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MAIN TOPIC

R. T. Blakelock · S. W. Beasley

The clinical implications of non-idiopathic intussusception

Abstract A pathological lesion can be identified at the leadpoint of intussusception in about 6% of episodes. Occasionally, general manifestations of an underlying disease indicate the specific cause of an intussusception (e.g., perioral pigmentation in Peutz-Jeghers syndrome), but usually the clinical features provide no clues as to the aetiology. Neonatal intussusception may be caused by a duplication cyst or Meckel's diverticulum. Beyond 12 months, the proportion of intussusceptions due to a pathological lesion at the leadpoint increases with age. There is an identifiable lesion in the majority of children over 5 years of age. Postoperative intussusception accounts for between 0.5% and 16% of intussusceptions, although it has a variety of causes; it typically follows retroperitoneal dissection. It is unusual for an intussusception due to a pathological lesion at the leadpoint to be reduced by enema. If it is reduced, the lesion may be seen at the time of reduction or fluoroscopy, or subsequently on ultrasonography.

Key words Intussusception · Leadpoint · Postoperative · Air enema

Introduction

The majority of episodes of intussusception are considered to be idiopathic in origin, in that there is no identifiable pathological lesion at the leadpoint. In the remainder, a definite pathological lesion can be identified by imaging or at the time of surgery. The reported incidence of a pathological lesion at the leadpoint ranges between 1.5% and 12%, with an overall incidence of about 6% (Table 1) [2, 3, 13, 14, 17–19, 31, 32, 34, 36]. Management usually involves resection of this lesion. There are few indications prior to surgery that a pathological lesion is present except in children who may have a recognisable condition that is known to predispose to intussusception, e.g., Peutz-Jeghers syndrome, Henoch-Schönlein purpura., and even less frequently, where a lesion has been observed radiologically or on ultrasonography (US) during attempted reduction. Age is a further factor that is related to the frequency of intussusception being caused by a pathological lesion. Rarely, intra-abdominal surgery, particularly when it involves retroperitoneal dissection, may precipitate intussusception in the early postoperative period, but in many of these cases there is no specific lesion at the leadpoint.

This review focuses on several aspects of non-idiopathic intussusception: (1) A description of the clinical clues that indicate there might be a pathological lesion at the leadpoint; (2) The relationship of age to the likelihood that an underlying pathological lesion has caused the intussusception; (3) Examination of the clinical features and possible aetiology of postoperative intussusception; (4) Whether pathological lesions at the leadpoint can be diagnosed on imaging during attempted enema reduction; and (5) The likelihood that a pathological lesion can be reduced by an enema.

Clinical manifestations of a pathological lesion at the leadpoint in intussusception

Vomiting, abdominal pain, and bloody stools are often considered to be the main symptoms found in children with intussusception, although only about 20% have this classic triad [5]. Lethargy and pallor are more subtle clinical features, recognition of which is important if a delayed diagnosis is to be avoided. These features appear to apply equally to non-idiopathic cases as they do to intussusceptions caused by an identifiable pathological lesion, such that the typical clinical presentation does not indicate the aetiology. The duration of symptoms at presentation does not seem to indicate that there is a

R.T. Blakelock · S.W. Beasley (✉)
Department of Paediatric Surgery, Christchurch Hospital,
Private Bag 4710, Christchurch,
New Zealand

Table 1 Incidence of pathological lesions at the leadpoint in intussusception [2, 3, 13, 14, 17–19, 31, 32, 34, 36]

Pathology of lesion	Frye 1970	Auldist 1970	Glerup 1972	Wayne 1973	Atkinson 1976	Ein 1976	Du 1978	Eklof 1980	Raudkivi 1981	Pang 1989	Ong 1990	Total
Meckel's diverticulum	2	10	10	19	6	14	1	9	8	6	27	112
Omphalo-mesenteric duct		1										1
Duplication cyst		3	1			5		1			4	14
Lymphoma/ lympho-sarcoma	1	3	1	5	2	1	1	1		4	5	24
Appendix	1		1		2	1	2	1				8
Henoch-Schonlein Purpura		1				1	1					3
Peutz-Jeghers familial polyposis	2		2	1		8		3		1	3	20
Postoperative		1	1			1					4	7
Benign intestinal neoplasms									2	6		8
Ectopic gastric mucosa		1										1
Ectopic pancreatic mucosa										1		1
Others		4	2	1			1	2		3	13	26
Total with lesion at leadpoint	6	24	18	26	10	31	6	17	10	20	56	224
Total intussuscep-tions in series	216	203	288	344	66	569	163	658	98	261	602	3,468
Percentage intussuscep-tions with lesion at leadpoint	2.8	12	6.3	7.6	1.5	5.4	3.7	2.6	10	7.7	9.3	6.5

