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CASE REPORT

ADVANCED

CLINICAL CASE

Pericardiectomy for Successful Treatment of Constrictive Pericarditis in a Pediatric Patient

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ABSTRACT

A 15-year-old girl with history of asthma and obesity presented with recurrent anasarca without systolic heart failure or significant renal disease. She was diagnosed with constrictive pericarditis and successfully underwent pericardiectomy with pericardial stripping and a waffle procedure. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;23:102009)

HISTORY OF PRESENTATION

A 15-year-old girl presented with worsening lower extremity edema, shortness of breath, and fatigue over 6 months. Vital signs were normal (blood pressure: 108/80 mm Hg; heart rate: 100 beats/min, SpO_2: 96%; respiratory rate: 24/min). Her breath sounds were absent at the bilateral lung bases. Heart sounds were distant and without rub or murmur. There was 1+ pitting edema of the lower extremities.

LEARNING OBJECTIVES

- To recognize the clinical characteristics of constrictive pericarditis, especially in the pediatric population, where it is a rare entity.
- To learn surgical treatment strategies for constrictive pericarditis.

PAST MEDICAL HISTORY

The patient had a history of asthma and obesity (103.2 kg; body mass index: 39.05 kg/m^2). Review of systems was negative for fever, rashes, joint swelling/pain, vomiting, diarrhea, or other constitutional symptoms. History was significant for a hospital admission 1 month prior for treatment of bilateral pleural effusions requiring drainage. Laboratory tests during the admission revealed positive antinuclear antibody (1:320) and anti-double-stranded DNA (29 IU/mL) findings, resulting in a presumptive diagnosis of systemic lupus erythematosus (SLE), for which she was treated with steroids.

DIFFERENTIAL DIAGNOSIS

Specific etiologies for pericarditis should be ruled out first, including SLE, tuberculosis, and other infectious etiologies.

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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

CP = constrictive pericarditis

HD = hospital day

SLE = systemic lupus erythematous

INVESTIGATIONS

Electrocardiogram (**Figure 1**) revealed normal sinus rhythm at 91 beats/min, normal axis in the frontal plane, low-voltage QRS complex, and T-wave inversion in the inferolateral leads. An echocardiogram revealed no structural abnormalities, normal biventricular systolic function, no evidence of pulmonary hypertension, and a small to moderate circumferential pericardial effusion with evidence of tamponade physiology. Specifically, there was augmentation of diastolic antegrade flow with expiration in the hepatic vein Doppler and 25% to 30% respirophasic variation in mitral and tricuspid inflow velocities (**Figures 2 and 3**). There was no diastolic free wall collapse of the right atrium or right ventricle. A "septal bounce" was appreciated (Video 1).

She had no evidence of renal disease or significant systemic inflammation. Right-sided thoracentesis yielded 2 L of transudative fluid. She was treated with ibuprofen and furosemide for presumptive chronic pericarditis with improvement in her dependent edema.

Laboratory examination showed mild anemia (9.6 g/dL) and elevated D-dimer (3.96 μg/mL). The antinuclear antibody result was positive at 1:320, and the anti-double-stranded DNA (IU/mL) result was weakly positive at 29. The erythrocyte sedimentation rate was slightly elevated at 31 mm/h (**Table 1**).

She underwent cardiac catheterization on hospital day (HD) 2 and was found to have elevated filling pressures (mean right arterial pressure: 24 mm Hg; left pulmonary capillary wedge pressure: 24 mm Hg) and right ventricle/left ventricle pressure discordance, consistent with CP. She underwent pericardiocentesis. Serosanguineous fluid (130 mL) was drained, after which she had equalization of left ventricular end-diastolic pressure/right ventricular end-diastolic pressure/left atrium/right atrium, with a left pulmonary capillary wedge pressure of 24 mm Hg, with a decrease to 22 mm Hg after pericardiocentesis (**Table 2**).

