Bowel ischemia in an infant with unspecified renovascular hypertension: Case report

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Abstract:

**Introduction:** Renovascular hypertension due to congenital multiple visceral arterial stenoses in neonates is rare. Management is challenging and has not been standardized. Medical control of blood pressure (BP) remains the first-line therapeutic approach. However, unwise control of BP in such cases may lead to disastrous situations.

**Case presentation:** We present a female neonate with hypertension due to unspecified vascular occlusive disease. The hypertension was managed medically by maintaining BP at “near normal levels” which led to bowel ischemia. The patient survived the short bowel syndrome and is now 2 years old. She is on full oral feeding and has reached acceptable growth parameters. Her BP has stabilized at around 110/70 mmHg without antihypertensive drugs. The patient has good organ functions and walks despite increased narrowing in stenotic areas and complete obliteration of the left iliac and femoral arteries seen on follow up CT angiography.

**Conclusions:** We suggest keeping BP at the highest levels permissible in similar clinical situations to prevent a state of bowel hypoperfusion. When alternative treatments for congenital multiple visceral arterial stenoses are not feasible, careful medical therapy and waiting approach for collaterals to develop may be appropriate.
**Introduction:**

Renovascular disease is an important cause of hypertension in children, with a reported incidence of 3-10% [1-5]. Medical management remains the first line of treatment [1-9]. However, “unwise” control of BP may lead to disastrous situations. We present the case of an infant with congenital multiple visceral arterial stenoses in which medical therapy contributed to the development of bowel ischemia.

**Case presentation:**

As per history obtained from the parents a full term female with unremarkable prenatal and family history who was born via normal spontaneous vaginal delivery with a birth weight of 2.5 kg was admitted to a provincial hospital at the age of 18 days with cardiogenic shock. She was diagnosed as having high BP, was treated as a case of cardiomyopathy, and was discharged home after 10 days on propranolol and captopril. Without a medical report from the previous hospital she was brought by her parents to our pediatric emergency room at the age of 45 days complaining of lethargy and poor oral intake. The patient looked malnourished and hypoactive. Her weight was 2.4 kg. She had a high systolic BP of 114 to 178 mmHg and diastolic BP of 57 to 82 mmHg. She was admitted to the pediatric intensive care unit (PICU) and required daily doses of 1.8 mg hydralazine, 9 mg propranolol, and 12 mg captopril to control her BP in a range of systolic 85 to 142 mmHg, and diastolic 43 to 75 mmHg. Laboratory studies showed the following: white blood cell (WBC) count of 32.8X10^9/L; hemoglobin of 102 g/L; platelets at 804X10^9/L; erythrocyte Sedimentation rate (ESR) of 2 mm/h; normal renal and liver profiles; normal urinalysis; serum renin of 625 nmol/L; serum cortisol of 526
nmol/L; and growth hormone of 58 µg/L. Echocardiography showed severe nonobstructive hypertrophy of both ventricles, with normal cardiac function. Doppler ultrasound of renal arteries revealed severe bilateral renal artery stenosis with a peak systolic velocity (PS) of 250 cm/sec and a resistive index (RI) of 0.89 (Figure 1). CT angiography revealed multiple arterial stenoses involving both renal arteries near the ostium (Figure 2), the superior mesenteric artery (Figure 3), the celiac artery, the hepatic artery, and both femoral arteries. The patient was stabilized on daily doses of 6 mg hydralazine and 9 mg propranolol to control her BP in a range of systolic of 97 to 114 mmHg and diastolic of 39 to 54 mmHg. The patient was discharged home on these medications with a plan to perform percutaneous transluminal angioplasty (PTA) of the stenosed arteries when she reached a weight of 5 kg. Two weeks after discharge, the patient presented to the pediatric emergency room septic with greenish vomiting, blood per rectum, and pneumoperitoneum. Laparotomy revealed bowel necrosis involving the ileum, cecum, and ascending colon. The necrosed bowel was resected, and a jejunostomy with a mucus fistula at the transverse colon was created. The multidisciplinary team treating the patient included a pediatric surgeon, a pediatric intensivist, a pediatric nephrologist, a pediatric gastroenterologist, a pediatric geneticist, a pediatric rheumatologist, a pediatric radiologist, and an interventional radiologist. The patient stayed in the hospital for about 8 months. Genetic analysis was normal, and metabolic disorders were ruled out. The skin biopsy was normal. The short bowel syndrome was managed successfully. The stoma was closed with a small-bowel-to-transverse-colon anastomosis. When the patient reached a weight of 5 kg, two attempts to perform PTA failed because of the very small caliber of the femoral arteries. During this 8-month-
period, all efforts were directed at keeping the systolic BP between 115 mmHg and 150 mmHg to prevent further episode of bowel hypoperfusion. Later, the patient was discharged on full oral feeding, with hydralazine 2 mg orally every 8 hours and propranolol 3 mg orally every 8 hours as needed if the systolic BP exceeded 150 mmHg. At present, the patient is 2 years old with normal cardiac, liver, and renal functions. She is on full oral feeding, and her weight is 11.5 kg. She has not required antihypertensive medications for the last 6 months. Recent CT angiography revealed increased narrowing of both renal arteries, the superior mesenteric artery, the celiac artery, and the hepatic artery, and complete obliteration of the left external iliac and left femoral arteries. However, a good set of collateral vessels were seen during the evaluation (Figure 4).