pathological leadpoint [31], although some authors have suggested that the duration of symptoms may be longer in this group of children [14, 37]. Apart from the age of the child at presentation, there are probably no distinguishing features of acute intussusception to indicate that the child has a pathological lesion.

The only clinical features that may be indicative of a pathological lesion at the leadpoint are other manifestations of an underlying disease that has more generalised signs and has a recognised association with intussusception. For example, a child with manifestations of Peutz-Jeghers syndrome, such as pigmentation of the lips and buccal mucosa, who develops clinical signs of intussusception is likely to have a hamartomatous polyp at the leadpoint. Likewise, a child with a family history of familial polyposis may have a adenomatous polyp as a leadpoint. In these examples, the underlying causative disease is usually known prior to the onset of symptoms. In other situations, the development of intussusception may be the first sign of disease. An example of this is Henoch-Schönlein purpura, where the abdominal features of intussusception may precede the development of the purpuric rash, polyarthralgia, or acute glomerulonephritis by a day or two. It should be remembered that these children may develop moderately severe abdominal pain without intussusception [9]. Children with cystic fibrosis who develop acute abdominal signs and symptoms should have intussusception included in their differential diagnosis [1], but it must be distinguished from appendicitis, the other acute abdominal condition that may occur in these children [25]. Fecal accumulation and so-called ''meconium ileus equivalent'' are much more common as a cause of abdominal symptoms in cystic fibrosis.

Influence of age on likelihood of a pathological lesion at the leadpoint

The majority of patients with intussusception present at between 3 and 12 months of age [3, 31, 32]. Before the age of 3 months, intussusception is rare and is likely to

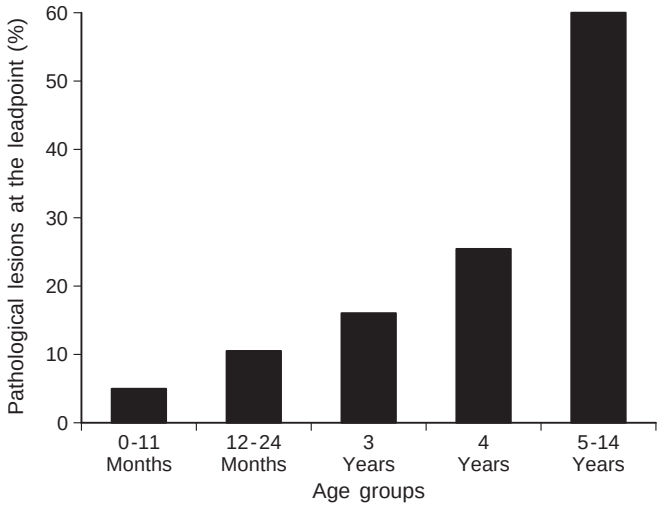

Fig. 1 Percentage of intussusceptions with pathological lesion at the leadpoint in different age groups [3, 31, 32]

be due to a congenital malformation, e.g. a cystic duplication or Meckel's diverticulum [4]. Beyond the normal peak incidence of idiopathic intussusception, the proportion of episodes of intussusception due to a pathological leadpoint increases with age (Fig. 1). Yet, paradoxically, in terms of absolute numbers, the 1st year of life has the greatest number of episodes of intussusception caused by a pathological lesion [31]. The most common lesion that causes intussusception is a Meckel's diverticulum, and this can present at any age. Lymphoma (or lymphosarcoma), the next most common pathology in some series, usually occurs in the older child [32, 37]. In adults, intussusception is only rarely idiopathic in origin. In one series, a lesion was identified in 32 of 33 (97%) patients over 14 years of age [32], compared with an overall incidence of 6.5% in children (Table 1).