FIGURE 1 Electrocardiogram on Admission: Sinus Tachycardia With Nonspecific T-Wave Abnormalities

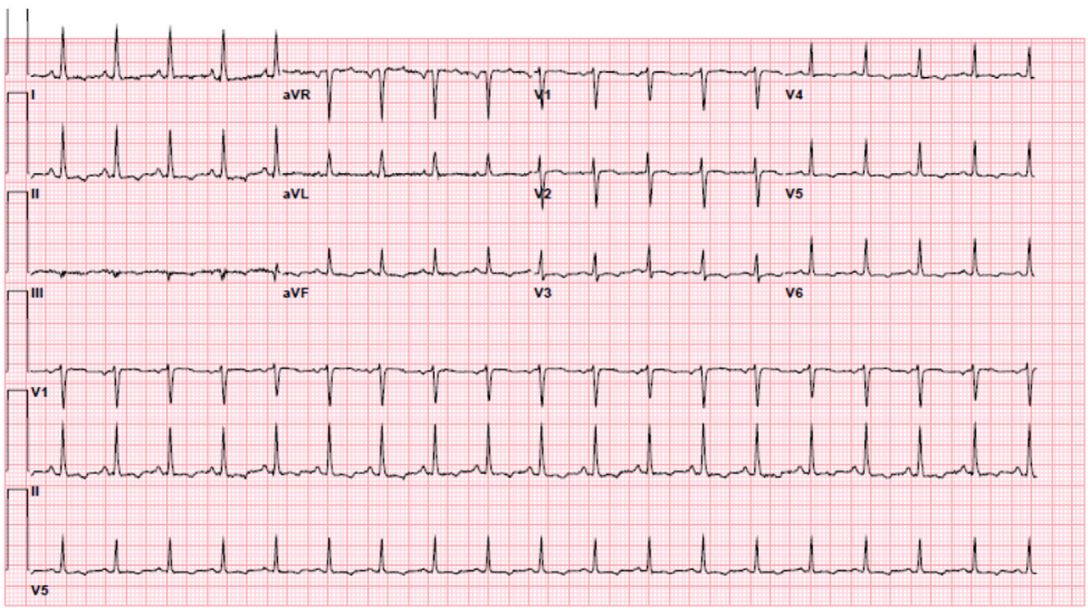
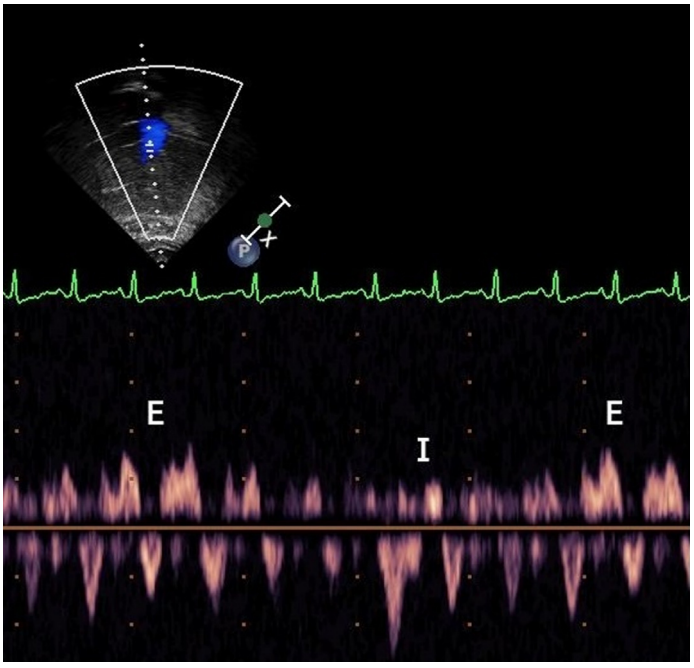


FIGURE 2 **Echocardiogram: Prominent Expiratory Diastolic Flow Reversal in Hepatic Veins**



E = expiration; I = inspiration; P and x = probe markers.

Cardiac magnetic resonance on HD3 demonstrated increased pericardial thickness up to 6 mm and pericardial enhancement, suggestive of inflammation and pericardial constriction (Figure 4). The hydroxychloroquine and mycophenolate were discontinued because of low suspicion of SLE.

MANAGEMENT

Initial management included steroids, hydroxychloroquine, and mycophenolate for presumptive SLE. Ibuprofen and furosemide were added for presumed chronic pericarditis treatment.

