

A Review of Ileal Atresia*

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The ileum has played quite a prominent part in the history of small bowel atresia; 295 years ago small intestinal atresia was reported¹ in a stillborn child, the terminal ileum being the site affected; and thirteen years later a girl who lived for 22 days was found at autopsy to have complete discontinuity of the lower ileum.² Bland Sutton based his classic review of intestinal atresia in 1889³ on the report of a 48 hour old child in whom he diagnosed ileal atresia and attempted operative relief probably the first recorded attempt. His correct diagnosis in a living child was achieved by exclusion; milk feeds had been tolerated thus excluding what he described as atresia of the pharynx; and meconium had been passed confirming a normal anus.

Recent experience suggesting that results in ileal atresia were significantly worse than those in jejunal atresia led us to review our material.

Material

A total of 53 jejunal atresias and 52 ileal atresias admitted between 1954 and 1977 were analyzed. Distribution between the sexes was similar, but it was noteworthy that 22 of the babies with jejunal atresia weighed less than 2.5 kg at birth and only eight of those with ileal atresia—the reverse of the findings of the review by the Surgical Section of the American Academy of Pediatrics.⁴ On the other hand major associated anomalies were more common in the ileal group (Table I). These included, predictably, five cases with major umbilical anomalies with or without ectopia vesicae and three congenital heart lesions. Less predictable were one child with oesophageal atresia and one with Downs syndrome.

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TABLE I
DISTRIBUTION OF CASES, INCIDENCE OF LOW BIRTH WEIGHT, MAJOR ASSOCIATED ANOMALIES AND PRE-OPERATIVE COMPLICATIONS

	Jejunum	Ileum
Total cases	53	52
Under 2.5 kg birth weight	22	8
Major associated anomalies	2	10
Pre-operative complications	18	35
Deaths	21	15

Pre-operative complications were also more common in the ileal than in the jejunal group occurring almost twice as often. These included 20 children with volvulus, which might have been the cause of the atresia or the result of it-the heavy packed blind end rotating on its mesentery; there were also 24 children who had evidence of intrauterine perforation, with calcification, a meconium cyst or frank meconium peritonitis; and nine children with pneumoperitoneum or abscess formation resulting from gut perforation after birth (Table II).

TABLE II
PRE-OPERATIVE COMPLICATIONS

	Jejunum	Ileum
Volvulus	7	13
Calcification, meconium cyst or meconium peritonitis	9	15
Perforation	2	7
Total	18	35

Every child with jejunal atresia had vomitted and all but one had abdominal distension. In ileal atresia ten of the 52 children had never vomitted and nine did not show abdominal distension.

The time of onset of the first symptoms was difficult to find in every case, but certainly half of the jejunal atresias were admitted under 24 hours compared with a quarter of the ileal atresia, suggesting the earlier appearance of symptoms (Table III) Furthermore, of the 52 patients with ileal atresia, 17 did not have a positive diagnosis of intestinal obstruction on admission, while in 49 of the 53 cases of jejunal atresia were accurately diagnosed.

Both jejunal and ileal atresia carry a high mortality; a slight improvement in the second half of the series applies to ileal atresia, but not to jejunal atresia.

TABLE III
AGE ON ADMISSION

	Jejunum	Ileum
Under 24 hours	27	14
24-47 hours	15	22
48-71 hours	5	7
74-95 hours	3	3
Over 96 hours	3	6

Of the 15 deaths from ileal atresia six seemed virtually unavoidable. Three had cloacal exstrophy and one had a severe congenital heart lesion. One child, who was operated on before he was twelve hours old, had a massive midgut volvulus and even though resection was delayed until a second look procedure he was left after resection of gangrenous bowel with insufficient small intestine for survival. Another child, also operated on at under twelve hours, died within 24 hours of fulminant *E. coli* septicaemia: his dilated proximal end had perforated within a few hours of birth.

Of the avoidable deaths two could be attributed to late referral, both over 72 hours old on admission, one having extensive gangrene due to a volvulus and one already having necrotising enterocolitis and *Klebsiella* septicaemia.

Two babies died in the immediate postoperative period and might have been saved by more vigorous support; one had an enormous meconium cyst and one had his oesophageal atresia repaired before the abdomen was explored to deal with the ileal atresia.

Two babies died as a direct result of an anastomotic leak and two more died aged two months and three months from late complications; one developing a mesenteric thrombosis during an epidemic of severe diarrhea and one inhaling vomitus during a period of subacute obstruction. One other baby died as a result of a persistent fistula from the anastomosis and only at postmortem was a duplicated colon found that was causing distal obstruction.

In addition to those patient; who died, three had pneumonia, two had septicaemia and two had complications attributable to some leakage at the anastomosis.

Discussion

There is a tendency to attribute a proportion of the mortality and morbidity in intestinal atresias to late referral, but this is not born out

in this series of ileal atresia. Of those 15 patients who died only two might have been saved had they been referred earlier. However, pneumonia, which occurred in only seven cases of ileal atresia compared with sixteen of jejunal atresia, was seen almost exclusively in those cases of ileal atresia referred when over 48 hours old, in three patients pneumonia may have contributed to their death and in two more their period of hospitalization extended beyond eight weeks. Delayed diagnosis may thus contribute to morbidity as well as to mortality.

Apart from delay in diagnosis, 17 patients were admitted with the wrong diagnosis, three were correctly but incompletely diagnosed, being referred on account of other major anomalies. Many of the remaining 14 had already vomited bile. Abdominal distension is rarely missed in a special care unit and leads to immediate referral to the surgeon; green vomitus is perhaps more often missed, and even when noted does not seem to carry the same urgency; further supporting evidence of intestinal obstruction is awaited. In seven of the 14 cases a record of the passage of normal meconium led to a false sense of security. It is perhaps not generally realized how often normal meconium may be passed even in distal obstruction.

However, our figures suggest that in more recent years cases of ileal atresia have been referred rather earlier and the paediatricians are entitled to claim responsibility for at least a part of the improved results.

From surgical point of view, major changes in operative technique have not been introduced. Anastomotic leak not only accounted for two deaths but also for complications such as fistulae, abscesses and obstruction in other cases. Like Nixon and Tawes⁵ we found most of the leaks were in babies under 2.5 kg and a modified technique may be required in this situation.

Massive intestinal resection is less common in ileal atresia than in jejunal atresia. Of twelve children with jejunal atresia who were left with a short small bowel, nine died, suggesting severely inadequate absorption, whereas of nine children with ileal atresia recorded as having short bowel only two died. Total as well as partial parenteral feeding may improve survival rates in these cases.

With an overall mortality of around 30 % we still do not have much to be proud of. There is a little room for improvement in the operative technique of anastomosis, but much more for improvement of the care of the associated anomalies and the control of infection.

REFERENCES

1. Goeller, G. C.: Abortus humani monstrosi. Norimb. Hist. Anatom. Misc. Acad. Nat. Curios., 1963.
2. Horch, C.: De puella cum coalitu intestini ilei nata, et per vaginam duos dei vivente. Frankof. Misc. Acad. Nat. Curios. 1966.
3. Bland Sutton, J.: Imperforate Ileum. Am. J. Med. Sci. 98: 457, 1889.
4. de Lorimier A., Fonkalsrud, E. W., Hays, D. M.: Congenital atresia and stenosis of the jejunum and ileum. Surgery 65: 819, 1969.
5. Nixon H. H., Tawes, R.: Etiology and treatment of small intestinal atresia. Surgery 69: 41, 1971.