

VALVULAR HEART DISEASE

CASE REPORT: CLINICAL CASE

Acute on Chronic Rheumatic Valvulitis



Natalie K. Craik, MD,^a Joseph Burns, MD,^a Nirica Borges, MD,^a Tam T. Doan, MD,^a
Amy E. Sanyahumbi, MD, MPH,^a Edward J. Hickey, MD,^a Debra L. Kearney, MD,^a Ryan H. Rochat, MD,^a
Eyal Muscal, MD,^a Thomas Glenn, MD^a

ABSTRACT

An 11-year-old boy presented in distress with tachypnea, holosystolic murmur, and a gallop. Echocardiography revealed mitral valve thickening and severe regurgitation. He required valve replacement with pathology consistent with acute on chronic valvulitis. This case underscores the importance of considering rheumatic heart disease, despite no preceding suspicious history. (JACC Case Rep. 2024;29:102618) © 2024 Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

HISTORY OF PRESENTATION

An 11-year-old Hispanic boy with no medical history presented with 3 days of chest pain and labored breathing. He experienced one episode of emesis but denied diarrhea, fever, rhinorrhea, and cough. His weight was 38.6 kg (42nd percentile), height 150 cm (57th percentile), and BMI 17.2 kg/m². Vital signs were as follows: blood pressure 106/72 mm Hg, pulse 116 beats/min, temperature 37.1 °C, respiratory rate 40 breaths/min, and oxygen saturation 99%. His physical examination was significant for tachypnea, a IV/VI holosystolic murmur with diastolic rumble, and

intermittent gallop best heard in the left sternal border.

PAST MEDICAL HISTORY

His mother reported his history of 2 prior viral infections with no concern for streptococcal infection. She denied other episodes of high fever, sore throats, skin infections, or travel outside of the country. She denied any prior cardiac symptoms; however, she noted he may have been self-limiting his activities as he primarily enjoyed sedentary pastimes and his school physical education was not rigorous.

DIFFERENTIAL DIAGNOSIS

This presentation prompted a broad differential diagnosis before obtaining an echocardiogram; however, chest pain, tachypnea, emesis, and new murmur with a gallop are alarming for acute heart failure. The most common etiologies are congenital heart disease, infectious myocarditis, dilated cardiomyopathy, and arrhythmia-induced cardiomyopathy.¹

LEARNING OBJECTIVES

- To recognize the clinical findings of rheumatic heart disease in a low-risk population.
- To understand the immediate medical and operative management for patients with acute on chronic rheumatic heart disease.

From the ^aBaylor College of Medicine & Texas Children's Hospital, Houston, Texas, USA.

The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the [Author Center](#).

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ABBREVIATIONS AND ACRONYMS

ARF = acute rheumatic fever
RHD = rheumatic heart disease

INVESTIGATIONS

At an outside institution, he was noted to have an elevation in brain natriuretic peptide and troponin, prompting transfer to Texas Children's Hospital. He was started on bilevel-positive airway pressure 10/5 on arrival. A chest radiograph was obtained (**Figure 1**), and his laboratory results confirmed an elevated troponin (0.048 ng/mL) and brain natriuretic peptide (3115 pg/mL).

His remaining laboratory work revealed mildly elevated C-reactive protein (2.9 mg/dL), increased transaminases (aspartate aminotransferase 89 mg/dL, alanine transaminase 70 mg/dL), mild hyperbilirubinemia (1.9 mg/dL), and mild microcytic anemia (mean corpuscular volume 72.4, hemoglobin 11.5 g/dL).

His echocardiogram demonstrated nodular thickening of his mitral valve, and severe mitral regurgitation via a flail anterior mitral leaflet consistent with acute valvulitis² (**Figure 2**). Tethering of his posterior mitral valve leaflet was also noted (**Figure 2**), possibly suggestive of chronic valvulitis.³

There was also severe dilation of the left atrium and mild to moderate dilation of the left ventricle with normal function. Based on upper mild tricuspid

regurgitation jet velocity, right ventricular pressures were half-systemic. Highest yield initial echocardiogram videos (including apical and parasternal long views with and without color) can be viewed on the online version of this paper. Educational figures were drawn for the family (**Figure 3**).

