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Intestinal intussusceptions in children population

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Abstract:

Intussusception is an invagination of a proximal segment of the bowel into an immediately adjacent, distal portion of the bowel. Ileocolic intussusception is one of the most common

causes of bowel obstruction in children. The peak incidence occurs in infants between five and seven months of age. The etiology of intussusception in children is typically idiopathic, approximately 5-10% of patients have a pathological lead point. Postoperative intussusception is one of the rare causes of intussusception. Children with ileocolic intussusception often present abdominal pain, palpable mass, rectal bleeding, fussiness, and emesis. The most effective imaging method for the diagnosis of intussusception is ultrasound. The treatment of ileocolic and ileoileal intussusceptions is different, the first type requires invasive treatment.

Keywords:

intussusception, bowel obstruction, postoperative intussusception, ileocolic intussusception, ileoileal intussusception

Introduction:

Intussusception is the most common cause of gastrointestinal obstruction in infants and young children. An intussusception is defined as the telescopic invasion of the intestine into its lumen followed by the displacement of the invaginating fragment distally in accordance with peristalsis [1]. Intussusception can occur in any part of the bowel, but the most typical localisation, which is of clinical relevance, is the ileocaecal region, where ileocolic intussusceptions are found [2]. The etiology of intussusception in children is typically idiopathic, often influenced by anatomical or infectious factors [1], only 5-10% of cases have an identified lesion that caused intussusception [3,4,5]. Ultrasound is the initial and crucial diagnostic method, also used during nonoperative procedures such as enema to confirm intussusception reduction [3]. Differential diagnosis of intussusception includes gastritis, acute appendicitis, hernia, testicular torsion, intestinal torsion [6,7,8] and other surgical conditions leading to intestinal obstruction (malrotation, intestinal atresias, meconium ileus, etc.) [9]. Early diagnosis of intussusception is important, as the entrapment of the mesenterium into the cavity results in venous congestion and intestinal circulation disturbances with swelling of the intestinal wall, resulting in gastrointestinal obstruction, intestinal wall necrosis and perforation with peritonitis and septic shock. [1].

Aim

The aim of the study is to summarize the current knowledge about intestinal intussusception in the pediatric population, which may be of clinical significance. Accumulated knowledge will avoid misdiagnosis with clinically significant intestinal abnormalities.

Method

A review of the scientific literature and publications of the PubMed database. Using medical records, we describe the essence of selected types of intestinal intussusception in children and their possible complications.

Results

1. Etiology

We distinguish some of the anatomical features of the gastrointestinal tract that are predisposed to intestinal intussusception in the paediatric population: lack of mature participation of the large intestine, reduced intestinal stiffness due to missing or underdeveloped teniae coli and anterior insertion of the rectum against cecum and lack of mature participation of the longitudinal muscle fibers of the colon at the level of the ileocecal valve [3].

Another common cause of pediatric intussusception is infectious etiology resulting in mesenteric lymphadenopathy [3]. A viral infection with accompanying hyperplasia of Peyer's patches and hypermobility has also been suggested as pathogenesis [10,11]. Approximately 30% of cases before intussusception had a viral disease [3].

Most of the clinically relevant pathological lead points are lymphoma and Meckel's diverticulum and others like polyps, duplications and bowel wall tumours such as lipomas, hamartomas, schwannomas, lymphangiomas and haemangiomas [2]. Intussusception can also be caused by noninfectious etiologies, such as intestinal allergies, Celiac disease, and Crohn disease similarly to infectious causes with hypertrophy of Peyer patches and/or mesenteric lymphadenopathy as a lead point [3].

2. Epidemiology

The age range in which intussusception typically occurs is not precisely defined, varying between 5 and 36 months of age [1]. The incidence of intussusception decreases with age, with only 30% of all cases occurring in children older than 2 years of age [3]. The worldwide incidence has been estimated to be 0.74 (range 0.09–3.28) per 1000 children, with a peak incidence among infants of five to seven months of age [2]. However, intussusception can occur at any age, also in utero (impaired blood supply due to intussusception is one of the postulated mechanisms of the development of congenital intestinal obstruction). Boys suffer from intussusception more often (about 2/3 cases) [1]. Intussusception is rare in neonates, especially premature neonates, its incidence is only 0.3% (0%–2.7%) of all cases of intussusceptions [9]. Intussusception in full term neonates and infants is usually ileo-colic (80%), in premature neonates mostly in small bowel (91.6%) – ileal or jejunal [9,12,13].

