INTESTINAL ISCHEMIA IN NEONATES AND CHILDREN

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Abstract

The article reviews the intestinal ischemia theme on newborn and children. The intestinal ischemia may be either acute - intestinal infarction (by vascular obstruction or by reduced mesenteric blood flow besides the occlusive mechanism), either chronic. In neonates, acute intestinal ischemia may be caused by aortic thrombosis, volvulus or hypoplastic left heart syndrome. In children, acute intestinal ischemia may be caused by fibromuscular dysplasia, volvulus, abdominal compartment syndrome, Burkitt lymphoma, dermatomyositis (by vascular obstruction) or familial dysautonomia, Addison's disease, situs inversus abdominus (intraoperative), burns, chemotherapy administration (by nonocclusive mesenteric ischemia). Chronic intestinal ischemia is a rare condition in pediatrics and can be seen in abdominal aortic coarctation or hypoplasia, idiopathic infantile arterial calcinosis.

Keywords: intestinal ischemia, newborn, children

Introduction

Acute mesenteric ischemia consists of the rapid, partial or complete, interruption of the blood flow in the irrigation area of the superior or inferior mesenteric artery, that results in intestinal infarction - the hemorrhagic necrosis of the intestines [1].

Intestinal ischemia is a rare condition in neonatology and paediatrics. In its etiology we distinguish the vascular occlusion and the nonocclusive mesenteric ischemia (NOMI), induced by intestinal vasoconstriction without thromboembolic occlusion [2] or the decreasing mesenteric blood flow.

In 1960, Ratner and Swenson reviewed one of the first series of acute mesenteric infarctions in infants and children [3].

Acute intestinal ischemia in the newborn

By vascular obstruction. Arterial thromboses are rare and severe in newborns, mostly caused by arterial catheterization [4-5]. There was reported a case of aortic thrombosis with mesenteric failure, not caused by catheterization, with symptoms of bilious vomiting, melena. Physical examination showed dehydration, meteorism. Diagnosis was confirmed by Doppler sonography. Shortly after, the newborn deceased by multiple organ dysfunction syndrome [6]. In such cases one can apply thrombolytic therapy [7], and if the treatment is contraindicated or fails, surgery is recommended [8].

Intestinal malrotation can cause intestinal infarction by volvulus (Figure 1).

By NOMI. A study on 387 patients with hypoplastic left heart syndrome showed that 6.5% of patients were diagnosed with mesenteric ischemia. In 80% of these patients, a low perfusion state and significant hypotension...
were documented within 48 hours prior to the diagnosis of bowel ischemia. 84% died shortly after onset of the gastrointestinal symptoms regardless of means of management. Others died because of complications of their primary cardiac disease. Therefore, the overall mortality was 97%. Mesenteric ischemia has been erroneously identified as *necrotizing enterocolitis* (NEC) (Figure 2 A, B). These two disorders may share some common pathological and clinical features but are distinctly different disease processes [9].

NEC remains one of the most devastating and poorly understood intestinal diseases in neonates. Ischemic injury of the bowel is a risk factor, together with genetics, microbial dysbiosis and immaturity (vasculature, motility, barrier, innate immunity). The reality is that ischemia is most likely involved in some intermediary role [10].

![Figure 1. Newborn. Intestinal infarction through volvulus.](collection of A. Budușan, Pediatric Surgery Clinic, Emergency Hospital for Children Cluj-Napoca)

![Figure 2 A, B. Newborn. *Necrotizing enterocolitis*.](collection of V. Negrea, Pediatric Surgery Clinic, Emergency Hospital for Children Cluj-Napoca)
Acute intestinal ischemia in children
By vascular obstruction. Fibromuscular dysplasia is a rare disorder of medium-sized arteries with no signs of inflammation or atherosclerosis. It is the most common cause of stenosis in children and young adults [11].

Midgut volvulus is the most common serious complication of malrotation, and it may cause intestinal ischemia [12-13].

Another cause of intestinal ischemia in children is abdominal compartment syndrome (ACS) (in contrast with adults, children with ACS have diverse primary diagnoses, with a significant number of primary extra-abdominal-mainly central nervous system-conditions; ischemia and reperfusion injury appear to be the major mechanisms for development of ACS in children) [14].

A case of a 14-year-old boy was reported who presented with septic shock and was found to have necrosis of the cecum because of a Burkitt lymphoma. The cause of the ischemia appeared to be mesenteric infiltration and subsequent vascular compromise of the bowel [15].

There was also a case of a 4-year-old boy with dermatomyositis and abdominal pain. An emergency operation showed an ulcer and perforation at the junction of the third and fourth portions of the duodenum because of the mesenteric vasculitis [16]. Rheumatologists should be alerted about this serious complication in patients with childhood or young adult dermatomyositis presenting with abdominal complaints [17].