MANAGEMENT: PREOPERATIVE COURSE

After admission to the cardiovascular intensive care unit, he began milrinone therapy and aggressive diuresis. He completed 48 hours of antibiotics empirically for endocarditis rule out. His anti-streptolysin O titer was positive, his anti-DNAse antibody was elevated, and he began penicillin V potassium prophylaxis. He had ongoing fatigue, tachycardia, mild respiratory distress, and recurrent episodes of brief self-resolving atrial tachycardia. On day 5 of admission, he developed a fever (38.7 °C) and thus met 1 major (carditis suggestive of mitral valvulitis) and 2 minor (C-reactive protein >3.0 mg/dL, fever) revised Jones criteria, thereby meeting criteria for acute rheumatic fever (ARF).

His Karius test, a liquid biopsy for infectious etiology, was negative. Given the degree of decompensated heart failure, although steroids were considered, they were ultimately not given due to concern for poor wound healing in the event of surgery.

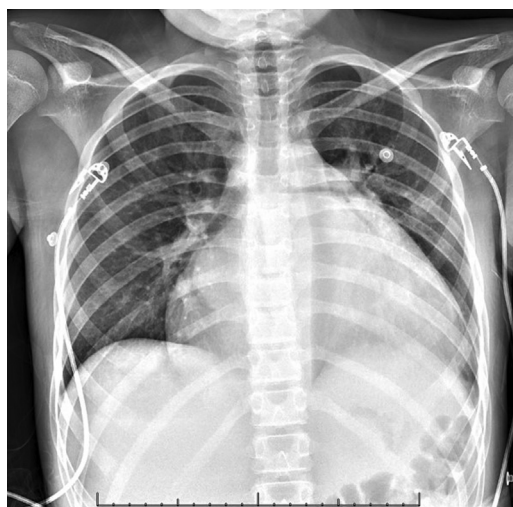
Because we were unable to meaningfully improve his cardiac function medically, he was presented for surgery and, on hospital day 13, he underwent cardiac surgery.

MANAGEMENT: PATHOLOGY/HISTOLOGY AND OPERATIVE COURSE

Intraoperatively, extensive pericardial gelatinous adhesions (**Figure 4**) and a thickened, inflamed mitral valve (**Figures 5A and 5B**) with no obvious vegetations were noted. Subsequent histology revealed areas of acute and chronic valvulitis (**Figures 5D and 5E**). There were many histiocytic aggregates focally suggestive of Aschoff bodies with Anitschkow cells, pathognomonic for acute rheumatic heart disease (RHD) (**Figures 5C and 5F**). Although prominent neutrophils raised concern for another infectious valvulitis, stains and polymerase chain reaction studies for acid-fast, bacterial, and fungal organisms were negative.

