Longitudinal intestinal lengthening and tailoring: results in 20 children

A Bianchi MD FRCS

J R Soc Med 1997;**90**:429–432 JOINT MEETING SECTIONS OF PAEDIATRICS AND SURGERY, 29 JANUARY 1997

SUMMARY

Longitudinal intestinal lengthening and tailoring (LILT) is increasingly favoured as a treatment for short-bowel syndrome. In a personal series, 20 children underwent LILT between 1982 and 1997.

There was negligible morbidity (hemiloop anastomotic stenosis in 2) and no operative mortality. At a mean follow-up of 6.4 years overall survival was 45%, and certain features were clearly related to outcome. Survivors had >40 cm of residual small bowel (commonly jejunum) and had little hepatic dysfunction despite parenteral nutrition of similar time and nature as non-survivors. Children who did not survive had <40 cm of residual small bowel and developed early lethal hepatic dysfunction of unclear aetiology. Outcome did not seem to be influenced by the presence of the ileocaecal valve or the length of residual colon.

Children born with short-bowel should be offered LILT at an early stage when still in good physical condition, so as to avoid liver-damaging intraluminal stasis and bacterial translocation and to enhance intestinal adaptation and hepatoprotective factors.

INTRODUCTION

The management of a child with short bowel is difficult, the course hazardous and the outcome uncertain. Notwithstanding, developments in bowel transplantation[1,2] and progressive imaginative concepts in autologous gastrointestinal reconstruction[3–5] sustain hope for the future. Here I analyse a personal series of 20 children with neonatal short-bowel syndrome who underwent longitudinal intestinal lengthening and tailoring (LILT) between 1982 and 1997.

SURGICAL METHOD

The technique for longitudinal bowel lengthening and tailoring is essentially that described in 1980[6] and modified in 1984[7] with bipolar bowel division and manual suture in preference to the GIA stapler. The dilated bowel is carefully mobilized with preservation of all blood vessels, and the longest segment possible is divided along the antemesenteric border. Distraction of the bowel to either side away from the midline and against the mesenteric attachment opens an inverted triangular space within the mesentery bordered by the mesenteric vessels and the bowel wall. Longitudinal bowel division along the mesenteric border develops two vascularized hemisegments which are tubularized to form hemiloops with a continuous inverting 6″0″ Maxon running suture knotting every fourth throw. Bowel continuity is established with a jejuno-colonic anastomosis and a wide oblique hemiloop anastomosis in the original S-shape, or the Aigrain spiral[8], whichever causes least traction on the blood supply. The stomach is drained through a nasogastric tube or a Stamm tube gastrostomy which is subsequently used for feeding.

EARLY MANAGEMENT

Since 1982, 20 children have undergone LILT for short-bowel syndrome. Surgery was undertaken following in-depth counselling and consultation with parents. 10 children, of whom 3 survive, were born with gastroschisis and 10, of whom 6 survive, had small-bowel atresia. The residual bowel was dilated (>5 cm in the neonate), non-propulsive and associated with intraluminal stasis and bacterial translocation. Cholestatic jaundice, often beginning early, was present in all patients. The hepatic dysfunction was progressive in 10 children, whereas in 9 liver function returned to normal as enteral absorption improved. One jaundiced child with improving liver function died from Gram-negative septicaemia. Details of the original anomaly, bowel length measured along the antemesenteric border of the unstretched bowel at the time of LILT, the presence of the ileocaecal (IC) valve, the length of residual colon, the age at LILT, the post-LILT length, and the eventual outcome are outlined in Tables 1 and 2. All children were maintained on intravenous nutrition delivered through a central venous catheter

Neonatal Surgical Unit, St Mary's Hospital, Whitworth Park, Manchester M13 0JH, England

Table 1 **Details of 9 survivors of 20 LILT patients**

Anomaly	Jejunum	IC valve	Colon	Age at LILT	Post-LILT	Outcome
SB atresia	46 cm	+	All	12 weeks	88 cm	Off IVA
Gastroschisis	45 cm	+	All	35 months	84 cm	Off IVA
Gastroschisis	45 cm	–	Left colon	15 weeks	90 cm	Off IVA
SB atresia	98 cm	–	Left colon	92 months	118 cm	Off IVA
SB atresia	45 cm	–	Mid-transv	41 months	90 cm	IVA*
SB atresia	38 cm	+	All	7 weeks	63 cm	Off IVA
SB atresia	90 cm	–	Mid-transv	13 weeks	120 cm	Off IVA
SB atresia	40 cm	–	Mid-transv	15 months	58 cm	Off IVA
Gastroschisis	110 cm†	–	Mid-transv	17 months	166 cm	IVA*