FIGURE 3 **Echocardiogram: Mitral Valve Inflow Variation**

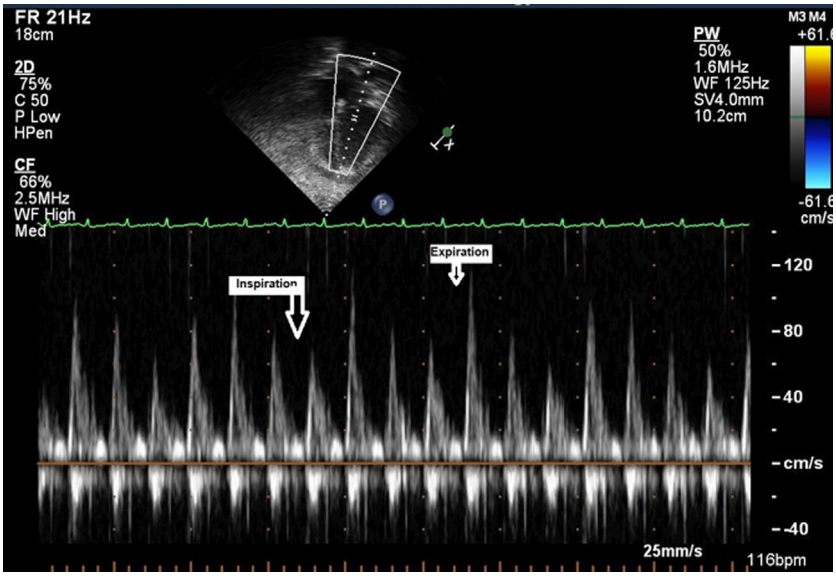


TABLE 1 Laboratory Test Results

	Normal	Initial Presentation	Transfer of Care
CBC			
WBC, $\times 10^3/\mu L$	6.2-15.5	–	8.6
Neutrophil, %	21.3-69.3	–	62.1
Lymphocyte, %	17.0-63.7	–	20.6
HGB, g/dL	10.3-13.2	–	9.6
PLTS, $\times 10^3/\mu L$	150-500	–	366
Inflammatory markers			
CRP, mg/L	0.0-5.0	0.9	–
ESR, mm/h	0-10	31	–
Ferritin, ng/mL	20-200	38.3	–
Procalcitonin, ng/mL	<0.49	–	0.04
Albumin, g/dL	3.5-4.9	4.1	–
Coagulation/cardiac studies			
D-dimer, μg/mL	0.00-0.50	–	3.96
BNP, pg/mL	0-100	260	–
Microbiology			
EBV PCR	Negative	Negative	–
CMV PCR	Negative	Negative	–
Parvovirus PCR	Negative	Negative	–
Histoplasmosis Ag	Negative	Negative	–
QuantiFERON-TB-Gold (QIAGEN)	Negative	Negative	–
Serum *Entamoeba histolytica* serology	Negative	Negative	–
Beta-d-glucan	Negative	Negative	–
HIV	Negative	Negative	–
Rheumatologic			
C3, mg/dL	84-168	150	143
C4, mg/dL	13-44	18	22
ANA	Negative	1:320	–
Anti-dsDNA	Negative	29	–
ANCA	Negative	Negative	–
Anti-Smith	Negative	Negative	–
ACE	Negative	Negative	–

ACE = angiotensin-converting enzyme; Ag = antigen; ANA = antinuclear antibody; ANCA = antineutrophil cytoplasmic antibody; BNP = B-type natriuretic peptide; CBC = complete blood count; CMV = cytomegalovirus; CRP = C-reactive protein; dsDNA = double-stranded DNA; EBV = Epstein-Barr virus; ESR = erythrocyte sedimentation rate; HGB = hemoglobin; PCR = polymerase chain reaction; PLTS = platelets; TB = tuberculosis; WBC = white blood cell.

TABLE 2 Catheterization Results

	Prepericardiocentesis	Postpericardiocentesis
R_p, units	1.22 (2.53 × m^2)	1.30 (2.70 × m^2)
R_s, units	12.45 (25.77 × m^2)	11.52 (23.85 × m^2)
Q_p/Q_s	1.001	1.00:1
R_p/R_s	0.10	0.11

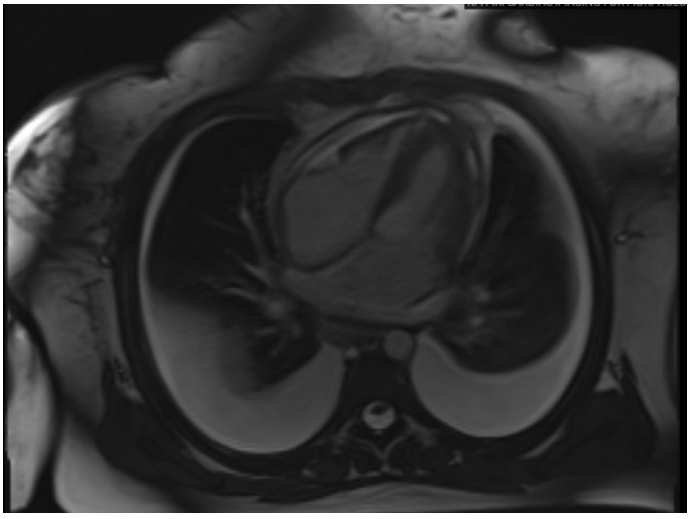
Values in parentheses are adjusted for body surface area.

