Case Report
Central Eur J Paed 2022;18(1):38-43
DOI 10.5457/p2005-114.314

Atresia of the Abdominal Aorta Presenting as Neonatal Arterial Hypertension: a Case Report with Literature Review

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Received: November 24, 2022; **Accepted:** January 16, 2022

Abstract

Objective – We present a case of neonatal arterial hypertension in a patient with atresia of the abdominal aorta diagnosed after an extensive diagnostic workup, emphasizing the importance of imaging methods in the detection of vascular malformations. The article reviews the vascular causes of neonatal arterial hypertension. **Case Report** – A case of a 5-day-old boy born after 38 weeks of gestation who was admitted to a tertiary level medical center due to arterial hypertension, tachypnea, and a heart murmur was reported. Abdominal Doppler US revealed high flow paraspinal vessels and huge bilateral subcutaneous collaterals extending from the pelvis towards the upper abdomen. Common iliac arteries and abdominal aorta at the level of renal arteries and below could not be differentiated. The main differential diagnostic options were stenosis of the abdominal aorta or paraspinal arterio-venous malformation. Magnetic resonance angiography revealed atresia of the abdominal aorta, with large caliber bilateral paraspinal collaterals, feeding the lower part of the body. Surgical treatment wasn't indicated at that time. At the present the patient is four years old, developing normally on antihypertensive therapy. **Conclusion** – Neonatal arterial hypertension is rarely idiopathic, thus identifying a possible underlying illness becomes imperative. Imaging methods have a pivotal role in the early detection of the causes of neonatal arterial hypertension.

Key Words: Arterial Hypertension ▪ Atresia Abdominal Aorta ▪ Ultrasound ▪ Magnetic Resonance Angiography ▪ Newborn.

Introduction

Neonatal arterial hypertension (AH) is defined as systolic blood pressure of at least the 95th percentile for gestational age, birth weight, and sex on three separate measurements (1). It occurs in up to 3% of neonates admitted to the Neonatal Intensive Care Unit. It is a potentially under-recognized condition due to scarcity of normative data on neonatal blood pressure values, exclusion of neonates from clinical trials of antihypertensive medications, and relative rarity of the condition (2, 3). Even though neonatal AH is rare, the list of causes is vast. Most common are umbilical artery thromb-emboli, followed by bronchopulmonary dysplasia, and vascular causes such as coarctation of the thoracic aorta (4). Clinical manifestation of neonatal AH is varying, the most common being a "silent" presentation as an asymptomatic neonate that has elevated blood pressure which is not life-threatening (4). Additional symptoms and clinical features of neonatal AH usually overlap with other illnesses such as apnea, muscular hypertonia, cyanosis, vomiting, seizures, etc. In rare cases, neonatal AH can present as congestive heart failure, hematuria, hypertensive retinopathy, and failure to thrive (5).

Neonatal AH is rarely idiopathic, usually, it is a result of an underlying condition, thus it is vital to distinguish the cause and determine the best method of treatment (4). A multidisciplinary approach is necessary for a definitive diagnosis, ranging from clinical observation, followed by laboratory tests, and imaging, most commonly ultrasound (US) and magnetic resonance angiography (MRA). In detecting the causes of neonatal AH, imaging has a pivotal role.

The purpose of this paper is to present a case report and review of the literature in the diagnosis of neonatal AH due to vascular abnormality.

Case Report

A 5-day-old boy was born after 38 weeks of gestation, to Caucasian nonconsanguineous parents. The pregnancy proceeded normally, without any description of pathomorphological changes in the fetus on the prenatal US.