Intussusception is seen occasionally in the newborn [35], and even in premature infants [39]. Intrauterine intussusception has been described [35, 39], and is thought to be a cause of intestinal atresia [21, 33].

Postoperative intussusception

Postoperative intussusception is a rare but well-recognised clinical entity. Its reported incidence ranges from 0.5% to 16% [3, 6, 11, 15, 23, 30] of all cases of intussusception and in 0.08% of all paediatric laparotomies [15]. Most cases occur after major abdominal surgery, especially procedures that involve extensive retroperitoneal dissection such as nephrectomy for Wilms' tumour [7, 10, 15, 23, 26, 38]. It has also been reported to follow other more distant procedures such as cervical lymph-node dissection, thoracic procedures, and ventriculo-atrial shunt insertion [28, 30, 38].

Whereas 77% (1,462/1,904) of idiopathic intussusception occur within the first 12 months of life (Fig. 1), about 67% of postoperative intussusceptions occur after the age of 12 months [15, 23]. Symptoms commence within 2 weeks of surgery in 71%–90% of cases [15, 23, 30, 38], in contrast to postoperative intestinal obstruction due to adhesions, which occurs beyond 2 weeks in 75% of cases. The clinical distinction between the two causes of obstruction may be difficult to make, and most children will be thought to have an adhesive obstruction initially. In post-operative intussusception the leadpoint tends to be proximal, involving the jejunum or proximal ileum [6, 24].

One reason postoperative intussusception may go unnoticed initially is that there may be no palpable abdominal mass or rectal bleeding. There is already a cause of abdominal pain and vomiting, due to the proximity of the previous abdominal or retroperitoneal surgery [6]. Vomiting and abdominal distension in abdominal operations are more likely to be associated with postoperative ileus early on, or adhesive small-bowel obstruction subsequently. The key to diagnosis is to consider the possibility of intussusception in a child who fails to progress as expected, particularly if there are persisting clinical and radiological findings consistent with a small-bowel obstruction. A contrast enema rarely demonstrates an intussuscipiens [26], as almost all commence in the small bowel and do not extend into the colon. Abdominal US may be helpful [8], and is probably the investigation of choice. These children require laparoscopy or laparotomy: most of these intussusceptions can be reduced manually, and resection is only rarely required [9].

The aetiology of postoperative intussusception is varies, and presumed causes include: a leadpoint from a bowel suture line or appendiceal stump; disordered intestinal motility secondary to extensive retroperitoneal dissection, postoperative oedema, and bowel handling; electrolyte disturbances affecting peristalsis; radiation therapy; chemotherapy (e.g., vincristine); anaesthesia; and drug effects [11, 22, 23, 26–28, 30, 31].

Can pathological lesions at the leadpoint be diagnosed radiologically, and can they be reduced?

There has been controversy as to how the child with a suspected pathological leadpoint should be managed. It has been claimed that it is not possible to reduce such an intussusception by enema [14, 29], but this is now known to not be so [16]. It is clear that an intussusception due to a pathological leadpoint can only rarely be reduced: there were only 3 pathological leadpoints reduced by enema in over 1,000 consecutive children with intussusception treated at the Royal Children's Hospital, Melbourne. The majority of pathological leadpoints are benign (Table 1). Reduction of a malignant lesion at the leadpoint has been reported, but this is extremely rare and is likely to be recognised at the time of reduction [12].

