Neonatal Intussusception: A Review

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ABSTRACT

Intussusception is defined as a process in which a segment of bowel invaginates into the adjoining intestinal lumen, causing bowel obstruction. It is the most common cause of intestinal obstruction in infants and children between 6 months to 18 months of age. In neonates and premature infants, it accounts for only 3% of intestinal obstruction and 0.3% (0–2.7%) of all cases of intussusception. Since neonatal intussusception is an uncommon entity, we conducted this review to examine its occurrence, clinical features, diagnostic modalities, and treatment options.

Key words: Intussusception; Neonatal intussusception; Neonatal intestinal obstruction; Neonatal acute abdomen

INTRODUCTION

Intussusception is defined as a process in which a segment of bowel invaginates into the adjoining intestinal lumen, causing bowel obstruction. With early diagnosis, appropriate fluid resuscitation, and therapy, the mortality rate from intussusception in children is less than 1%.

It is the most common cause of intestinal obstruction in infants and children between 6 months to 18 months of age. In neonates and premature infants, it accounts for only 3% of intestinal obstruction and 0.3% (0–2.7%) of all cases of intussusception [1-5]. Although small bowel intussusceptions are very rare in infants, it is common in neonates and premature infants [1, 2].

Intussusceptions occur at the rate of 1-4 in 2000 infants and children [6,7]. Although seen in all pediatric age groups and even in adults, it is most commonly seen in first two years of life (75%). Ninety percent cases occur before three years of age. More than 40% present between 3-9 months of age. Male to female ratio is 2:1 to 3:2 [6, 8]. Since neonatal intussusception is an uncommon entity, we conducted this review to examine its occurrence, clinical features, pathophysiology, and management.

METHODS

A search of the Pub-Med database was conducted on intussusception in neonatal age group published between 1975 and 2017. We included only English language literature. Search terms included intussusception, neonatal intussusception, neonatal intestinal obstruction, and neonatal acute abdomen. Any publication related to the subject intussusception in patients of neonatal age group was included for review regardless of study type, as well as abstracts of relevant articles. The papers were analyzed for their relevance to the topic. All papers related to neonatal intussusception were included in the study. Full texts of the articles, if possible, were accessed to assess their relevance to the subject.

RESULTS

More than 50 papers published on intussusceptions were found. Of these, 22 had mentioned neonatal intussusceptions (table 1). Majority of the ar-
icles on neonatal intussusceptions were individual case reports (19), and remaining were case series with review of literature (4). The publications covered a time period of 42 years, and still, covered only around 70-80 cases during this long-time span, demonstrating the rarity of this clinical entity.

After reviewing all the articles, it was observed that neonatal intussusception is difficult to diagnose as many cases resemble a clinical picture of necrotizing enterocolitis (NEC) [7, 9-11]. Most of the time, the diagnosis is made on the operating table when the baby is operated for complications like intestinal perforation or obstruction [7, 9, 12-13].

**Etiology.** The etiology of neonatal intussusception is not very clear. This is especially true for premature neonates [3]; however, in full term neonates, a lead point is present in approximately 58% of patients [2], such as duplication cyst, hamartoma, Meckel's diverticulum, or mesenchymoma [8, 14]. In premature infants, it is suggested that common perinatal risk factors resulting in intestinal hypoperfusion/hypoxia, dysmotility, and stricture formation may act as a lead point for intussusception [3]. In premature neonates, lead point has been found to be about 8% [15].-inspired meconium has also been found to be causing intussusception [14]. It has been observed that hypoxic events may play a crucial etiologic role in the pathogenesis of late onset neonatal intussusception [16].

**Clinical features.** Intestinal obstruction in neonatal period may be due to duodenal or small intestinal atresia or stenosis, malrotation, meconium ileus, meconium plug syndrome, anorectal malformations, Hirschsprung's disease, ileus related to sepsis, and other rare causes [17, 18].

Intussusception is an extremely rare cause of intestinal obstruction in neonatal age group and coupled with its presentation similar to NEC, the diagnosis of Neonatal intussusception is easily missed [11, 19-21]. Since the initial treatment protocol for intussusception (Surgical intervention) and that of NEC (Conservative management) are diametrically opposite [11, 19-21], it becomes important to accurately diagnose and differentiate these conditions. Though surgical intervention may be needed in patients of NEC, it is mostly in stage III of Bell's classification [22]. Unless the initial presentation of NEC is pneumoperitoneum, the initial management is conservative. The delay in making a diagnosis of intussusception in neonates is considerable with a mean of 10-19 days [15, 19]. This delay in diagnosis leads to compromised bowel and further complications [24].