His birth weight was 2760 g (10th percentile), birth length 48 cm (10th percentile), head circumference 33.5 cm (25th percentile), and Apgar score 10/10. He was admitted to a tertiary level medical center due to suspected sepsis and a heart murmur (a 2/6 systolic-diastolic murmur). At the age of 30 hours, he became tachypneic with a hyperactive precordium. Echocardiography revealed opened ductus arteriousus, aortic regurgitation, and hemodynamically insignificant foramen ovale. His arterial blood pressure was higher, ranging from 100/75 to 130/100 mmHg (90th percentile for a newborn 92/65 mmHg). His chest X-ray was normal. Abdominal Doppler US revealed high flow paraspinal vessels. In addition, huge bilateral subcutaneous collaterals extending from the pelvis towards the upper abdomen were seen (Fig. 1). Femoral arteries were normal, but the abdominal aorta below renal arteries and pelvic arteries could not be seen. Differential diagnostic options were stenosis of the abdominal aorta or paraspinal arterio-venous malformation (AVM). MRA revealed the atresia of the abdominal aorta, with large caliber bilateral paraspinal collaterals, bigger on the right side, feeding the lower part of the body (Fig. 2). The right renal artery originated from the right internal iliac artery, the left renal artery from collaterals at the same level. An extensive collateral system between both external iliac arteries and the epigastric arteries and the internal thoracic artery was seen. Antihypertensive therapy with oral amlodipine 0.05 mg/kg/12 hours was started immediately. The blood pressure ranged from 76-123/51-98 mmHg and the amlodipine dose was gradually raised to 0.2 mg/kg/12 hours. He had some minor facial dysmorphic features (sloping forehead, hypotelorism), toenails hypoplasia, mild muscular hypotonia, and transient hypercalcemia. His hearing, thyroid function, and ocular status were normal. Chromosomal microarray analysis was performed but did not confirm any chromosomal microduplication or microdeletion. Parents refused any further molecular genetic investigations. A surgical procedure was not possible or indicated at that age.

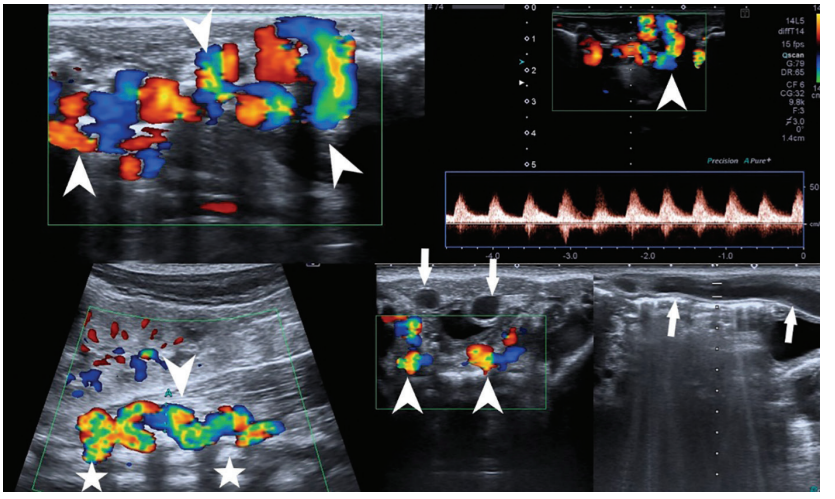

Fig. 1. *Ultrasound with Doppler of the abdominal vessels showed tortuous paraspinal vessels with a right-side predominance (arrowheads) and two thick collaterals originating from iliac arteries (arrows) and running cranially in the anterior abdominal wall (star – along the spine).*

