Chest pain in a child? The diagnosis is NOT always in the bag.

John Martinez, MD; Pierre Wong, MD; Darren Berman, MD; John Wood, MD; Ram Subramanyan, MD, PhD; Jennifer A. Su , MD

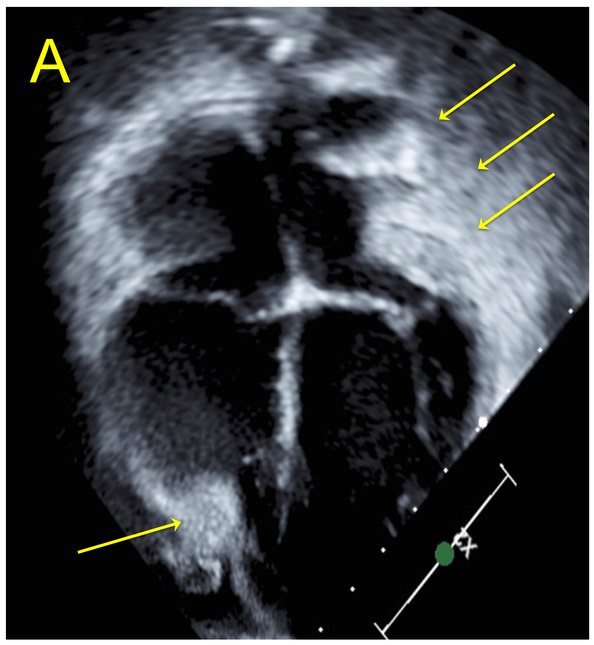
Introduction

We report a case of chest pain in a child, initially thought to be due to a mediastinal mass. The mass was discovered to be a pulmonary sequestration and she was found to have congenital absence of the pericardium as the cause of chest pain due to resultant external compression of her coronary system.

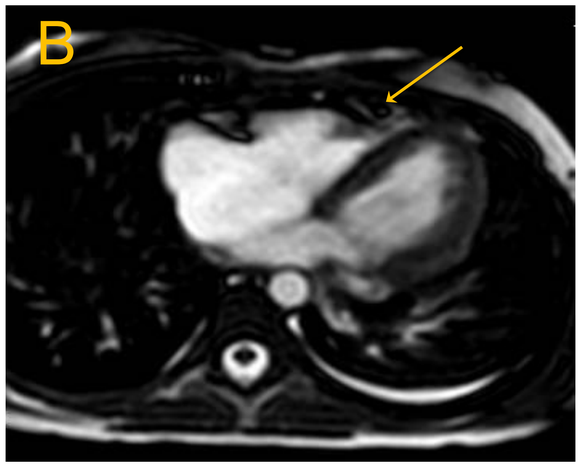
Case Description

A nine-year-old female presented with acute left-sided chest pain and emesis. Her troponin level was elevated at 0.43 ng/ml (normal <0.01 ng/ml). Her EKG showed sinus rhythm with abnormal repolarization pattern and echocardiogram demonstrated normal biventricular function with a curiously shaped right ventricle (RV) initially concerning for an aneurysm (**Figure A**).

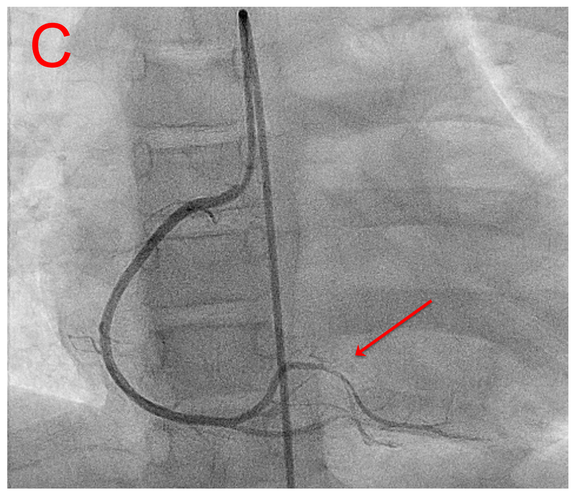
A chest X-ray was obtained and revealed widening of the superior mediastinum raising concern for potential mass lesion. The patient was admitted for evaluation of a mediastinal mass potentially causing extracardiac coronary compression.



