

Hypertension in a Pediatric Patient With Repeat Aortic Coarctation Repair

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A 15-year-old male adolescent with a history of repeated procedures to correct aortic coarctation presented with hypertensive urgency.

Introduction

Coarctation of the aorta comprises approximately 5% to 8% of congenital heart defects and is often associated with valvular malformations.¹ These defects are typically diagnosed early and are managed with surgical repair, balloon angioplasty, or endovascular stent placement. However, as the following case illustrates, complications can occur in this population despite early intervention.

Case

A 15-year-old male adolescent presented to the pediatric ED after repeated blood pressure (BP) checks by the school nurse revealed consistently elevated systolic and diastolic pressures. The patient's hypertension was associated with symptoms of intermittent headache and light-headedness. His medical history was remarkable for a congenital aortic coarctation and a bicuspid aortic valve. The patient had undergone a subclavian flap repair prior to 1 month of age, followed by a balloon dilata-

tion 1 year later for recurrent coarctation. The rest of the patient's medical history was unremarkable, including normal renal function. He denied illicit drug or alcohol use, sexual activity, or trauma.

On evaluation, the patient's cardiac examination revealed a regular rate and rhythm with normally split S2; there were no rubs, murmurs, or gallops on auscultation. He had normal and equal pulses of the upper and lower extremities bilaterally. The patient presented without cyanosis. He was alert and oriented with normal upper and lower extremity reflexes. The neurological examination, including cranial nerve, strength, and gait testing, was unremarkable. The gastrointestinal examination showed a soft, nondistended abdomen, with no pulsatile masses. There was no abnormal swelling of his extremities. Although the physical examination findings were unremarkable, the patient's vital signs were concerning as BP in his right upper extremity was as high as 208/110 mm Hg, while BP in his right leg was 130/68 mm Hg.

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DOI 10.12788/emed.2016.0010

The patient was given oral acetaminophen for his headache, after which there was significant improvement in his upper extremity BP. His chemistry panel, including creatinine levels, liver function, and urine electrolytes, were all within normal values. An electrocardiogram was significant for left ventricular hypertrophy without arrhythmia or conduction abnormalities (**Figure**). After discussion with the patient's cardiologist, he was discharged home in the care of his parents, who were instructed to have close outpatient follow-up with the cardiologist and undergo advanced imaging studies.

The patient followed up with his cardiologist, who ordered cardiac magnetic resonance imaging (MRI). The MRI showed mild narrowing of the distal aortic arch with a minimal and clinically insignificant pressure gradient. Based on the MRI findings, the patient was referred to a pediatric nephrologist, who performed a 24-hour ambulatory BP evaluation. The results of this study showed the patient to have systolic hypertension at the 95th percentile for his age and height. Based on the patient's athletic predilection, β -blockers were avoided, and he was instead started on the angiotensin-converting enzyme (ACE) inhibitor lisinopril, along with annual follow-up cardiac evaluation.

Discussion

The authors' initial concern for this patient was the possibility of a recurrent coarctation causing a significant pressure gradient between the upper and lower extremities with associated symptoms. A review of the literature demonstrates such an occurrence is not uncommon in this patient population, especially in patients with a history of early intervention (ie, within the first year of life).²

Causes and Incidence

One of the factors believed to contribute to recurrent coarctation is insufficient growth versus retraction of the manipulated tis-

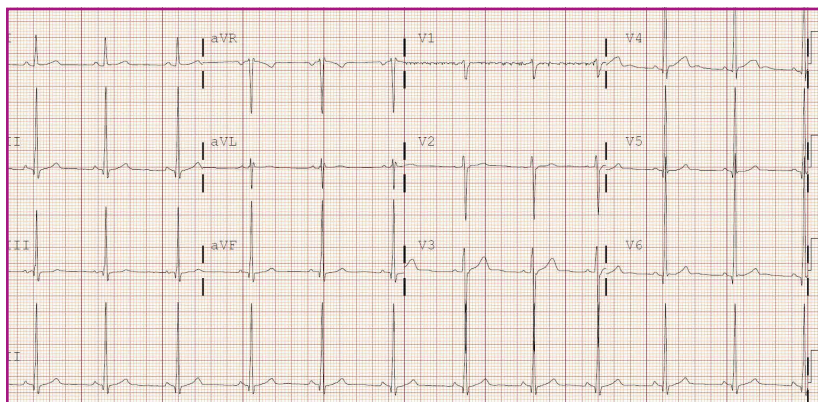


Figure. Electrocardiogram of patient demonstrating left ventricular hypertrophy but no signs of arrhythmia or conduction abnormalities

sues over time. The rates of recurrence vary based on the initial technique used for repair. These recurrences have been found to be approximately 6% in patients who had subclavian flap repairs; 31% for those who had balloon angioplasty alone; and approximately 20% in patients who had aortic stenting.³⁻⁵ As seen in this case, balloon angioplasty is usually performed in patients requiring revascularization. However, up to 32% of these patients will require further intervention due to subsequent recurrence.⁶

Evaluation

Although emergency physicians (EPs) have numerous diagnostic modalities available to evaluate patients with suspected aortic coarctation, as long as the patient is in no acute distress, much of the work-up can be performed on an outpatient basis—in conjunction with the primary- and subspecialty-care team. Regarding appropriate imaging modalities, echocardiography with Doppler or 3D reconstruction of MR angiogram can be useful in detecting both anatomical abnormalities as well as the associated gradient dysfunction; computed tomography can be used for assessing the anatomy.⁷ All of these modalities can also be used to evaluate late-term complications of aortic coarctation pathology, including aortic aneurysms. To help ensure

good outcome, the EP should always keep the possibility of recurrence in the differential when evaluating these patients, regardless of the number of previous interventions attempted.

Hypertension

As this case illustrates, patients with a history of coarctation repair often develop high BP. Unfortunately, up to 23% of these patients will go on to have BP above the 95th percentile.⁵ Moreover, a significant number of patients in this population will also suffer from exercise-induced hypertension, even when at-rest BP is controlled with antihypertensive medications.⁸

β -blockers, angiotensin-receptor blockers, and ACE inhibitors are considered the first-line medications for hypertension in adults and adult-sized patients with this condition.⁹

Since a high proportion of patients as young as age 7 years may develop high BP postrepair,¹⁰ the EP should discuss the initiation of an antihypertensive agent with the patient's care team prior to discharge. It is also important to keep in mind that elevated BP is present to a significant degree even in patients without recurrent obstruction. The negative sequelae associated with uncontrolled hypertension is well known, and patients with congenital anatomical anomalies are at higher risk for such negative outcomes.

Conclusion

This case illustrates a common presentation of a teenaged patient with a chronic medical condition due to a corrected congenital cardiac defect. It also demonstrates the unique and early opportunity the EP has to evaluate and provide appropriate intervention for patients with potentially life-threatening diseases.

Patients with a history of corrective vascular surgery due to congenital heart

malformations are an at-risk population. Therefore, during evaluation, the EP should always keep in mind that these patients have a higher prevalence of related abnormalities at earlier ages than the general population. Steps initiated in the ED prior to discharge, in collaboration with the patient's primary- and specialty-care team, can assist in expediting appropriate outpatient management of any sequelae. If a patient does not have a cardiologist, a referral to one should always be made prior to discharge.

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