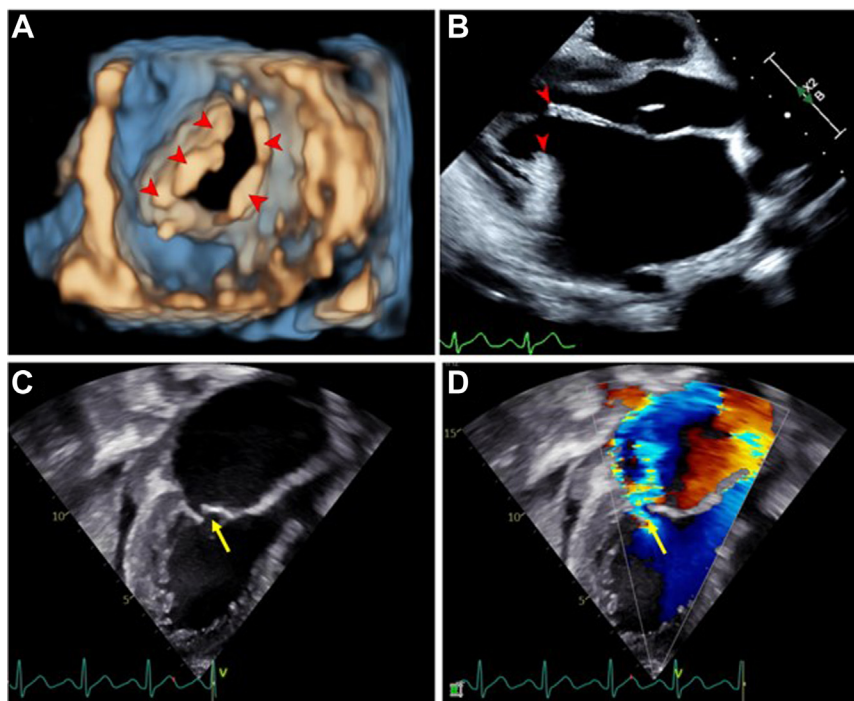
The mitral valve was successfully replaced with a 27-mm On-X valve, and tricuspid annuloplasty was performed.

FIGURE 1 Chest Radiograph



Initial chest radiograph demonstrated severe cardiomegaly, left bronchial compression, and diffuse pulmonary edema consistent with mitral valvulitis and severe regurgitation.

FIGURE 2 Echocardiographic Findings



(A) Three-dimensional en face view of the mitral valve leaflets viewed from the left ventricle showing nodular thickening of both leaflets (arrowheads) in diastole. (B) Parasternal long-axis view of the mitral valve leaflets in diastole showing thickening of both leaflets (arrowheads). (C) Apical long-axis view of the mitral valve leaflets in systole showing evidence of flail anterior mitral leaflet (arrow) and poor coaptation with the tethered posterior leaflet. (D) Apical long-axis view of the mitral valve leaflets in systole showing severe mitral regurgitation via a posteriorly deviated jet through the flail anterior mitral leaflet (arrow).

MANAGEMENT: POSTOPERATIVE

He was weaned off milrinone and begun on enalapril and spironolactone due to moderately depressed function postoperatively, which was continued through discharge. He remained an inpatient due to heparin bridging to therapeutic warfarin. He is to continue penicillin V indefinitely.

DISCUSSION

ARF describes the autoimmune response to group A *Streptococcus*.³ RHD describes the long-term valvular disease that can occur after ARF.^{4,5} In high-income settings, the rate of RHD has steeply declined.⁵ A multicenter evaluation from 2008 to 2018 reported on 947 cases in children between 4 and 17 years of age in the United States.⁶ Estimates among higher-risk

populations, including American Indians, report an incidence of 0.5% in adults.⁷

RHD classically involves the left-sided valves due to endothelial inflammation; in children, mitral regurgitation predominates in the first decade of life.⁸ Patients with a history of ARF are at a higher risk of recurrence, with estimates from 15% to 34%,⁹ and therefore require antibiotic prophylaxis for secondary prevention.⁸ This patient's presentation and histology are unusual for high-income countries. Although there is laboratory evidence of a preceding streptococcal infection, he was largely asymptomatic until his acute illness. His histologic and echocardiographic findings are suggestive of both acute and chronic valvulitis. Of note, this patient now has 2 definitive risk factors which adult studies have demonstrated increase risk of recurrence of ARF (under 23 years of age and prior heart failure) and 1 potential risk factor

