

Strong Pursuit of the Diagnosis Despite its Attempt to Evade: Neonatal Critical Coarctation of the Aorta

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Abstract

Coarctation of the aorta is a congenital heart disease that can be found in various age groups but is often missed in the neonatal period. Neonates can present in severe distress or some may even be asymptomatic and present decades later into their life. We present a patient that presented in severe respiratory distress and in cardiovascular shock and after fervent pursuit of the diagnosis, revealed itself to coarctation of the aorta.

Keywords: *Coarctation; Aorta; Congenital Heart Disease*

Introduction

Coarctation of the aorta accounts for 5 - 8% of congenital heart disease and is the most commonly undiagnosed form as well [4]. This can be diagnosed in utero and often lends a better prognosis than if presented antenatally. If diagnosed antenatally, it can often be missed in the first 2 days after discharge from the nursery as the ductus arteriosus has yet to close and only half will present with a murmur [3]. Depending on how rapidly and how dependent the lesion may be, the neonate can present with mild symptoms of tachypnea, poor feeding, poor perfusion to cardiovascular collapse and congestive heart failure [3,4].

Case Presentation

Our patient was a 10-day old male who presented to the emergency department in respiratory distress. Mother had prenatal care given only by a local midwife throughout his pregnancy and was told that the pregnancy was going well and had no issues. He was delivered at 37 weeks of gestation in his home via water birth in the bathtub. Amniotic fluid per mother was thought to be yellow. No vital signs were monitored during the delivery process. No newborn screening was obtained.

Parents report that he was doing well until 5 days of life when they noticed his respiratory distress. He started having lethargy and had a weaker cry. They brought him to see the midwife at which point, she again said that everything was expected. At 7 days of life, he started having increased labored breathing and had poor feeding. Parents also reported diaphoresis, frequent tiring with feeding, cold extremities, rib retractions and cola-like urine.

On physical exam, the newborn appeared lethargic, mottled and pale. He had a soft and sunken anterior fontanelle and icteric conjunctivae. His mucous membranes were dry. He had decreased air entry and bilateral crackles could be heard diffusely. He had subcostal, intercostal and suprasternal retractions without rhonchi or wheezing. He was tachypneic at 70 bpm with end expiratory grunting. He was tachycardic at 170 bpm and without murmur. His PMI was at the 5th intercostal. He had delayed capillary refill of 5 seconds with very

thread-like and weak brachial pulse. Femoral and dorsalis pedis pulses were absent. He was very diaphoretic. His abdomen was soft, non-tender but was mildly distended. He had normal bowel sounds. His liver edge could be palpated about 5 cm below the costal margin. He did not have splenomegaly. He did not have any gross deformities, cyanosis, ecchymosis or purpura. His cranial nerves II-XII were grossly intact.

He was afebrile. 4 limb blood pressure showed 100/69 with MAP 99 on left arm, 133/82 with MAP 102 on right arm, 55/37 with MAP 42 on left leg, and 57/30 with MAP 36 on right leg. CXR showed cardiomegaly with severe bilateral pulmonary edema. His capillary blood gas showed severe metabolic acidosis with an HCO₃ of 12 and a base excess of -15. He also had elevated lactic acidosis of 10.8. LDH was vastly elevated at 821. Given the CXR findings, hepatomegaly as well as metabolic acidosis, his clinical presentation was consistent with severe congestive heart failure. Given these findings, patient was started on inotrope infusion and within 30 minutes, patient had improved peripheral circulation and stronger pulses. An echocardiogram was immediately ordered to further evaluate as the most likely differential at this time was an interrupted aortic arch or aortic coarctation given the significant difference in the 4-limb blood pressure. Initial echocardiogram had shown a wide-open arch up to the branch of the mainstem left bronchus and then the arch was opened just distal to that again without confirming an aortic abnormality. There was severe depressed left ventricular function with ejection fraction of 11%. Patient was continued on inotropes and as his cardiovascular collapse had improved, due to our strong clinical suspicion, we again asked the pediatric cardiologist to once more interrogate the aorta. A second echocardiogram was done and at this point, showed critical coarctation of the aorta, hidden behind the left mainstem bronchus. The patient was immediately begun on prostaglandin and diuresis was begun. Hepatomegaly and cardiomegaly had improved. Patient was then transferred to another hospital for higher level of care where he had an end-to-end coarctation repair via lateral thoracotomy. He continues to follow up with our cardiology clinic and has been progressing well.

