

Post-Cardiac Injury Syndrome after Surgical Repair of Atrial Septal Defect: Reporting a Rare Occurrence

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Abstract

A 16-year-old male with past medical history of congenital atrial septal defect surgical repair presented with recurrent pericarditis secondary to post-cardiotomy injury syndrome (PCIS). After failing medical therapy, he ultimately underwent pericardiectomy for symptom resolution. PCIS is underdiagnosed in children and should be considered in patients with recurrent chest pain.

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Abstract:

A 16-year-old male with past medical history of congenital atrial septal defect surgical repair presented with recurrent pericarditis secondary to post-cardiotomy injury syndrome (PCIS). After failing medical therapy, he ultimately underwent pericardiectomy for symptom resolution. PCIS is underdiagnosed in children and should be considered in patients with recurrent chest pain.

Key Words: recurrent pericarditis, multimodality imaging, atrial septal defect, post-cardiac injury syndrome, pericardiectomy

Abbreviations:

ASD: atrial septal defect

CRP: C-reactive protein

ESR: erythrocyte sedimentation rate

hs-CRP: high-sensitive C-reactive protein

NSAIDs: non-steroid anti-inflammatory drugs

PCIS: post-cardiac injury syndrome

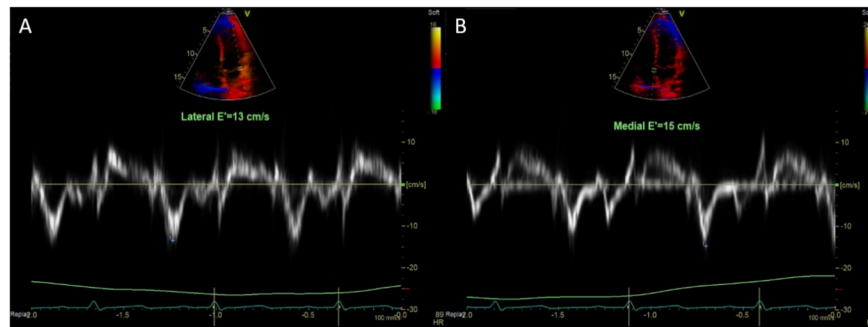
RP: recurrent pericarditis CMR: cardiac magnetic resonance imaging Echo: Echocardiography

Introduction:

Post cardiac injury syndrome (PCIS) is the development of pericardial diseases in patients with prior history of cardiac surgery, electrophysiology procedures, myocardial infarction, or percutaneous coronary intervention. The incidence of PCIS varies and is dependent on underlying cause.¹ Cardiac surgery is the most common cause (10-40%) followed by electrophysiology procedures (1-2%), and coronary intervention (0.1-1%).¹ The clinical presentation is varied as symptoms may develop even after a few years therefore making diagnosis and associated management difficult. Here, we present a case of an adolescent male who developed PCIS induced recurrent pericarditis (RP) after surgical repair of an atrial septal defect (ASD).

Case Presentation:

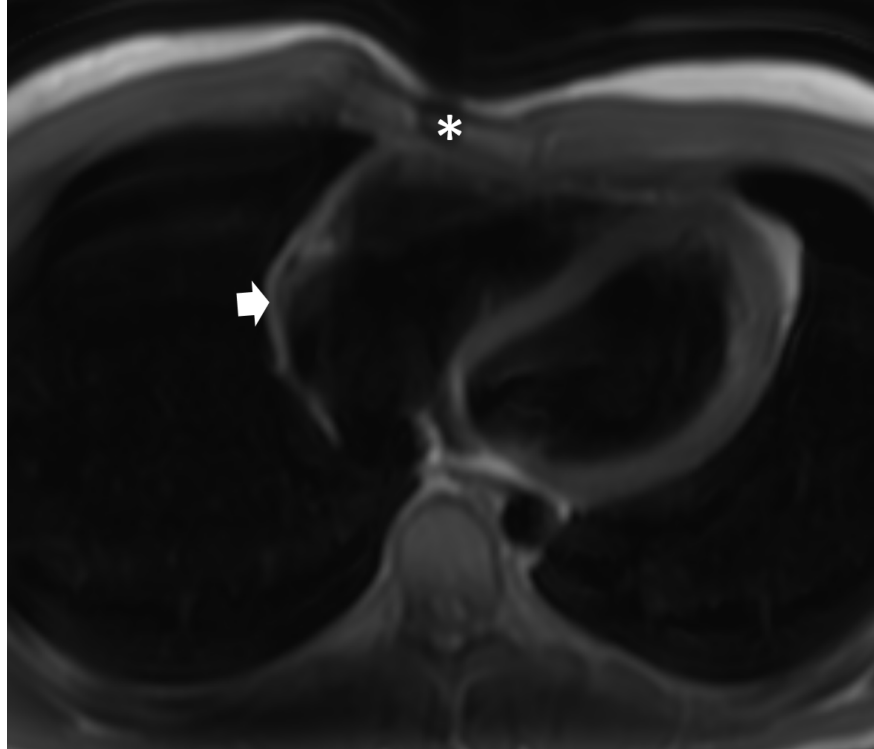
A 16-year-old male with past medical history of pericarditis that developed after surgically repaired ASD presented for recurrent episodes of chest pain. He described his chest pain as 7/10, pleuritic, sharp, mid-sternal, and worse when supine. Pain was non-radiating without any associated shortness of breath, fever or chills. Physical examination and vital signs were within normal limits. Cardiovascular exam showed regular rate, normal rhythm, S1, S2 sounds, and no pericardial rub. His chest pain began a few months after surgery, and at that time he was diagnosed with acute idiopathic pericarditis. After diagnosis, he frequently experienced flares limiting his daily activities. During these episodes, he was intermittently treated with a combination of non-steroidal anti-inflammatory drugs (NSAIDs), colchicine, and prednisone.



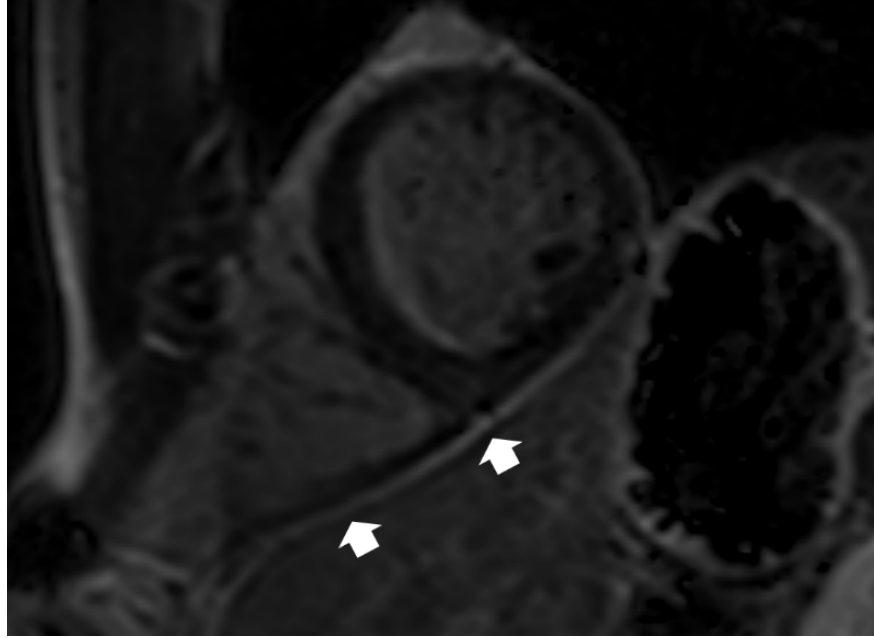
The patient underwent further workup. Chest x-ray (CXR) showed no focal pulmonary consolidation, cardiac enlargement or pericardial calcification. Inflammatory markers including erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and high-sensitive C-reactive protein (hs-CRP) were elevated. High-sensitivity troponin and brain natriuretic peptide (BNP) levels were within normal limits. A 12-lead electrocardiogram (ECG) demonstrated normal sinus rhythm. Echocardiogram displayed diastolic septal bounce and the presence of mild constrictive physiology with annular reversal (**Figure 1**) .