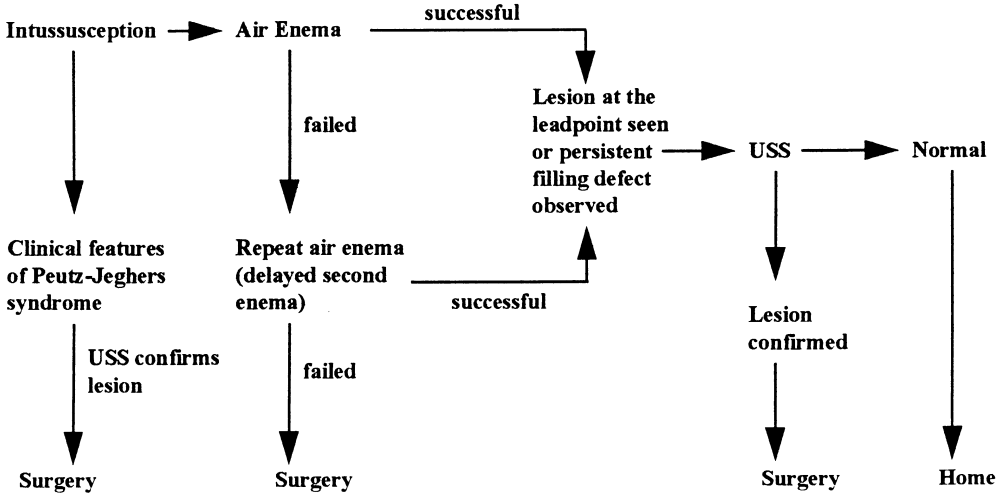

Fig. 2 Algorithm for management of intussusception where a pathological lesion at the leadpoint is a likely consideration (*USS* ultrasonography)

In a review of 12 children with pathological lesions at the leadpoint, Miller et al. [29] reduced the intussusception with an air enema in 7 children, but identified the lesion in only 2 of these at initial flouroscopy. In the other 5 cases that were successfully reduced, the lesions were not found at the time of the air enema, but the use of a either a barium enema, US, or a combination of both demonstrated a lesion at the leadpoint. Three children were unsuccessfully reduced, and the lesion at the leadpoint was not detected at initial fluoroscopy. Fluoroscopy failed to reveal an intussusception or a pathological lesion in a further 2 patients who required surgery for persisting small-bowel obstruction. These authors have recommended US in children suspected of having a pathological leadpoint, recurrent intussusception, or persistent abnormal filling defects on fluoroscopy [29].

It would seem appropriate to attempt enema reduction in all patients with suspected intussusception regardless of age, provided they do not have clinical evidence of peritonitis, septicaemia, or other signs of necrotic bowel [20]. In the older child, there must be a high index of suspicion that there is a pathological lesion at the leadpoint, and this must be looked for during enema reduction (Fig. 2). Where suspicion remains high despite successful enema reduction, it would not be unreasonable to perform US post-reduction. Likewise, further imaging (preferably by US) during the same hospital admission or shortly thereafter would seem appropriate if there is a persistent filling defect post-reduction. A persistent lesion identified on subsequent examination is an indication for surgical intervention.

References

1. Agrons GA, Corse WR, Markowitz RI, Suarez ES, Perry DR (1996) Gastrointestinal manifestations of cystic fibrosis: radiologic-pathologic correction. Radiographics 16: 871–893
2. Atkinson GO, Gay BB Jr, Naffis D (1976) Intussusception of the appendix in children. Am J Roentgenol 126: 1164–1168
3. Auldist AW (1970) Intussusception in a children's hospital: a review of 203 cases in seven years. Aust N Z J Surg 40: 136–143
4. Beasley SW (1996) Intussusception. In: Puri P (ed) Newborn surgery. Butterworth-Heinemann, London, pp 404–407
5. Beasley SW, Auldist AW, Stokes KB (1988) The diagnostically difficult intussusception: its characteristics and consequences. Pediatr Surg Int 3: 135–138
6. Bodycomb J, Beasley SW, Auldist AW (1987) Post-operative intussusception. Pediatr Surg Int 2: 108–109
7. Brown RA, Stunden RJ, Millar AJ, Cywes S (1986) Postoperative intussusception in the paediatric patient. J R Coll Surg Edinb 31: 169–170
8. Carnevale E, Graziani M, Fasanelli S (1994) Post-operative ileo-ileal intussusception: sonographic approach. Pediatr Radiol 24: 161–163
9. Choong CK, Beasley SW (in press) Intraabdominal manifestations of Henoch-Schönlein purpura. J Paediatr Child Health
10. Cohen MD, Baker M, Grosfeld JL, Siddiqui A (1982) Postoperative intussusception in children with neuroblastoma. Br J Radiol 55 651: 197–200
11. Cox JA, Martin LW (1973) Postoperative intussusception. Arch Surg 106: 263–266
12. Devred P, Faure F, Padovani J (1984) Pseudotumoral cecum after hydrostatic reduction of intussusception. Pediatr Radiol 14: 295–298
13. Du JN (1978) Ten years' experience in the management of intussusception in infants and children by hydrostatic reduction. Can Med Assoc J 119: 1075–1076
14. Ein SH (1976) Leading points in childhood intussusception. J Pediatr Surg 11: 209–211
15. Ein SH, Ferguson JM (1982) Intussusception–the forgotten postoperative obstruction. Arch Dis Child 57: 788–790
16. Ein SH, Shandling B, Reilly BJ, Stringer DA (1986) Hydrostatic reduction of intussusceptions caused by lead points. J Pediatr Surg 21: 883–886
17. Eklof OA, Johanson L, Lohr G (1980) Childhood intussusception: hydrostatic reducibility and incidence of leading points in different age groups. Pediatr Radiol 10: 83–86
18. Frye TR, Howard WH (1970) The handling of ileocolic intussusception in a pediatric medical center. Radiology 97: 187–191
19. Gierup J, Jorulf H, Livaditis A (1972) Management of intussusception in infants and children: a survey based on 288 consecutive cases. Pediatrics 50: 535–546
20. Glover JM, Beasley SW, Phelan E (1991) Intussusception: effectiveness of gas enema. Pediatr Surg Int 6: 195–197
21. Grosfeld JL, Clatworthy HW Jr (1970) The nature of ileal atresia dur to intrauterine intussusception. Arch Surg 100: 714–717