3. Clinical Presentation

In the pediatric population, intussusception is one of the most common abdominal emergencies and is readily diagnosed based on a typical pattern of abdominal pain, palpable mass and “currant jelly” bloody stools in children under 2 years old [3,5,14]. The classic pediatric triad is quite rare and occurs in less than 15% of cases [3]. Children with intussusception often present with nonspecific complaints, such as abdominal pain, fussiness, and emesis. These symptoms can make the diagnosis challenging [10]. The development of symptoms depends on the degree of strangulation and obstruction of the bowel [5]. Children under 2 years of age usually show symptoms such as screaming or irritation, and it is also characteristic for the child to lie on his or her knees in the chest and to have acute abdominal colic. Between bouts, the child may be sluggish when the pain is stronger, but it may also return to activity when the pain is less [3].

4. Diagnosis

Physical examination reveals a palpable mass, but it is present only in about 60% of cases, usually in the upper right quadrant of the abdominal cavity or in the upper abdomen and usually shaped like a sausage [3]. The diagnostic modality of choice is ultrasonography, being a quick, simple and non-invasive way to diagnose intussusception, with a very high sensitivity (98–100%) and specificity (88–100%) [15].

A characteristic feature may be the externally visible ring around the intussusceptum caused by a swelling of the intussusception forming the target or the doughnut sign [3,16]. On the transverse view we can notice pseudo-kidney appearance which is formed by the layers of the intussusception [3]. Color Doppler may show decreased blood flow to the intussusceptum if bowel is ischemic. Ultrasound, however, has its limits when the presence of massive air in cases of bowel distension or morbid obesity, both of which reduces the chance to make a correct diagnosis of intussusception [3,17].

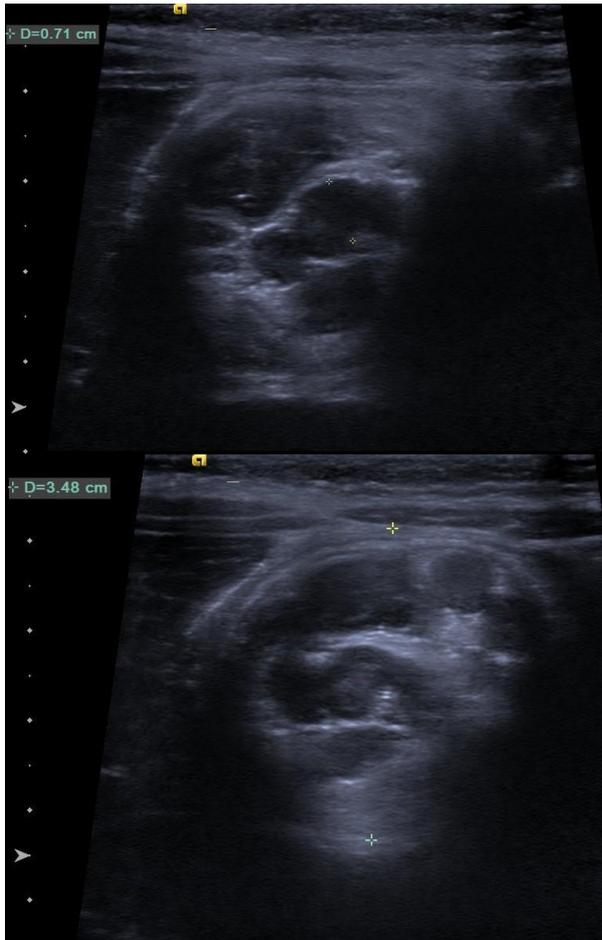
Imaging methods are necessary for diagnosis. Many studies have shown that conventional abdominal X-ray has a range of sensitivity and specificity much lower than that of ultrasound, which is very sensitive and specific with a high negative predictive value (99.0% -99.7%). [10, 18, 19, 20]. Plain abdominal X-rays and computerised tomography are no longer commonly used to diagnose intussusception, and the exceptional use of contrast studies has been described [2]. Ultrasound is the first-line imaging modality for the evaluation of intussusception, because of no ionizing radiation [10,21]. Ultrasound is also an effective instrument used in the infusion treatment of the intussusception. Reducing intussusception under ultrasound control with a saline intake is an effective and safe method that prevents the child from being exposed to a significant amount of radiation [22].