FIGURE 3 Educational Diagram

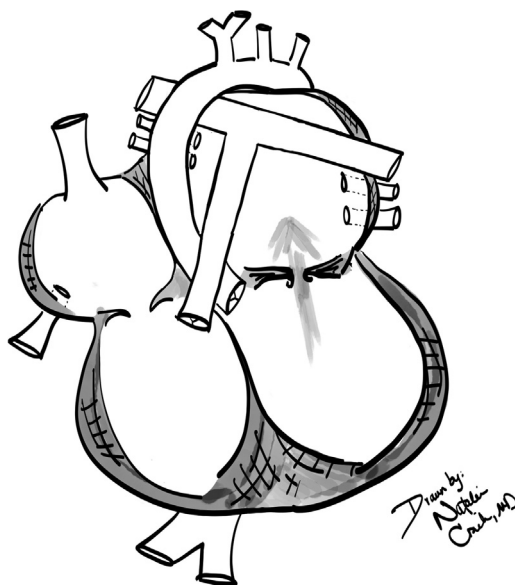


Diagram used for family and bedside provider education, demonstrating cardiomegaly with significant left atrial dilation, thickened mitral valve leaflets, and severe mitral regurgitation.

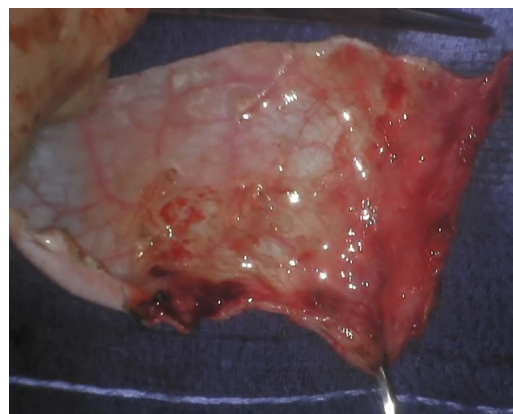
(poor adherence to antibiotic prophylaxis)¹⁰ in the setting of historical lapses of insurance coverage.

This case underscores the importance of consideration of RHD in pediatric patients in the United States and the need for awareness of the increased incidence of RHD in non-White children.⁴ Furthermore, children already affected by social determinants of health (including living in socioeconomically deprived areas) are at a higher risk for more severe disease,⁴ likely attributable to structural and societal factors, including access to health care, health literacy, transportation, and myriad of other barriers to care.

FOLLOW-UP

After discharge, he demonstrated normalization of ventricular function and a low mitral valve mean gradient (2.6 mm Hg). He has continued to require

FIGURE 4 Intraoperative Picture



Gross intraoperative photograph of pericardium with extensive gelatinous adhesions.

titration of his warfarin dose and follows up regularly in clinic. The family lacked insurance for the first months after his surgery, making INR titration and echocardiogram monitoring financially and logistically demanding for the family. He remains adherent with all medications.