SB=small bowel.
*Still on partial intravenous alimentation
†Originally measured at 27 cm

Table 2 **Details of 11 non-survivors of 20 LILT patients**

Anomaly	Jejunum	IC valve	Colon	Age at LILT	Post-LILT	Outcome
Gastroschisis	35 cm	–	Mid-transv	18 weeks	62 cm	Hep fail
Gastroschisis	30 cm	–	Mid-transv	3 weeks	54 cm	Hep fail
SB atresia	33 cm	+	All	6 weeks	51 cm	Hep fail
Gastroschisis	39 cm	+	All	4 weeks	54 cm	Hep fail
SB atresia	58 cm	–	Mid-transv	14 weeks	73 cm	Hep fail
SB atresia	33 cm	–	Mid-transv	22 weeks	63 cm	Hep fail
SB atresia	60 cm*	–	Left	10 weeks	98 cm	Hep fail
Gastroschisis	36 cm	–	Mid-transv	11 weeks	58 cm	Hep fail
Gastroschisis	25 cm	–	Mid-transv	17 weeks	50 cm	Hep fail
Gastroschisis	28 cm	–	Mid-transv	8 weeks	56 cm	Hep fail
Gastroschisis	90 cm	–	Mid-transv	7 weeks	123 cm	Gram-neg sepsis

Hep fail=Hepatic failure
*Originally measured at 26 cm

(Nutracath, 6.6 F Broviac). Venous access was carefully managed and access sites were preserved by repeated use of the same site for replacement of blocked or infected feeding catheters. In the latter part of the series continuous bowel sterilization with enteral kamamycin (10 mg/kg four times daily) or tobramycin (10 mg/kg four times daily) as well as metronidazole (10 mg/kg four times daily) was instituted at diagnosis and maintained over the long term. Enteral feeding was started early with breast milk or a predigested formula (Pregestimil, Mead Johnson BV, Nijmegen, Netherlands) and was determined by bowel tolerance. Intravenous nutrition was tailored according to enteral absorption.

FOLLOW-UP

All children recovered rapidly from the operative intervention, with mouth-to-anus transit by the third to fifth postoperative day. None of the suture lines leaked and there were no interloop fistulae. In the early part of the series 2 children required further laparotomy because of stenosis at the hemiloop anastomosis. Adoption of a wide oblique anastomosis subsequently avoided this complication. Post-LILT there was a reduction in the incidence of bacterial translocation and less central-line-related sepsis. All children showed an initial increase in enteral absorption, allowing reduction in intravenous alimentation.

At a mean follow-up of 6.4 years (range 0.5–15), overall survival is 45%. Of the 9 long-term survivors, 5 have been followed for more than 5 years. 7 of the 9 progressed to full enteral absorption and dispensed with intravenous feeding, usually within 8–16 weeks from operation; 2 still require partial parenteral support. All are able to take an unrestricted diet, often demonstrating hyperphagia. These 9 children were free of serious hepatic dysfunction and eventually showed near-normal physical

and mental growth, rapidly catching up on developmental milestones. Liver indices steadily returned to normal as enteral function improved. No child has developed cholelithiasis or urinary oxalate stones. 2 children with high urinary oxalate excretion have been prescribed a low oxalate diet. All are given vitamin B_{12} supplements parenterally. The number of stools decreased from 10–15 liquid motions per day to an average of 1–4 semisolid motions. All children over 4 years old are continent of faeces. 3 have gone through normal puberty without additional intravenous nutritional supplements.

After early postoperative improvement the second group of 10 children showed steady overall deterioration. Hepatic dysfunction (cholestatic jaundice) of early onset, frequently antedating LILT, followed a relentless and rapid progression. The serum alkaline phosphatase and aminotransferases rose, as did bilirubin, and serum albumin began to fall. The initial improvement in enteral absorption reversed and dependence on intravenous nutrition again increased. These children showed poor weight gain and development. Sepsis became more troublesome, with pharyngeal and skin organisms as well as gut-related bacteria. All eventually developed splenomegaly and portal hypertension and died from end-stage hepatic failure. One other child, whose enteral absorption was improving, died from Gram-negative septicaemia.