5. Ileocolic Intussusception

Ileocolic intussusception is an invagination of the terminal ileum through the ileocecal valve into the colon. Most cases in infants and young children up to the age of 3 years are idiopathic, usually with hypertrophied lymphoid tissue in the terminal ileum acting as a lead point. Some intussusceptions, especially in older children, are with a pathological lead point [23,24].

Ileocolic intussusception is a common cause of acute abdominal pain in children and the most common cause of obstruction in young children [23,25]. The invagination of the terminal ileum into the colon causes constipation, compression of the blood vessels and resulting ischemia. Typical clinical symptoms are abdominal pain, vomiting and bloody stools. Less than 25% of cases have a classic triad of symptoms. [23,26,27]. If intussusception is long-lasting, perforation occurs, diffuse peritonitis and shock evolve, and death is possible [23,24]. Authors of a study from 2018 from Uganda reported a mortality rate of 32% for patients with this condition [23,28]. Early diagnosis and appropriate treatment are critical to prevent this outcome. Radiology is important for a correct and rapid diagnosis of this disease, as the clinical symptoms are often unclear and the laboratory values are unspecified (Fig

1.)[23]. Though in most cases idiopathic, possibility of a patologic mass as the lead point of intussusception should always be considered [29,30]. The ileocolic intussusception in most cases requires invasive treatment: enema or in some cases surgery [15]. Ultrasound-guided or fluoroscopic pneumatic or hydrostatic enema is successful in 85 to 90% of cases [3,31]. If the non-operative reduction is ineffective or there are signs of peritonitis the surgical intervention may be required.



(Fig. 1.) Ileocolic intussusception with enlarged lymph nodes in a 3-year-old boy.

6. Small Bowel Intussusception (SBI)

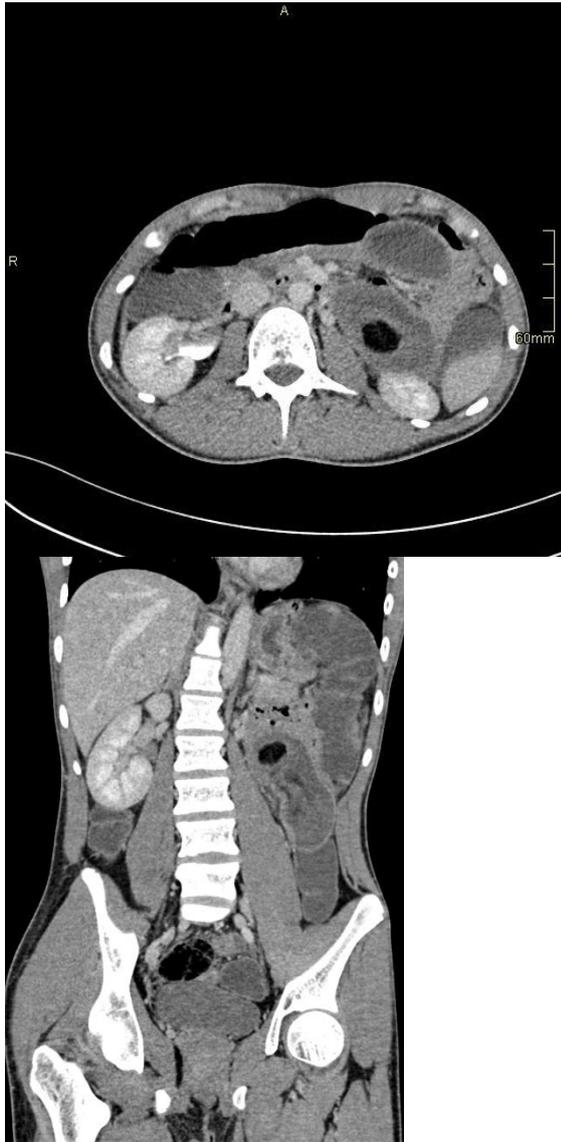
Small bowel intussusception (SBI) includes jejuno-jejunal, ileo-ileal, jejunoileal and duodenojejunal intussusceptions [32,33]. SBIs occur mainly as ileoileal intussusceptions, which are located in the paraumbilical region [15]. Pediatric SBI occurs less frequently than ileocolic intussusception. SBI represents 1–10% of all intussusceptions, but the incidence increases with the age of children [33]. Ileoileal intussusception is more common in neonates