R_p = pulmonary vascular resistance; R_s = systemic vascular resistance; Q_p = pulmonary flow; Q_s = systemic flow.

The decision was made to pursue pericardiectomy with pericardial stripping and a waffle procedure.[1] On HD 9, she underwent pericardiectomy off cardiopulmonary bypass with full sternotomy. The opening central venous pressure was 35 mm Hg. The pericardium was entered in the midline, where 150 mL of pericardial and 1.5 L of pleural fluid were drained. The parietal and visceral pericardium were both thickened. The visceral pericardium was glistening and completely encapsulating the heart, restricting

motion. The parietal pericardium was stripped in its entirety, except for strips left along each of the phrenic nerves (Figure 5). The visceral pericardium was then tediously peeled away from the right atrial and ventricular surface. The entire surface of the left ventricle was scored in a grid-like fashion using a number 15 scalpel, completing the waffle operation and allowing the left ventricle to expand (Figure 6). Care was taken not to injure the coronary vessels. A perioperative transesophageal echocardiogram showed mild mitral regurgitation, mild tricuspid regurgitation, and normal biventricular function. Postoperatively, she had prolonged chest tube drainage and the same echo findings. A pericardial biopsy finding was consistent with fibrous pericarditis.

FIGURE 4 Cardiac Magnetic Resonance With Pericardial Thickening and Enhancement



DISCUSSION

Constrictive pericarditis (CP) is a rare entity within the pediatric population, with limited published case reports. We present a rare case of a 15-year-old girl with CP who was successfully treated by pericardiectomy with stripping and a waffle procedure.[1] Her presentation was significant for persistent, recurrent anasarca without significant systemic inflammation, renal disease, and systolic heart failure. The incidence of pericarditis has widely ranged in the literature between 11% and 54%.[2-4] CP accounts for approximately 7% of all cases and is uncommon in pediatric patients.[5]

This patient presented with tamponade physiology without strong evidence to support an underlying etiology such as SLE. Pericardial effusions can be seen in SLE but are usually small and rarely compromise hemodynamics.[6] However, Kahl[7] reported that 13% of their cohort of 75 patients with SLE pericarditis had tamponade physiology, and Rosenbaum et al[8] reported 21.9% of 41 patients.

Initial conservative management of CP is similar to the treatment of acute pericarditis, typically using a combination of anti-inflammatory agents (colchicine and a nonsteroidal anti-inflammatory drug) for 2 to 3 months. Most cases are self-limited and respond to supportive care, treatment of the inciting etiology, and medication.[9]

A pericardiectomy is the only definitive treatment option for patients with chronic CP who have persistent and prominent symptoms. A correct and early diagnosis of CP is extremely important because successful pericardiectomy can be curative.[9]

Overall survival after pericardiectomy for CP differs significantly among the major etiologic subgroups and is best suited for patients with idiopathic and miscellaneous constriction, intermediate for postsurgical constriction, and poor for postradiation constriction.[10] The excellent survival after pericardiectomy for idiopathic constriction emphasizes that, when an isolated entity, CP can be treated safely with pericardiectomy.

FOLLOW-UP

The patient has remained asymptomatic and had slow resolution of her echocardiographic findings over the past 36 months.

