopathy; Dilated cardiomyopathy.

ABBREVIATIONS

STC = Saw Tooth Cardiomyopathy
LV = Left Ventricle
LVNC = Left Ventricular Non-Compaction
MRI = Magnetic Resonance Imaging
LGE = Late Gadolinium Enhancement
LBBB = Left Bundle Branch Block
ALCAPA = Anomalous Origin Of Left Coronary Artery From Pulmonary Artery
ARNI = Angiotensin Receptor-Neprilysin Inhibitor

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