DISCUSSION

The problems relating to the short-bowel state, the management of the associated intestinal insufficiency and the intricacies of the intestinal adaptation response have been repeatedly reviewed[9–11]. Management of the short-bowel patient consists essentially of intravenous alimentation and early enteral nutrition, with pharmacological and surgical manipulation of the residual bowel to reduce stasis and bacterial translocation and enhance intestinal adaptation and function. Central venous access for parenteral nutrition is a major surgical responsibility. Loss of venous access is a life-threatening complication: infected or blocked feeding catheters should not be removed but replaced as often as necessary, through the same venous access site.

Surgical techniques for autologous gastrointestinal reconstruction have been varied and imaginative[3] and since 1980 LILT has received considerable attention and application. The procedure is designed to tailor the residual dilated small bowel to propulsive dimensions and to increase isoperistaltic small-bowel length. The LILT procedure has been reported safe with only a few complications[12,13] and many children have benefited from a sustained improvement in enteral absorption[14–16]. This report confirms such experience. All children showed an early improvement in enteral absorption, allowing reduction in intravenous alimentation. Bowel-related bacterial translocation became less troublesome and central line sepsis decreased with improving general health.

The main features that characterized the survivors was the presence of >40 cm of residual dilated small bowel, together with only minor hepatic dysfunction: presence of the ileocaecal valve, length of residual colon, and age at LILT seemed not to affect the eventual outcome. It is noteworthy, however, that most children in this group underwent LILT quite late, having survived the hazardous neonatal period; thus, they could possibly represent a self-selected group of potential long-term survivors. For these children, operative resolution of the bowel-related problems (dilatation and non-propulsion, stasis and bacterial translocation) was the only residual barrier to intestinal adaptation and enhanced enteral absorption. As overall bowel function improved, liver indices steadily returned to normal despite continuing parenteral nutrition. These observations bring into question the term 'TPN jaundice' and the implication that the intravenous feeding solutions are largely responsible for the cholestatic jaundice in short-bowel syndrome.

Analysis of this series suggests that long-term survival for children who develop short-bowel syndrome during the neonatal phase is largely determined by their liver status. Death from other causes is uncommon. The onset of cholestatic jaundice reflects severe hepatocellular dysfunction which is an ominous occurrence of unclear aetiology. Hepatic dysfunction seems to correlate with bowel length, in so far as this reflects functional small-bowel mass. Thus we can postulate in short-bowel syndrome a substantial reduction in the enterocyte-related 'hepatoprotective factors' passing to the liver through the portal circulation. Alternatively or in association, the loss of ileal Peyer's patches and mesenteric lymphoid tissue may lead to impaired immunological recognition and response. There is evidence that the number and distribution of T-lymphocytes in gut-associated lymphoid tissue (GALT) is critical. In the normal newborn rabbit, increasing spontaneous bacterial translocation coincides with small-bowel colonization and reaches a peak at 6 days[17,18]. Bacterial translocation then rapidly declines as T-lymphocyte-related immunity increases, representing maturation and stabilization of the mucosal barrier[17–19]. The increased T-lymphocyte proliferation following bacterial exposure of germ-free animals supports Crabbe *et al.*[20] in their suggestion that the normal mechanism for GALT stimulation and T-cell proliferation is early spontaneous translocation of intraluminal organisms, potentially occurring predominantly through Peyer's patches. Indeed, colonization of the ileum in the newborn is normally higher than that of the jejunum[19], and it is the ileum and ileal mesentery that carry the greater load of lymphoid tissue. In a report on lymphocyte subpopulations

after extensive small-bowel resection in the rat, Barrena *et al.*[21] did not find any alteration of humoral and B-cell immunity, but noted a substantial reduction in cellular (T-cell related) immunity following 80% jejunoileal resection. This reduction was similar whether the animals were fed enterally or parenterally and despite sustained nutrition; so the immunological response seemed unaffected by the mode and adequacy of nutrition. Their findings suggest that removal or loss of large amounts of lymphoid tissue along with the bowel during extensive midgut loss or resection might be associated with failure or inadequate development of an immune response even when nutritional status is preserved. In this context we might speculate that the reduction in GALT and the reduced T-lymphocyte response to gut-related translocation results in a failure of development of hepatic immunocyte populations, leaving the liver vulnerable to bacterial and viral antigens. In line with these concepts it is also noteworthy that liver dysfunction in children with antenatal short bowel never occurs at a time when the bowel is sterile, but follows on small-bowel colonization and the onset of bacterial translocation.