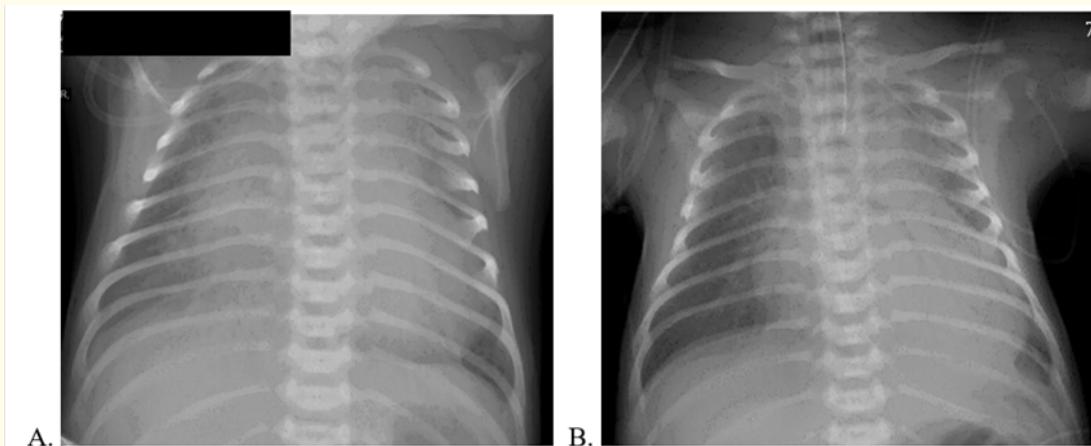


Figure 1: A) CXR at presentation. B) CXR after inotrope support and diuresis.

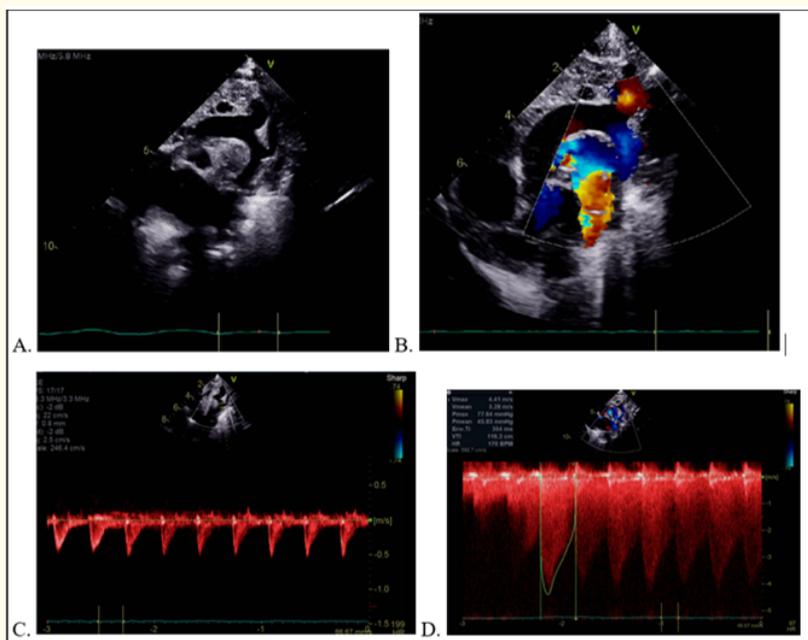


Figure 2: A) Coarctation of the distal part of thoracic aorta. B) Echocardiogram with doppler showing constriction. C) Doppler wave form before inotrope support. D) Emergence of saw tooth pattern due to increased pressure gradient after inotrope support.

Discussion and Conclusion

Coarctation of the aorta is a narrowing of the aorta most commonly found distal to the left subclavian artery and accounts for 5 - 8% of all congenital heart disease [2,3]. Interrupted aortic arch is the most severe form and is classified based on where the arch is blunted [2]. We found this to be a fascinating case that highlights the importance of clinical suspicion and while we must explore all available differential diagnosis, if the clinical signs and symptoms all point towards a diagnosis, how resolute we must be to pursue it.

Coarctation of the aorta often presents after the patient is discharged from the newborn nursery where most babies appear well on the first few days. At 2 - 3 weeks of life, the PDA closes and causes the ductal dependent aortic coarctation to have a sudden increase in left ventricular afterload leading to eventual left ventricular dilatation and congestive heart failure [1,3]. Those with preductal defects tend to present earlier in life and with more severe symptoms as compared with post-ductal coarctation more commonly appear late into adult life [1]. Neonates can often present with lethargy, tachypnea, paleness, cool extremities, poor feeding as well as decreased urine output. Cardiac murmur may not be present in up to 50% affected infants but a harsh systolic ejection murmur may be auscultated along the left sternal border with radiation to the back, especially over the affected aortic segment. Carotid, brachial and radial pulses may be strong while femoral, popliteal and dorsalis pedis pulses may be weak or even absent [3]. This can progress to cardiovascular collapse, heart failure and metabolic acidosis [4].

Ancillary studies to diagnose coarctation of aorta include 4 limb blood pressure where a pressure gradient > 20 mmHg is significant for aortic narrowing although this method lacks specificity [1,3]. CXR can show cardiomegaly and pulmonary congestion. EKG can be normal at first then show rightward axis deviation due to right ventricle hypertrophy from increased afterload. Prenatal ultrasound may show a fetus with a hypoplasia of the left heart in severe cases but may be completely normal in mild cases leading to delayed diagnosis and worsened mortality and morbidity on such patients [4]. Transthoracic echocardiograph is the confirmatory test for diagnosis and to identify the narrowed aortic defect.

Management of such affected neonates should begin with cardiovascular resuscitation including inotropes and prostaglandin to keep PDA open. If patient is asymptomatic, patient may be monitored conservatively in lieu of an elective surgery but in a case of critical coarctation like ours, patient must be surgically corrected at the earliest possible time.

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