CONCLUSIONS

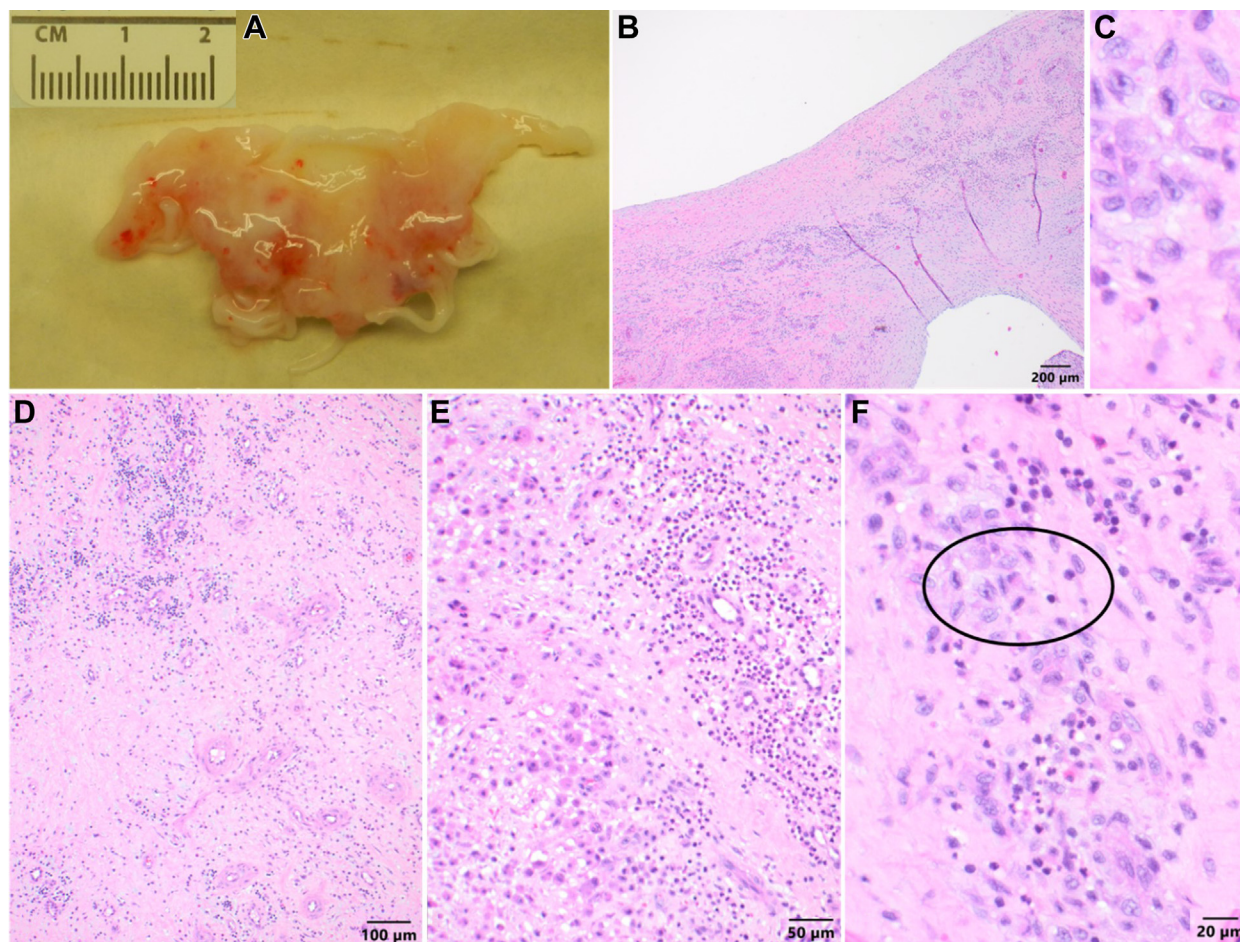
Even in the United States, it is imperative to reinforce prompt evaluation and treatment of pharyngitis and skin infections for primary prevention of RHD and, in affected patients, to ensure adequate penicillin prophylaxis to prevent recurrence and sequelae.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Natalie K. Craik, Texas Children's Hospital, Legacy Tower, 6651 Main Street, Floor 14, Houston, Texas 77030, USA. E-mail: Natalie.craik@bcm.edu.

FIGURE 5 Histology and Histopathology



(A) Gross photograph of thickened, inflamed anterior mitral valve leaflet and chordae tendineae. (B) Low power view of thickened leaflet with fibrosis and inflammation. (C) High power view of Anitschkow histiocytes with typical caterpillar-like nuclei. (D) Chronic valvulitis with fibrosis, lymphocytic infiltrates, and neovascularization. (E) Acute valvulitis with histiocytic infiltrates suggesting Aschoff bodies (lower left) and focally prominent neutrophils (upper right). (F) Mixed inflammatory infiltrates with focal Anitschkow histiocytes (circle).

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KEY WORDS child, mechanical valve, rare, social determinants of health, socioeconomic, United States

APPENDIX For supplemental videos, please see the online version of this paper.