Hypertension as a Presentation of Coarctation of the Abdominal Aorta

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Introduction

Coarctation of the aorta is a narrowing of the descending aorta. It is typically located at the insertion of the ductus arteriosus just distal to the left subclavian artery; however, there is a spectrum. Narrowing can be located at any location along the aorta, more rarely the abdominal aorta. Coarctation accounts for 4-6% of all congenital heart defects with a reported prevalence of approximately 4 per 10,000 live births.

There is a wide variety in timing and how a patient can clinically present with coarctation of the aorta. If there is a severe narrowing that is dependent on a patent ductus arteriosus (PDA), patient is likely to present within one week of life when PDA closes. When this happens there is an abrupt decrease in systemic circulation and an increase afterload on the heart. This can lead to cardiogenic shock and require a more emergent intervention. If narrowing is less severe and adequate systemic flow is maintained, then presentation can occur later in life. A common presentation in these cases could be unexplained hypertension or pain in legs with exercising (claudication). Both presentations will commonly have a delay in femoral pulses.

Coarctation of the aorta is treated via surgical repair or balloon dilation of narrowed segment of aorta. Repair is recommended for indications listed in Table 1.

Case

A 2-year-old female presented to the ED with four days of fever with minimal cough and congestion. Upon physical exam, patient was found to have blood pressure of 156/100. With multiple attempts, blood pressure remained elevated and patient was transferred to children's hospital.

On arrival, patient had physical exam that was significant for weak femoral pulses bilaterally and a 3/6 systolic murmur heard over the midclavicular line on anterior chest. **Vital signs were notable for a large pressure gradient: upper extremity BP of 154/102, lower extremity BP of 83/55**. Patient had a normal CBC, CMP, UA. Renin and aldosterone, in addition to urine metanephrines were all within normal limits. Patient had an ECHO that showed no abnormalities in the heart anatomy or vessels. Renal U/S and Doppler also showed normal anatomy and vessels. However, radiology commented that observed turbulent blood flow and an inability to assess the full aorta may be indicative of a possible abnormality of the abdominal aorta.

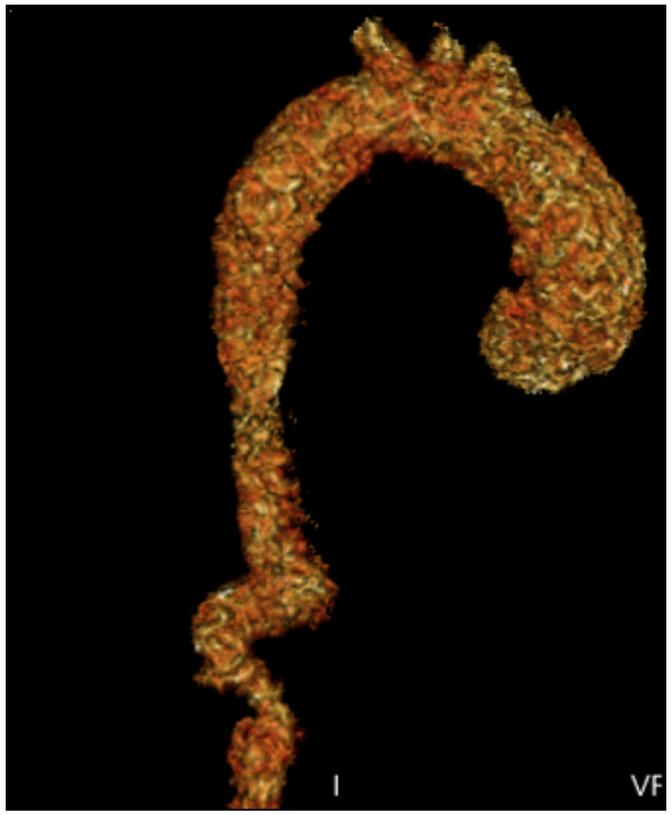

Figure 1 – Patient CT angiogram presenting "corkscrew" deformity of descending aorta

Table 1 – Indications for Repair

- **Critical coarctation (with little to no blood flow to systemic circulation)**
- **Coarctation gradient >20 mmHg**
- **Radiologic evidence of clinically significant collateral flow**
- **Systemic hypertension attributable to coarctation**
- **Heart failure attributable to coarctation**

Case (cont.)

Patient had CT angiogram (illustrated in Figure 1) completed which showed "corkscrew"/spiral deformity of the descending aorta. Patient was discharged and returned for surgery within one week. Patient underwent left posterolateral thoracotomy, repair of aortic coarctation with 14 mm GorTex jump graft, and is currently following up with cardiology outpatient. Patient was able to be weaned off antihypertensives within 1-2 months post surgery. Patient was indicated for surgery based on coarctation gradient >20 mmHg and systemic hypertension.

Discussion

Hypertension is a common presentation of less severe coarctations of the aorta, especially once patient is older than the neonatal period. Family reported to team that patient was being followed by outpatient primary care physician for hypertension, that lifestyle modifications were initiated, and follow-up was planned.

Essential hypertension is the most common etiology for hypertension in the pediatric population. The current suggestion by the U.S. Department of Health and Human Services (Flynn et al., 2018) is that children >6 years old do not need extensive work up for hypertension if they have positive family history, obesity on exam, or any other concerning physical exam finding for secondary hypertension. Given this patient's age (<2 years old) and physical exam finding of delayed femoral pulses, she would benefit from a further work up for secondary hypertension, with focus on coarctation of the aorta, which we initiated.

It is important to remember that classification of hypertension in the pediatric population is based on age, gender, and height. There are three stages of HTN: Elevated BP, Stage 1, and Stage 2. Each stage has a different recommendation for follow up and when further testing should be initiated. Our patient had Stage 2 hypertension on admission.

References

Flynn, J. T., Kaelber, D. C., & Baker-Smith, C. M. (2018). Subcommittee on Screening and Management of High Blood Pressure in Children. Clinical Practice Guideline for Screening and Management of High Blood Pressure in Children and Adolescents (vol 140, e20171904, 2017). *Pediatrics*, *142*(3).

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