22. Gross K, Desanto A, Grosfeld JL, West KW, Eigen H (1985) Intra-abdominal complications of cystic fibrosis. J Pediatr Surg 20: 431–435
23. Holcomb GWD, Ross AJD, O'Neill JAJ (1991) Postoperative intussusception: increasing frequency or increasing awareness? South Med J 84: 1334–1339
24. Hulbert WC Jr, Valvo JR, Caldamone AA, Putnam TC, Emmens RW, Rabinowitz R (1983) Intussusception following resection of Wilms' tumor. Urology 21: 578–580
25. Jacobs S, Beasley SW, Robertson C, Stokes KB (1988) Appendicitis in cystic fibrosis: can the morbidity be reduced? Pediatr Surg Int 3: 414–417
26. Kaste SC, Wilimas J, Rao BN (1995) Postoperative small-bowel intussusception in children with cancer. Pediatr Radiol 25: 21–23
27. Kiesling VJ Jr, Tank ES (1989) Postoperative intussusception in children. Urology 33: 387–389.
28. McGovern JB, Gross RE (1968) Intussusception as a postoperative complication. Surgery 63: 507–513
29. Miller SF, Landes AB, Dautenhahn LW, Pereira JK, Connolly BL, Babyn PS, et al (1995) Intussusception: ability of fluoroscopic images obtained during air enemas to depict lead points and other abnormalities. Radiology 197: 493–496
30. Mollitt DL, Ballantine TV, Grosfeld JL (1979) Postoperative intussusception in infancy and childhood: analysis of 119 cases. Surgery 86: 402–408
31. Ong NT, Beasley SW (1990) The leadpoint in intussusception. J Pediatr Surg 25: 640–643
32. Pang LC (1989) Intussusception revisited: clinicopathologic analysis of 261 cases, with emphasis on pathogenesis. South Med J 82: 215–228
33. Rachelson MH, Jernigan JP, Jackson WF (1955) Intussusception in the newborn infant. J Pediatr 47: 87–94
34. Raudkivi PJ, Smith LH (1981) Intussusception: analysis of 98 cases. Br J Surg 68: 645–648
35. Todani T, Tabuchi K, Tanaka S (1975) Intestinal atresia duc to intrauterine intussusception: analysis of 24 cases in Japan. J Pediatr Surg 10: 445–451
36. Wayne ER, Campbell JB, Burrington JD, Davis WS (1973) Management of 344 children with intussusception. Radiology 107: 597–601
37. Wayne ER, Campbell JB, Kosloske AM, Burrington JD (1976) Intussusception in the older child—suspect lymphosarcoma. J Pediatr Surg 11: 789–794
38. West KW, Stephens B, Rescorla FJ, Vane DW, Grosfeld JL (1988) Postoperative intussusception: experience with 36 cases in children. Surgery 104: 781–787
39. Yoo RP, Touloukian RJ (1974) Intussusception in the newborn: a unique clinical entity. J Pediatr Surg 9: 495–498