On the evidence of this series some might argue that LILT should be reserved for the self-selected survivors who have come through the neonatal phase and who have reached a static phase in bowel adaptation and function[22]. However, the results also highlight the fact that death in neonatal short-bowel syndrome is largely due to progressive hepatocellular failure, often of early onset and aggravated by recurrent sepsis and bacterial translocation. It therefore seems appropriate, particularly for high-risk children with <40 cm small bowel, to try to prevent, eliminate or reduce sepsis-related factors—i.e. bowel dilatation, non-propulsion and stasis—and to enhance small-bowel adaptation. This implies a policy of early surgery soon after birth when the child is in good physical condition and free of liver injury.

REFERENCES

1 Reyes J, Tzakis AG, Todo S, Nour B, Starzl T. Small bowel and liver small bowel transplantation in children. *Semin Pediatr Surg* 1993;**2**:289–300

2 Grant D on behalf of the International Intestinal Transplant Registry. Current results of intestinal transplantation. *Lancet* 1996;**347**: 1801–3

3 Bianchi A. Autologous gastro-intestinal reconstruction. *Semin Pediatr Surg* 1995;**4**:54–9

4 Warner BW, Chaet MS. Non-transplant surgical options for management of the short bowel syndrome. *J Pediatr Gastroenterol Nutr* 1993;**17**:1–12

5 Collins JB, Georgeson KE, Vincente Y, Kelly DR, Figueroa R. Short bowel syndrome. *Semin Pediatr Surg* 1995;**4**:60–73

6 Bianchi A. Intestinal lengthening: a technique for increasing small intestinal length. *J Pediatr Surg* 1980;**15**:145–51

7 Bianchi A. Intestinal lengthening: an experimental and clinical review. *J R Soc Med* 1984;**77**:35–41

8 Aigrain Y, Cornet D, Cezard JP, Boureau M. Longitudinal division of small intestine: a surgical possibility for children with the very short bowel syndrome. *Z Kinderchir* 1985;**40**:233–6

9 Georgeson KE, Breaux CW Jr. Outcome and intestinal adaptation in neonatal short bowel syndrome. *J Pediatr Surg* 1992;**27**:344–50

10 Shanbhogue LKR, Molenaar JC. Short bowel syndrome: metabolic and surgical management. *Br J Surg* 1994;**81**:486–99

11 Vanderhoof JA, Langnas AN, Pinch LW, Thompson SS, Kaufman SS. Short bowel syndrome. *J Pediatr Gastroenterol Nutr* 1992;**14**:359–70

12 Huskisson LJ, Brereton RJ, Kiely EM, Spitz L. Problems with intestinal lengthening. *J Pediatr Surg* 1993;**28**:720–2

13 Thompson JS, Vanderhoof AJ, Antonson L. Intestinal tapering and lengthening for short bowel syndrome. *J Paediatr Gastroenterol Nutr* 1985;**4**:495–7

14 Weber TR, Powell MA. Early improvement in intestinal function after isoperistaltic bowel lengthening. *J Pediatr Surg* 1996;**31**:61–4

15 Thompson JS, Pinch LW, Murray N, Vanderhoof JA, Schultz LR. Experience with intestinal lengthening for the short bowel syndrome. *J Pediatr Surg* 1991;**26**:721–4

16 Figueroa-Colon R, Harris PR, Birdson E, Franklin FA, Georgeson KE. Impact of intestinal lengthening on the nutritional outcome for children with short bowel syndrome. *J Pediatr Surg* 1996;**31**:912–16

17 Van Camp JM, Tomaselli V, Drongowski R, Coran AG. Bacterial translocation in the newborn rabbit: effect of age on frequency of translocation. *Pediatr Surg Int* 1995;**10**:134–7

18 Urao M, Teitelbaum DH, Drongowski RA, Coran AG. The association of gut-associated lymphoid tissue and bacterial translocation in the newborn rabbit. *J Pediatr Surg* 1996;**31**:1482–7

19 Urao M, Moy J, Van Camp JM, Drongowski R, Altabba M, Coran AG. Determinant of bacterial translocation in the newborn: small bowel versus large bowel colonization. *J Pediatr Surg* 1995;**30**: 831–6

20 Crabbe PA, Brazin H, Eyssen H, Heremans JF. The normal microbial flora as a major stimulus for proliferation of plasma cells synthesizing IgA in the gut. The germ-free intestinal tract. *Int Arch Allergy Appl Immunol* 1968;**34**:362–75

21 Barrena MJ, Eizaguirre I, Aldazabal P, *et al.* Lymphocyte subpopulations after extensive small bowel resection in the rat. *J Pediatr Surg* 1995;**30**:1447–9

22 Pokorny WJ, Fowler CL. Isoperistaltic intestinal lengthening for short bowel syndrome. *Surgery Gynecol Obstet* 1991;**172**:39–43