(Figure 1).

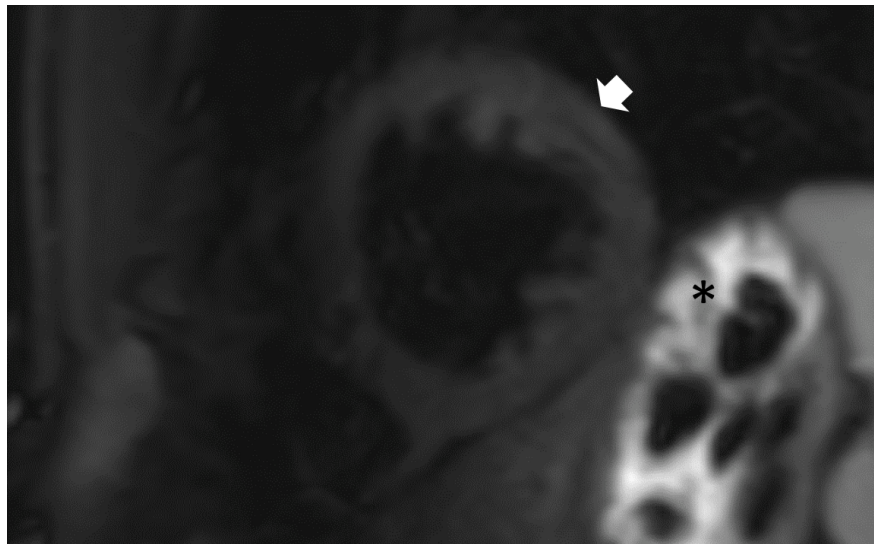
Cardiac magnetic resonance imaging (CMR) identified mild pectus deformity (Haller Index: 3.6) and mild late gadolinium enhancement (LGE) in the absence of pericardial edema (**Figures 2,3,4**) .



(Figure 2)



(Figure 3)



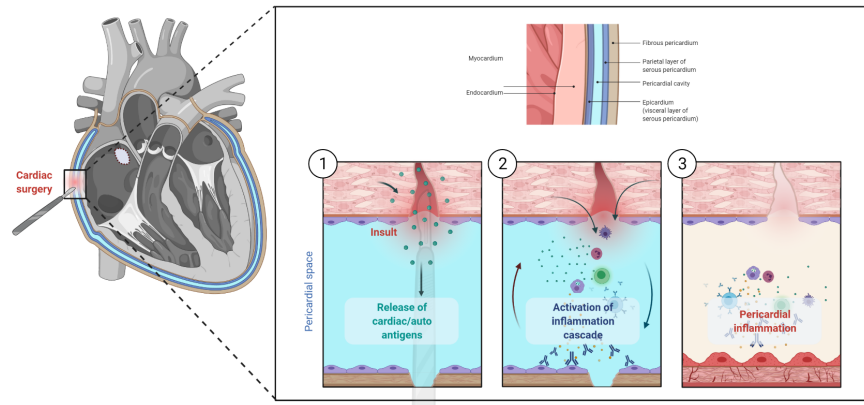
(Figure 4)

Based on his clinical and imaging findings, he was diagnosed with recurrent pericarditis complicated by constrictive physiology. Given his frequent recurrences, he was treated with ibuprofen 800 mg three times a day, colchicine 0.6 mg twice a day and prednisone 40 mg daily. He was counseled on the need for strict exercise restriction and adherence to medication. Additionally, the criteria for advancing the therapy to biologics (anakinra) or pericardiectomy was also discussed. Unfortunately, 4 months after his visit, he developed another recurrence. Given his significant limitation in quality of life (QoL), he elected to undergo pericardiectomy. Surgical pathology revealed organized pericarditis without any evidence of necrosis or necrobiosis.