Figure 3. 5-year-old girl. Small intestine rotated around Ladd’s bands; cyanotic bowel downstream of bands.
(collection of N. Bratu, Pediatric Surgery Clinic, Children Clinical Emergency Hospital “Marie Curie” Bucharest)

Figure 4. 16-year-old girl. Intestinal infarction through volvulus on postoperative peritoneal adhesion.
(collection of A. Budușan, Pediatric Surgery Clinic, Emergency Hospital for Children Cluj-Napoca)
Familial dysautonomia (Riley–Day syndrome) is an autosomal recessive hereditary neuropathy characterized by impaired development and progressive degeneration of the sensory and autonomic nerves. Patients present various symptoms, including cardiovascular instability, recurrent pneumonia, gastrointestinal dysfunction, decreased sensitivity to pain and temperature. Familial dysautonomia is a very common disorder in the Ashkenazi Jewish [18]. There is a case report of a 14-year-old boy with familial dysautonomia in whom a small-bowel infarction developed during a dysautonomic crisis; the patient presented hypotension, prolonged fever and abdominal distension. Probably, the bowel infarction was caused by prolonged hypoperfusion. Abnormal systemic cardiovascular regulation in patients with familial dysautonomia may affect splanchnic blood flow, which led to the fatal consequences in this case [19].

An 11-year-old boy with a history of abdominal pain, diarrhoea, emesis and fever, presented with dehydration, shock and acute mesenteric ischaemia. Final diagnosis was Addison’s disease [20].

A 5-year-old girl, diagnosed with situs inversus abdominus and polysplenia syndrome, developed during the surgical correction of malrotation (Ladd’s procedure performed in reverse fashion) transverse colon ischaemia by spasm of superior mesenteric artery, probably caused by bowel evisceration, intraoperative hypotension, and situs inversus abdominis [21].

Splanchnic ischaemia can be an important problem in pediatric patients with burns. A case report presented of an 11-year-old boy with 70% full-thickness burns who sustained multiple episodes of severe gastrointestinal haemorrhage due to both extensive ischaemic enterocolitis and severe gastric ulceration which required surgical intervention on several occasions. Causative mechanisms of splanchnic ischaemia in this patient including increased mesenteric vascular resistance, abdominal compartment syndrome and enteric feeding, are considered [22]. Another case presented extensive ischemic necrosis of the large and small bowel in a 2 year-old child, who sustained 20% total burn surface area partial-thickness flame burn [23].

There was reported a case of a 18-year-old girl with NOMI (necrosis from the ileum to the ascending colon) on the 16th day after the start of chemotherapy administration (prednisolone, L-asparaginase, vincristine and bortezomib) for a third hematological relapse of acute lymphoblastic leukemia [24].

### Chronic intestinal ischemia in pediatric patients

Chronic mesenteric ischemia is characterized by inadequate blood flow to the bowel resulting from stenosis on mesenteric arteries [25]. Untreated, bowel ischemia may cause malnutrition or acute bowel ischemia with infarction [26]. Chronic mesenteric ischemia is particularly rare in childhood. Because of the nonspecific nature of symptoms produced and absence of pathognomonic findings by physical examination or by routine laboratory testing, its recognition is difficult and its true incidence is unknown. The finding of both renal and visceral artery disease in three of the four patients underscores the need for adequate evaluation of mesenteric vessels before renovascular procedures are undertaken in this age group [27].

The incidence of chronic mesenteric ischemia associated with abdominal aortic coarctation or hypoplasia has been reported as 22% [28]. A case of 13-year-old female was reported with partially duplicated aorta with the anterior aorta containing the splanchnic and renal arteries and the posterior segment perfusing the lower extremities (treated with balloon angioplasty) [29].

For another case, a 12-year-old girl with idiopathic infantile arterial calcinosis, angioplasty has been used successfully to treat chronic mesenteric ischemia [30].

| Table I. Reported causes of intestinal ischemia in newborn and children. |
|-----------------------------------------------|
| **NOMI** | **By vascular obstruction** | **By NOMI** |
| **Neonatesgy** | Acute | Severe infection [6]  
Perinatal asphyxia [6]  
Severe dehydration [6]  
Malrotation [12-13] |
| | | Hypoplastic left heart syndrome [9] |
| **Pediatric patients** | Acute | Fibromuscular dysplasia [11]  
Malrotation [12-13]  
Abdominal compartment syndrome [14]  
Burkitt lymphoma [15]  
Dermatomyositis [16] |
| | | Familial dysautonomia [19]  
Addison’s disease [20]  
Situs inversus abdominus (intraoperative) [21]  
Burns [22,23]  
Chemotherapy administration [24] |
| | Chronic | Abdominal aortic coarctation or hypoplasia [28,29]  
Idiopathic infantile arterial calcinosis [30] |
Conclusions

The information existing in the field literature is poor. It is necessary to pay attention to this pathology, especially when it comes about this age group. The paraclinical examinations, especially those with vascular imaging, bring an important contribution to the correct diagnosis. Overall, the surgical treatment has the same principles as for adult intestinal ischemia.

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