and older children (>5 years old) [15]. In children population, SBIs usually have idiopathic cause, but the percentage of intussusceptions due to a pathological lead point increases with age [3,15]. The basic method of SBI diagnosis is ultrasonography (Fig.2.). The use of CT allows for a certain diagnosis and enables the diagnosis of complications, such as mesenteric thrombosis or small intestine torsion (Fig.3.)[33]. Most cases of SBIs are transient and could be observed dynamically until spontaneous reduction [32]. SBI tend to decrease spontaneously in children with gastroenteritis and/or upper respiratory tract infection, without clinical suspicion of intussusception [34]. Typically, SBI occurs in the subacute presentation, therefore it is difficult to diagnose preoperatively, which increases the risk of ischemic complications [33].

The small bowel is the most common site of intussusception in adults [35]. Adult SBIs have a pathological lead point (such as Meckel diverticulum and adhesions) in up to 90% of cases [3, 36]. About 60% of SBI in adults are caused by benign lesions, the remaining ones arise on the basis of a malignant lesion or are classified as idiopathic intussusception. [35]. In literature, about 50% of malignant lesions causing SBI were metastatic melanomas [3]. Primary melanoma of the small bowel is a rare, but also could be pathological lead point to intussusception [36].



(Fig.2.) Ultrasonography - Small bowel intussusception in a 2-year-old girl.



(Fig.3.) CT- small bowel intussusception with a fat-containing tumor as a lead point in a 16-year-old boy.

7. Intussusception with a pathological lead point

Pediatric intussusceptions have a pathological lead point in about 5-10% of cases [3,37]. Pathological lead points include Meckel's diverticulum, lymphoma, polyps, duplication cysts, and intestinal wall tumors (lipoma, hamartoma, lymphangioma, adenomyoma), appendicitis and digestive malformations [2,10,38]. Intussusception is also associated with certain general diseases, such as rheumatoid purpura, cystic fibrosis, coagulation disorders, hemophilia, and celiac disease [38].

Intussusception could arise secondary, the most common due to the presence of a Meckel's diverticulum [2,38]. Meckel's diverticulum is the most common gastrointestinal malformation with the incidence in 2% of the population [39]. Meckel's diverticulum usually causes gastrointestinal tract bleeding, bowel obstruction from volvulus or intussusception [40]. About 4% of patients with Meckel's diverticulum experience symptoms [39]. Meckel's diverticulum presents with gastrointestinal bleeding, cramps, tenderness around the navel, intestinal obstruction causing pain, gas, diarrhea, constipation, vomiting and diverticulitis [40]. Management of intussuscepted Meckel's diverticulum require resection with subsequent anastomosis, usually using a laparotomy or with laparoscopic assistance [2].

Neoplasia is a rare etiology of intussusception in children. The most common neoplastic cause of intussusception in the pediatric population is lymphoma [3]. The cases report in the literature usually concerns Burkitt's lymphoma or large B-cell lymphoma [5]. Typical symptoms of gastrointestinal tract lymphoma are abdominal pain, nausea, vomiting, diarrhea, and malabsorption [41]. Ultrasound is usually used in the diagnosis of lymphoma of the gastrointestinal tract, MRI or CT is intermittently helpful in diagnosing [2]. Children with Burkitt's lymphoma with symptoms of intussusception often have completely resectable disease [5]. Surgical intervention is the method of choice in treatment of Burkitt's lymphoma complicated by an intussusception, with performing complete resection of tumor with subsequent anastomosis [2].