FIGURE 5 Specimens of Visceral and Parietal Pericardium

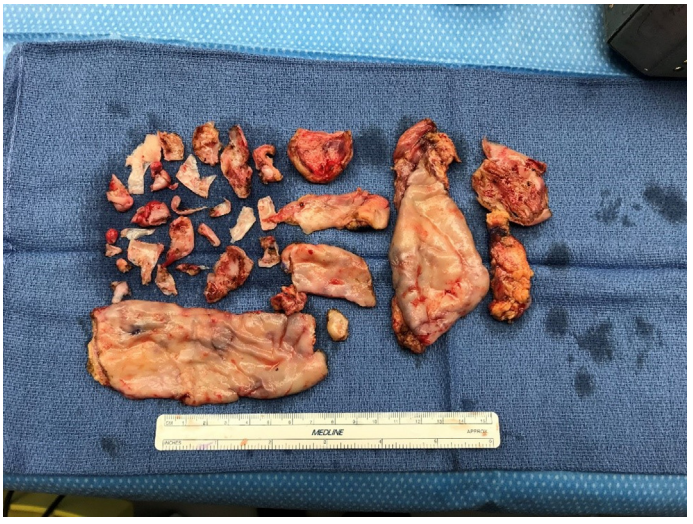
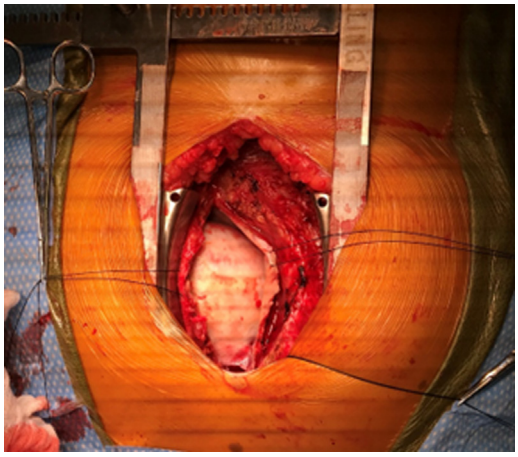


FIGURE 6 Sternotomy Revealing Pericardial Thickening



CONCLUSIONS

In patients with recurrent anasarca and shortness of breath, the diagnosis of CP should be considered. In this case, pericardiectomy with Waffle procedure was the treatment strategy of choice.

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REFERENCES

1. Heimbecker RO, Smith D, Shimizu S, Kestle J. Surgical technique for the management of constrictive epicarditis complicating constrictive pericarditis (the waffle procedure). *Ann Thorac Surg*. 1983;36(5):605-606. https://doi.org/10.1016/s0003-4975(10)60693-5

2. Man BL, Mok CC. Serositis related to systemic lupus erythematosus: prevalence and outcome. *Lupus*. 2005;14(10):822-826. https://doi.org/10.1191/0961203305lu2187oa

3. Ryu S, Fu W, Petri MA. Associates and predictors of pleurisy or pericarditis in SLE. *Lupus Sci Med*. 2017;4(1):e000221. https://doi.org/10.1136/lupus-2017-000221

4. Zhao J, Bai W, Zhu P, et al. Chinese SLE Treatment and Research group (CSTAR) registry VII: prevalence and clinical significance of serositis in Chinese patients with systemic lupus erythematosus. *Lupus*. 2016;25(6):652-657. https://doi.org/10.1177/0961203315625460

5. Strauss AW, Santa-Maria M, Goldring D. Constrictive pericarditis in children. *Am J Dis Child*. 1975;129(7):822-826. https://doi.org/10.1001/archpedi.1975.02120440048010

6. Tincani A, Rebaioli CB, Taglietti M, Shoenfeld Y. Heart involvement in systemic lupus erythematosus, anti-phospholipid syndrome and neonatal lupus. *Rheumatology (Oxford)*. 2006;45(suppl 4):iv8-iv13. https://doi.org/10.1093/rheumatology/kel308

7. Kahl LE. The spectrum of pericardial tamponade in systemic lupus erythematosus. Report of ten patients. *Arthritis Rheum*. 1992;35(11):1343-1349. https://doi.org/10.1002/art.1780351115

8. Rosenbaum E, Krebs E, Cohen M, Tiliakos A, Derk CT. The spectrum of clinical manifestations, outcome and treatment of pericardial tamponade in patients with systemic lupus erythematosus: a retrospective study and literature review. *Lupus*. 2009;18(7):608-612. https://doi.org/10.1177/0961203308100659

9. Welch TD. Constrictive pericarditis: diagnosis, management and clinical outcomes. *Heart*. 2018;104(9):725-731. https://doi.org/10.1136/heartjnl-2017-311683

10. Talwar S, Nair VV, Choudhary SK, et al. Pericardiectomy in children <15 years of age. *Cardiol Young*. 2014;24(4):616-622. https://doi.org/10.1017/s1047951113000814

KEY WORDS constrictive pericarditis, pericardiectomy

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