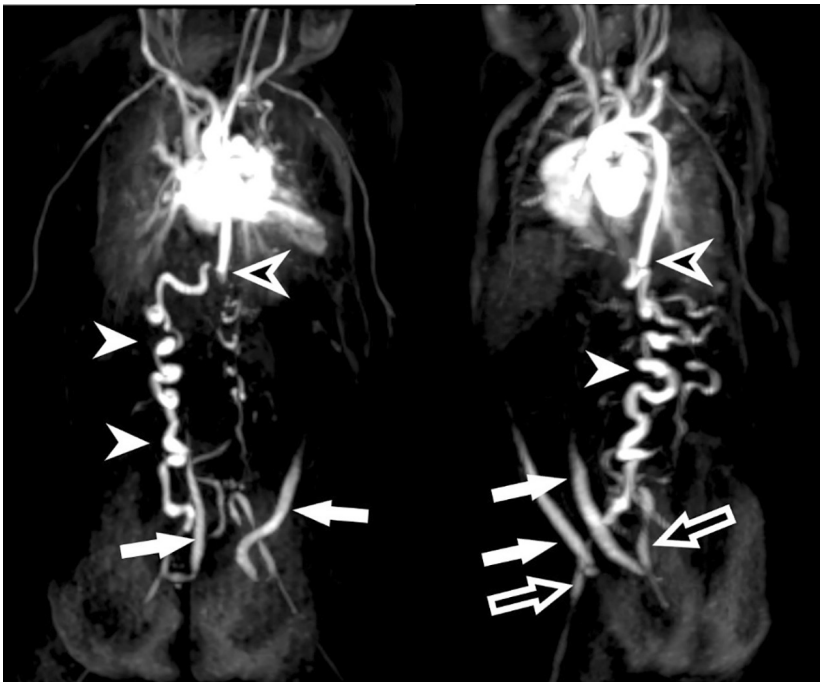

Fig. 2. *Magnetic resonance angiography: note the abrupt ending of the abdominal aorta (arrowheads), no abdominal aorta and common iliac arteries, but the presence of external iliac arteries (arrows), paraspinal collaterals (white arrowhead), and collaterals in the arterial abdominal wall (white arrows).*

He was regularly followed up clinically by measuring arterial blood pressure and by echocardiography which showed slow progress of left ventricular hypertrophy. Amlodipine dose was adjusted according to the baby's weight and at the age of eight months, metoprolol was introduced. At present, the boy is almost 4 years old. He is treated regularly with amlodipine 0.2 mg/kg/12 hours and metoprolol 0.7 mg/kg/12 hours. His arterial blood pressure is 100-105/65 mmHg (90. percentile for his height and age is 105/61 mmHg). His left ventricle is still hypertrophic. His neurodevelopmental status is normal.

Discussion

Since its first description in the 1970s, neonatal AH has become an emerging challenge for neonatologists and pediatricians (6). Despite the advances in medicine, a lack of clear guidelines of management when compared to the adult population still represents a great challenge and is formed mainly on case series and expert opinions (4, 6).

The confirmation of the diagnosis of the neonatal AH should be based on the detection of elevated blood pressure in all four extremities on three separate occasions taking into consideration antenatal, perinatal, and postnatal factors, which affect the neonatal blood pressure (3, 4). In most cases diagnosing neonatal AH warrants identifying the underlying illness. A detailed history provides information about the most common cause for neonatal AH i.e. umbilical catheter placement (1, 4). Physical examination should be performed focusing on cardiovascular, abdominal, and genitourinary systems, which may in combination with general appearance indicate the primary etiology of neonatal AH as well as to detect its effects on organ dysfunction (3). A laboratory evaluation includes serum electrolyte status, blood urea nitrogen, creatinine, blood gas analysis, pH, bicarbonate, plasma renin activity, aldosterone, cortisol, thyroid hormones, and urinary catecholamines (4).

In our case, the patient had a negative family history and didn't have the umbilical catheter placed at any time since birth. The physical examination showed mild dysmorphic features, though the chromosomal microduplications or microdeletions were excluded and his further neurodevelopmental status was adequate. The presentation of tachypnea elevated blood pressure, and a hyperactive precordium several hours after birth in combination with a heart murmur warranted further diagnostic: a chest X-ray focusing on the cardiac status, echocardiography, the abdominal US including Doppler of the aorta and renal vasculature, and MRA for the

evaluation of big vessel malformations (4, 7, 8). In our case, the normal chest X-ray and described findings on echocardiography narrowed the differential diagnosis to an underlying vascular condition.

Vascular causes responsible for the development of neonatal AH generally include conditions that cause narrowing of the abdominal aorta that consequently leads to renal function impairment. The narrowing can be diffuse or segmental. A diffuse narrowing is seen in hypoplasia or in the atresia of the aorta where the abdominal aorta does not develop at all (8-12). A segmental narrowing is seen in the coarctation of the abdominal aorta. Occlusion secondary to the thromboembolic phenomenon (8), aneurysm of the abdominal aorta (14), and midaortic syndrome (15) are also described as causes for narrowing. Only a few case reports were published in the literature describing the abovementioned morphological and anatomical changes of the abdominal aorta (8-12).