**Discussion:**

Renovascular hypertension in infants is caused by a large group of vascular disorders. These disorders include renal venous thrombosis, thromboembolism of the renal artery, external compression of the renal artery, fibromuscular dysplasia, neurofibromatosis, Takayasu's arteritis, Kawasaki disease, William's syndrome, midaortic syndrome, idiopathic arterial calcification, and so called unspecified group of vascular occlusive diseases [1,5]. Management of hypertension is individualized and depends primarily on the causative disorder [2-4]. However, regardless of the causative disease, control of BP is a priority to prevent the possible complications of hypertension. Reproducible BP measurements above 90/60 mmHg are widely accepted as the definition of hypertension in the term neonate [8]. Although many sick neonates are treated for hypotension and hypertension, the normal physiological BP to ensure appropriate organ perfusion is
uncertain [3]. Intestinal angina in patients with multiple arterial stenoses is rare. Stanley et al [2] reported only two cases with classic intestinal angina out of 24 cases with splanchnic arterial lesions. Christine et al [7] reported only one case of bowel ischemia out of 102 cases of idiopathic mid-aortic syndrome. Our patient fell in the group of unspecified vascular occlusive disease. Medical control of the BP with a PTA, or vascular reconstructive surgery at a later date seemed to be an appropriate treatment strategy. The BP was brought to "near normal" levels. However, in the presence of superior mesenteric artery stenosis, these "near normal" levels were insufficient to ensure adequate bowel perfusion, and the patient developed bowel ischemia.

When the patient passed the critical period of sepsis and short bowel syndrome and reached a weight of 5 kg, she underwent two attempts to perform PTA. These attempts were unsuccessful because of the very small diameter of the femoral arteries. Open vascular surgery was unfeasible because of lack of experience. The only choice was to wait and see. Although radiological follow up showed increasing narrowing in the stenotic areas, the patient showed clinical improvement over time. This could be explained by the development of collateral circulation seen on follow up doppler ultrasound and CT angiography. Christine et al [7] reported oliguric renal failure in only 4% of 102 cases of idiopathic mid-aortic syndrome and stated that effective collateral circulation develops over time. Srinivasan et al. [10] documented the presence of collateral circulation in 51% of the 68 angiograms performed for 43 children with renovascular hypertension due to fibromuscular dysplasia and neurofibromatosis type 1.

Conclusions:
Management of hypertension in infants caused by unspecified vascular occlusive disease is challenging. A multidisciplinary approach is important. Although intestinal angina is a rare complication, doctors and parents should be aware of it. Parent’s education is essential to prevent late presentation. Keeping BP at "high permissible" levels may prevent bowel hypoperfusion. Aggressive angioplastic interventions and open reconstructive surgeries are not indicated when the BP is medically controlled and the organs have normal functions. Careful medical treatment and waiting for collateral circulation to develop may be appropriate in such difficult clinical situations.

Consent

Written informed consent was obtained from the parents of our patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

Authors’ contributions

HS interpreted the patient’s radiologic data. MZ, MN, and SJ were major contributors in writing the manuscript. All authors read and approved the final manuscript.

References:


Figure legends:

**Figure 1:** Doppler ultrasound of renal arteries: severe bilateral renal artery stenosis with a peak systolic velocity (PS) of 250 cm/sec and a resistive index (RI) of 0.89

**Figure 2:** CT angiogram: stenosis of both renal arteries near the ostium (arrows).

**Figure 3:** CT angiogram: stenosis of the superior mesenteric artery (arrow).

**Figure 4:** Follow up CT angiogram: complete obliteration of the left external iliac and femoral arteries (arrow), and development of collateral circulation (dotted arrows).