The patient with intussusception is usually an infant, often one who has upper respiratory infection. The presenting symptoms include vomiting, abdominal pain, passage of blood and mucous in stool, and lethargy. In infants, the hallmark physical findings in intussusception include sausage-shaped mass in right hypochondrium and emptiness in the right lower quadrant (Signe de dance). If obstruction is complete, abdominal distention is frequently present. Contrary to this, neonatal intussusception presents with features mimicking NEC like abdominal distension, feeding intolerance, vomiting, and bloody stools [1, 15, 23, 25]. In full term neonates, ileocolic intussusception is the most common type [3]. However, in premature neonates, small bowel intussusception, primarily ileal, is more common than ileo-colic type [15, 19].

**Radiological features.** The radiological imaging in neonatal intussusception is generally unremarkable. On plain X-ray abdomen, features of obstruction like multiple air-fluid levels or dilatation of bowel loops may be seen [15, 24, 26].

Contrast enema may be useful in diagnosing intussusception in term neonates, as they are more likely to have a colonic component, it is of not much help in pre-term neonates [1, 3, 15, 23, 27-29]. There is a greater risk of bowel perforation while performing contrast enema as the bowel is frequently compromised till the time of investigation [24].

Ultra-sonography (USG) is an important investigation for diagnosing Neonatal intussusception. It has numerous advantages like absence of harmful radiation exposure to the neonate, lead point identification and ability to diagnose ileo-ileal and other small bowel intussusceptions [1, 15, 16, 19, 24, 30]. Sometimes, in neonates, the sigmoid colon lies in the right side along with distended small bowel loops, overlapping the intussusception [31]. This may obscure the diagnosis of intussusception on USG. This point needs to be kept in mind while dealing with such patients.

**Differential Diagnosis.** Neonatal Intussusception usually presents with features of abdominal distention, bilious vomiting and failure to thrive. These symptoms often mimic other differentials like NEC, intestinal atresia, intestinal malrotation or band obstruction [1-3, 6, 11, 13, 21].
Neonatal Intussusception: A Review

Management - Early and accurate diagnosis of neonatal intussusception is of essence in the management [1]. On the basis of previous reports, some attempts may be taken to differentiate NEC from neonatal intussusception [1]. NEC usually begins during the first 5 days of life. Thereafter, it may either evolve rapidly to bowel gangrene and perforation, which may lead to death; or it may follow a protracted course to intestinal perforation or gradual resolution of symptoms. Clinically, the infant’s general condition usually parallels the abdominal condition. In case of the neonatal intussusception, the onset of symptoms usually occurs after the 1st week of life. The persistent gastro-intestinal symptoms do not lead to deterioration of the general condition until there is bowel perforation. Hence, the possibility of intussusception should be highly suspected in a baby who was diagnosed with NEC, but who the patients has a more stable course than would normally be expected. In such a scenario, early surgical intervention is warranted to avoid further damage to viable bowel [1, 3, 14].

Ultrasonography of the abdomen has a high sensitivity in identifying the same and is a valuable tool while making a diagnosis [1, 15, 19].

Most of the cases of neonatal intussusception are initially treated as NEC, hence, very few articles in current literature find mention of USG as a diagnostic modality. As in any neonate with acute abdomen, neonatal intussusception is initially managed with intravenous fluids and antibiotics, and other symptomatic treatment. The possibility of bowel perforation reaches up to 50-60% in case of delayed presentation of neonatal intussusception [15, 19]. Prematurity is an added risk factor for mortality [14]. Owing to the misdiagnosis of NEC, majority of these patients develop surgical complications like intestinal obstruction and intestinal perforation. This leads to surgical intervention, where a diagnosis of intussusception is made. Consequently, exploratory laparotomy and manual resection, whichever applicable, remains the treatment of choice. Besides, since the diagnosis