8. Post-operative Intussusception (POI)

Post-operative intussusceptions (POI) are rare entities [42]. POI is a forgotten cause of post-operative obstruction and usually is misdiagnosed as post-operative adhesive obstruction [43]. POI is potentially fatal complication which can occur after virtually any type of abdominal surgery, such as retroperitoneal tumor resection, Ladd's procedure, diaphragmatic operation, and pancreatectomy and so on [44, 45]. The incidence of POI is 0.01–0.25 % of children after laparotomies [45]. The POI concerns in the literature in most cases small bowel intussusceptions with ileoileal intussusception predominance; a few ileocolic intussusception; Emil S et.al. was the first to describe post-operative colocolic intussusception [42,45]. During the first two weeks postoperatively in 90% of patients with POI occur bilious vomiting, high nasogastric tube output, abdominal pain or abdominal distension [43]. Early recognition of POIs may be hampered by the absence of the typical triad of intussusception (abdominal colicky pain, abdominal mass and bloody stool) and presence of postoperative ileus [45]. Diagnosis of POI is challenging and needs a very high index of suspicion. In the diagnosis of

POI abdominal radiograph, abdominal ultrasonography, contrast study and computerized tomography (CT) scan are used [43]. Abdominal ultrasound might have a lower accuracy in diagnosing postoperative intussusception than diagnosing idiopathic intussusception [46]. CT is an imaging diagnostic method used to exclude or confirm POI in suspected patients [45]. Treatment of POI require immediate surgical intervention [46]. Operative management usually involves manual reduction with resection and anastomosis in select cases [43].

9. Treatment

Treatment of intussusception depends on the type and area of occurrence [3]. The treatment of ileocolic and small bowel intussusceptions is different. In patients with ileoileal intussusception without any indication for surgery, ‘wait-and-see’ policy is preferred, in contrast the ileocolic intussusceptions require surgical intervention [15].

Early diagnosis of intussusception and prompt management reduces associated risks and the need for surgical intervention [47]. The gold standard treatment of the pediatric intussusception is hydrostatic or pneumatic reduction. Its effectiveness is estimated from 40% to 90%, mainly depending on the duration of symptoms and radiological experience [48]. Close observation is required due to the heightened possibility of recurrence within the first 24 hours [3].

Invasive management is generally indicated if peritonitis, bowel perforation, or shock occurs or when appropriate radiological facilities are unavailable or when contrast enema reduction is unsuccessful [47]. Primary surgery may be indicated despite a higher risk of complications and intestinal resection in cases of failure in enema reduction or if a successful result is not expected due to clinical conditions or long-lasting symptoms [48]. Open laparotomy with manual reduction of intussusception is used in the surgical management, although laparoscopy may be safer and just as effective and may result in shorter hospitalization [47]. Performing a reduction is inadvisable when preoperative colonoscopy, imaging studies or intraoperative appearance strongly suggests the presence of malignancy or ischemic bowel changes are suspected with risk of perforation with even gentle manipulation. Then the entire segment should be resected en bloc [3].

Conclusion

Intussusception is one of the most common causes of intestinal obstruction and acute abdomen in children. Early diagnosis of intussusception in children is difficult due to non-

specific symptoms. Particular attention should be paid to symptoms such as abdominal pain, vomiting, palpable mass and bloody stools. The imaging examination necessary for the diagnosis of intussusception is ultrasound, characterized by high sensitivity (98–100%) and specificity (88–100%). It is important to distinguish ileocolic intussusception from ileoileal intussusception during an ultrasound, because they differ in treatment method. The etiology of intussusception is most often idiopathic, but they may arise from previous pathological lesion or appear postoperatively. The treatment involves ultrasound-guided/fluoroscopic/pneumatic or hydrostatic enema or surgery management depending on the type of intussusception and the stage of the disease.

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