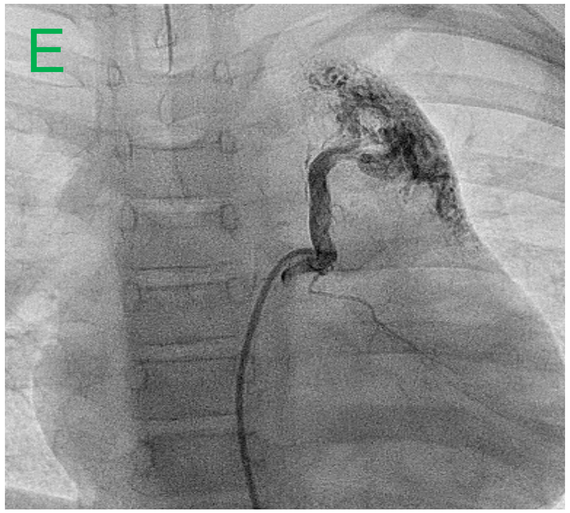
Cardiac MRI was performed revealing that the RV apex was functionally amputated by an external linear compression, suggesting a congenital defect in the pericardium (**Figure B**). The mass lesion appeared to be a pulmonary sequestration, with a left upper lobe consolidation with venous drainage to the innominate and hemiazygos veins, although arterial supply was unclear.

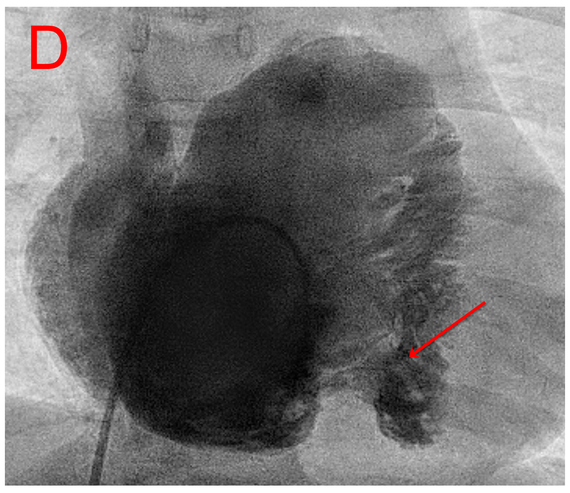


Cardiac catheterization was pursued, and angiography confirmed external compression near the RV apex consistent with a pericardial defect impeding blood flow to the right coronary system (**Figures C and D**).



Aortic angiography demonstrated an arterial vessel arising from the thoracic descending aorta which perfused a left upper anterior lobe pulmonary sequestration (**Figure E**). The patient underwent pericardiectomy and surgical resection of the pulmonary sequestration. Intraoperatively, she was noted to have a very deficient pericardium which covered mainly the superior and inferior venae cavae as well as the right pulmonary veins. The patient recovered from surgery uneventfully and was discharged home with complete resolution of chest pain. She has not had recurrence of symptoms.





Discussion

Congenital pericardial agenesis is a rare entity with a male predominance occurring in less than one in ten thousand.[1] Patients are often asymptomatic, and diagnosis is made incidentally when obtaining imaging for other reasons, such as in the evaluation of a mediastinal mass. These defects are typically left-sided and occur in isolation, though may be associated with other cardiopulmonary lesions, namely, pulmonary sequestrations.[1-4] Sequestrations largely manifest in infancy and are usually located between the left lower lobe and the diaphragm in contrast to the left upper lobe as was observed in our case. This association has been reported previously in adults but is herein presented in a healthy child.[2-4] Symptomatic pericardial defects are often discovered in the evaluation of chest pain secondary to compression of the coronary system by a rim of defective pericardium. Several imaging modalities may aid with diagnosis – however surgical visualization is definitive as imaging relies largely on the presence of indirect evidence of an absent pericardium.[1] Treatment options for symptomatic lesions include total or partial pericardiectomy. [1]

Conclusions

Congenital absence of the pericardium is a rare diagnosis often made incidentally with a known association with pulmonary sequestrations. It should be considered in the differential diagnosis of anterior mediastinal lesions, particularly when associated with chest pain and acute troponin elevation.

References:

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