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Patients</th>
<th>Result</th>
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</thead>
<tbody>
<tr>
<td>Tepmalai et al. [7]</td>
<td>2017</td>
<td>1</td>
<td>Neonatal intussusception in premature baby presenting with perforation</td>
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<tr>
<td>Mannai et al. [8]</td>
<td>2017</td>
<td>1</td>
<td>Acute intussusception in new born: A rare cause of intestinal obstruction</td>
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<td>Saldanha, et al. [12]</td>
<td>2016</td>
<td>1</td>
<td>Ileo-colic intussusception in premature neonate</td>
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<tr>
<td>Joshi et al.[9]</td>
<td>2015</td>
<td>1</td>
<td>Jejunal atresia due to intra-uterine intussusception</td>
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<tr>
<td>Kumar Das et al.[12]</td>
<td>2015</td>
<td>1</td>
<td>Meckel’s diverticulum causing neonatal intussusception</td>
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<tr>
<td>Prakash et al. [11]</td>
<td>2015</td>
<td>1</td>
<td>Intussusception in premature neonate -often misdiagnosed</td>
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<tr>
<td>Altuntas et al. [11]</td>
<td>2015</td>
<td>1</td>
<td>Ileo-ileal intussusception in premature neonate presenting as NEC</td>
</tr>
<tr>
<td>Taskinlar et al. [21]</td>
<td>2014</td>
<td>3</td>
<td>Diagnostic challenges between neonatal intussusception and NEC</td>
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<td>Ohuoba et al. [30]</td>
<td>2013</td>
<td>1</td>
<td>Perinatal survival in neonate with intestinal volvulus with intussusception</td>
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<td>Agarwal et al. [25]</td>
<td>2013</td>
<td>1</td>
<td>Fungal infection in neonate with suspicion of intussusception</td>
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<tr>
<td>Moreira-Pinto et al. [18]</td>
<td>2012</td>
<td>1</td>
<td>Neonatal intussusception with Beckwith Wiedemann Syndrome</td>
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<td>Ma et al.4 [4]</td>
<td>2014</td>
<td>1</td>
<td>Neonatal intussusception due to CMV</td>
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<td>Shad et al. [31]</td>
<td>2011</td>
<td>1</td>
<td>Ileo-colic intussusception in pre-mature neonate</td>
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<td>Al-Jahdali et al. [5]</td>
<td>2009</td>
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<td>Colo-colic intussusception with intestinal malrotation</td>
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<td>Loukas et al. [3]</td>
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<td>Intussusception in a Premature Neonate: A Rare Often Misdiagnosed Cause of Intestinal Obstruction</td>
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<td>Slam et al. [14]</td>
<td>2007</td>
<td>1</td>
<td>Sequential intussusception in pre-term neonate</td>
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<td>Ueki et al. [16]</td>
<td>2004</td>
<td>14</td>
<td>Role of hypoxic events in pathogenesis of Intussusception</td>
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<td>Martinez et al. [19]</td>
<td>2004</td>
<td>23</td>
<td>Intussusception in a preterm neonate; a very rare, major intestinal problem -systematic review of cases</td>
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<td>Margenthaler et al. [26]</td>
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<td>Goo et al. [29]</td>
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<td>Wang et al. [1]</td>
<td>1998</td>
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<td>Prenatal and neonatal intussusception</td>
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<td>Mooney et al. [23]</td>
<td>1996</td>
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<td>Cunningham et al. [27]</td>
<td>1980</td>
<td>1</td>
<td>Intussusception due to saccular duplication cyst</td>
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<tr>
<td>Patriquin et al. [28]</td>
<td>1977</td>
<td>12</td>
<td>Neonatal Intussusception</td>
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Table 1 - Studies describing the patients of neonatal intussusception.
is delayed in many patients; and small bowel intussusception is more common in neonates, the use of pneumatic and contrast enemas have no defined therapeutic role [19].

CONCLUSION

Neonatal intussusception is an uncommon entity, which may be confused with other neonatal abdominal emergencies. Although its symptoms may mimic many other conditions like NEC or malrotation of gut, a high degree of suspicion may help in arriving at a proper diagnosis. Since USG appears to be helpful in establishing the diagnosis, it may be used as a screening tool in every patient of neonatal intestinal obstruction and NEC. High index of suspicion, proper evaluation, and timely intervention may fetch satisfactory results.

By increasing the use of USG abdomen in neonatal small bowel pathologies, especially NEC, early diagnosis of neonatal intussusception may be possible. In such a scenario, evaluation of pneumatic and contrast enema may be conducted for framing guidelines.

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REFERENCES


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