Based on Doppler US findings of very prominent paraspinal vessels two differential diagnoses emerged in our case; paraspinal AVM and hypoplasia of the abdominal aorta. The paraspinal AVM was less likely due to the male sex of the infant (the AVM carries a female predominance) and the absence of neurological impairment. Abdominal aorta and iliac vessels could not be traced in the whole length during the examination also because of bowel air superposition (13). However, paraspinal collaterals and the thick subcutaneous bilateral collaterals in the abdominal wall originating from the external iliac artery suggested, that the most likely cause of AH was hypoplasia of the abdominal aorta. MRA was performed as the method of choice and confirmed that atresia of the abdominal aorta was the cause of AH. In addition, it provided detailed information about the anatomical course of the collaterals.

Atresia of the abdominal aorta is an extremely rare cause of neonatal AH. The most likely explanation for hypoplasia/atresia is either over-fusion or failure in the fusing of two embryonic dorsal aortas (arisen from the endocardial heart tube around the 18th day of embryonic folding), which migrate towards each other and fuse with the loss of their intervening wall around the 25th day of development (8). In this aspect, patients with an abdominal aortic anomaly need to be evaluated for other underlying syndromes or diseases as certain conditions have been known to be associated with the aortic pathology (8). Williams syndrome (elastin gene mutation) for instance, as well as some vasculopathies (such as Takayasu's arteritis, neurofibromatosis, and fibromuscular dysplasia), viral-mediated injury of mesodermal tissues during embryonic development (rubella) to name some from the vast list of causes (8). Even a case report of hypomelanosis of Ito and a separate case report of vanishing twin syndrome was described in association with hypoplasia of the abdominal aorta (8).

The etiology of atresia of the abdominal aorta in our case is still not clear, partly also due to the parents' refusal of further genetic tests, including whole-exome sequencing, which could rule out syndromic diseases. However, the general appearance and presence of other organ anomalies strongly suggest an underlying condition. According to the literature, the prognosis of infants with atresia of the abdominal aorta is poor as the long-term complications depend mostly on the level of atresia and the extent of collateralization. The management of neonatal AH associated with atresia of the abdominal aorta is somewhat challenging and should be tailored to each infant's clinical status, in consultation with a pediatric nephrologist and cardiologist (1). Treatment should be directed to resolve the underlying cause of neonatal AH e.g. surgical treatment, if possible. Surgical treatment of renovascular causes of neonatal AH has some merit as it offers definitive treatment. The choice of the optimal surgical technique depends on the anatomy, location, and state of the affected and collateral vessels as well as the child's age and clinical condition. The new surgical techniques include mesenteric artery growth which improves circulation, tissue expander–stimulated lengthening of arteries, aortic-bypass using polytetrafluoroethylene, renal artery reimplantation, and autotransplantation (16). In our case, the surgery at that moment was not an option due to good conservative management of the

neonatal AH, the child's clinical condition, and the complex anatomy of vessels.

If the underlying cause cannot be resolved, the antihypertensive treatment should be carefully titrated from intravenous (as a first-line treatment to prevent organ damage in cases of extremely high blood pressure) to oral doses. The problem arises when selecting an optimal antihypertensive regimen, as most of the commercially available medications were not tested on neonates and exhibit unpredictable pharmacokinetics and pharmacodynamics in neonates (6). When discharged with antihypertensive treatment, home blood pressure monitoring is a good tool for the supervision and adjustment of therapy doses between visits to the doctor as long–term monitoring of infants with neonatal AH is essential (3). Sometimes even despite all treatment options (medicamentous and surgical) the neonatal AH can persist and lead to early death (3, 8). However, the child in our case is four years old and he is doing well, developing physically and mentally in normal ranges. Neonatal AH is under control by antihypertensive treatment with stable, non-progressing left ventricle hypertrophy.

Conclusion

Early etiological determination of neonatal AH is important and imaging plays a crucial role in detecting the possible causes. US with Doppler is the first-line imaging method, which directs further examinations. MRA can differentiate different types of congenital and iatrogenic changes of the abdominal aorta and its branches and thus has an important impact on treatment management.

Authors' Contributions: Conception and design: DK; Acquisition, analysis and interpretation of data: VPT, NB, GN, and DK; Drafting the article: VPT and DK; Revising the article critically for intellectual content: VPT, NB, GN and DK; Approved final version of the manuscript: VPT, NB, GN, and DK

Conflict of Interest: The authors declare that they have no conflict of interest.

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