At 6-month follow-up, he reported significant improvement in his functional capacity, and QoL He reported no further flares. ESR, CRP, and hs-CRP were normal. ECG demonstrated normal sinus rhythm. Repeat transthoracic echocardiogram revealed no constrictive physiology but displayed development of new mild tricuspid regurgitation. His colchicine was tapered from 0.6 mg to 0.3 mg daily and was instructed to follow up in 6 months for further tapering.

Discussion:

PCIS is well reported in adults, however it is rare in children. The proposed pathophysiological mechanism of PCIS includes release of cardiac antigens in the pericardial space precipitating chemokines and anti-myocardial antibodies production. Ongoing inflammation at the epicardial level results in an inflammatory cascade (**Figure 5**).^{1,2}



(**Figure 5**)

Indications for ASD closure in childhood are well documented, and treatment is known to be effective.³ Complications including development of cardiac tamponade, recurrent pericardial effusion or pericardial effusions requiring drainage after ASD closure are rare.^{1,4-6} Furthermore, these pericardial complications are usually reported in the peri-procedure period, as compared to our patient who developed symptoms several months after surgery.⁷ RP concomitantly with constrictive physiology in these patients after a latent period has been sporadically reported in the literature. A prior study of 15 cases on RP in children and adolescents concluded that surgical ASD closure (n=6) was the predominant underlying etiology.⁶ Interestingly, the risk of development of PCIS presenting as RP is similar between pediatric and adult populations.⁸

Echocardiogram and CMR supplemented clinical information to attain diagnosis in our patient. CMR findings: mild LGE, right ventricular tethering, diastolic septal bounce, and respirophasic shift supported the diagnosis of recurrent pericarditis complicated by constrictive physiology in our case.¹ Short courses or abrupt cessation of anti-inflammatory therapy without adequate tapering may have resulted in residual pericardial inflammation, increasing his risk of further flares. Treatment of pericarditis requires NSAIDs and colchicine (as first line), steroids (second line), biologics such as interleukin-1 receptor blockers or disease modifying anti-rheumatic drugs (third line) and pericardiectomy (fourth line in refractory cases).^{9,10}

Our case represents a rare occurrence of RP with constrictive physiology treated with pericardiectomy in an adolescent patient with prior surgical ASD repair. Medical management of PCIS can be complicated due to its prolonged duration and notable side effects. Further, it can be an underdiagnosed or under-reported condition among adolescents. Patients with ASD repair are living longer and require monitoring for complications.¹¹ Therefore, in order to improve their outcomes it is imperative to timely identify the development of PCIS and adequately manage it.

Conclusion:

Recurrent pericarditis along with constrictive physiology may occur in adolescent patients with prior ASD repair after a prolonged symptom free interval. Therefore, adolescents with repaired congenital heart defects should be carefully screened for development of PCIS. Management can be challenging and often requires advanced imaging (CMR) to guide appropriate therapy. Treatment may necessitate addition of biologics to anti-inflammatory therapy or further escalation to pericardiectomy in refractory cases. Further research is needed to identify at-risk patients and design preventive strategies.

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Figures:

Figure 1: Mitral annular early diastolic velocities ((lateral (A) and medial (B)) demonstrating annulus reversus.

Figure 2: Black blood imaging demonstrates no overt pericardial thickening (*arrow*).

Note additional finding of mild pectus chest wall deformity (*).

LA=Left Atrium

LV=Left Ventricle

RA=Right Atrium

RV=Right Ventricle

Figure 3: T1-weighted (fat suppressed) phase sensitive inversion recovery sequence showing mild, predominantly basal pericardial delayed enhancement (*arrows*).

LV=Left Ventricle

RV=Right Ventricle

Figure 4: T2-weighted short-tau inversion recovery sequence, showing absence of significant pericardial edema (*arrow*). In contrast, there is increased signal detected within the adjacent gastrointestinal tract (*).

LV=Left Ventricle

RV=Right Ventricle

Figure 5: Pictorial representation of pathophysiology of post cardiac injury syndrome. *Figure created using BioRender.*