

Serial transverse enteroplasty procedure (STEP) for bowel lengthening in parenteral nutrition-dependent children

Interventional procedures guidance

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Your responsibility

This guidance represents the view of NICE, arrived at after careful consideration of the evidence available. When exercising their judgement, healthcare professionals are expected to take this guidance fully into account, and specifically any special arrangements relating to the introduction of new interventional procedures. The guidance does not override the individual responsibility of healthcare professionals to make decisions appropriate to the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

All problems (adverse events) related to a medicine or medical device used for treatment or in a procedure should be reported to the Medicines and Healthcare products Regulatory Agency using the [Yellow Card Scheme](#).

Commissioners and/or providers have a responsibility to implement the guidance, in their local context, in light of their duties to have due regard to the need to eliminate unlawful discrimination, advance equality of opportunity, and foster good relations. Nothing in this guidance should be interpreted in a way that would be inconsistent with compliance with those duties. Providers should ensure that governance structures are in place to review, authorise and monitor the introduction of new devices and procedures.

Commissioners and providers have a responsibility to promote an environmentally sustainable health and care system and should assess and reduce the environmental impact of implementing NICE recommendations wherever possible.

1 Guidance

- 1.1 The evidence on the safety of the serial transverse enteroplasty procedure (STEP) for bowel lengthening in parenteral nutrition-dependent children is adequate; however, there is limited evidence of its efficacy. Therefore, this procedure should not be used without special arrangements for consent and for audit or research.
- 1.2 Clinicians wishing to undertake STEP for bowel lengthening in parenteral nutrition-dependent children should take the following actions.
 - Inform the clinical governance leads in their Trusts.
 - Ensure that parents or carers understand the uncertainty about the procedure's efficacy, in addition to its risks. Provide parents or carers with clear written information. In addition, use of NICE's information for the public recommended.
- 1.3 Patient selection should be carried out in the context of a multidisciplinary team experienced in the management of short bowel syndrome (SBS). These teams should consider case selection carefully so that the procedure is only performed on children who are likely to remain dependent on parenteral nutrition.
- 1.4 Clinicians undertaking STEP for bowel lengthening in parenteral nutrition-dependent children should submit data on all patients to the Boston Children's Hospital's International STEP Data Registry.

2 The procedure

2.1 Indications and current treatments

- 2.1.1 SBS is a rare but serious condition which may either be associated with congenital conditions or follow bowel resection. Patients with short bowel syndrome (SBS) have a rapid intestinal transit time resulting in malabsorption of enteral nutrition and subsequent malnutrition. Most patients with SBS require partial or total parenteral nutrition (TPN), although some will develop adequate intestinal function and no longer require TPN. Intestinal failure combined with TPN can cause liver failure and recurrent sepsis leading to death.
- 2.1.2 SBS can be caused by structural or functional abnormalities.
- 2.1.3 Surgical procedures which prolong bowel transit time and aim to increase the absorption of nutrients include resection of dilated segments of the small intestine, tapering enteroplasty, intestinal plication, or the Bianchi intestinal loop lengthening procedure. Some patients require small bowel and liver transplants.

2.2 Outline of the procedure

- 2.2.1 Serial transverse enteroplasty procedure (STEP) is a surgical technique that aims to lengthen the small intestine of patients with SBS so that they can benefit from enteral nutrition.
- 2.2.2 The procedure is done under general anaesthesia. The procedure relies on the principle that the blood supply to the small bowel originates from the mesentery and traverses the bowel perpendicular to its long axis. An endoscopic stapler is passed through the mesentery and the bowel is simultaneously stapled and dissected on alternating sides in a direction perpendicular to its long axis. The small bowel is left with a zigzag appearance.

2.3 Efficacy

- 2.3.1 In a case series of 38 patients (29 with SBS) at 13-month follow-up, the mortality rate following STEP was 8% (3 out of 38) and 3 patients (8%) went on to require a bowel or liver and bowel transplant. One patient in a case series of 8 developed cholestasis which caused sepsis, liver failure, and subsequently death at 3-month follow-up.
- 2.3.2 The same case series of 38 patients reported an increase in mean bowel length from 68 ± 44 cm at baseline to 115 ± 87 cm in 27 patients where this outcome was measured. A second case series of 5 patients showed an increase in bowel length from 61 cm at baseline to 98 cm immediately following the procedure. A third case series of 8 patients, with 5 patients treated with STEP, reported an increase in mean bowel length of 17 cm, from an average of 62 cm to 79 cm.
- 2.3.3 A case report of a patient treated with STEP for SBS and D-lactic acidosis reported that a normal bowel transit time (2.5 hours) was achieved at 7-day follow-up.
- 2.3.4 In the case series of 38 patients, 21 of whom were on TPN at baseline, there was a mean improvement in the total percentage of calories tolerated enterally from 31% at baseline to 67% ($p < 0.01$) at 13-month follow-up. Of these 21 patients, 10 were completely weaned off TPN (follow-up not stated). In the case series of 5 patients, it was reported that the mean percentage of nutrition received enterally improved from 49% at baseline to 80% in 3 patients at 17-month follow-up ($p < 0.05$). The case series of 8 patients reported that, of the 5 treated with STEP alone, 'more than 50%' were completely weaned off TPN and 1 patient had significantly decreased dependency (not otherwise defined). In two case reports, STEP allowed for 75% and 100% of calorific intake to be achieved enterally at 11 and 7 months, respectively. For more details, see the [overview](#).
- 2.3.5 One Specialist Adviser commented that STEP is one of a range of surgical options, none of which has been shown convincingly to provide benefit. Most of the Specialist Advisers stated that the prognosis of SBS is uncertain and that some patients improve spontaneously without treatment.

2.4 Safety

- 2.4.1 A case series of 5 patients and a case report of 1 patient reported no long-term complications at 15- and 7-month follow-up, respectively.
- 2.4.2 A patient in one case report required nasogastric fluid aspiration postoperatively. This was bilious at first, becoming clear over time. For more details, see the [overview](#).
- 2.4.3 The Specialist Advisers considered this procedure to have low morbidity. They considered the potential risks associated with this procedure to include staple-line leak, bowel obstruction, pleural effusion, haematoma, abscess formation, progression to transplant and mortality.

2.5 Other comments

- 2.5.1 It was noted that safety data are based on small case series but they compare favourably with other surgical treatments for this high-risk group of patients. In view of this, the Committee considered the data adequate in the context of these severely ill children.

3 Further information

Sources of evidence

The evidence considered by the Interventional Procedures Advisory Committee is described in the [overview](#).

Information for patients

NICE has produced [information for the public on this procedure](#). It explains the nature of the procedure and the decision made, and has been written with patient consent in mind.

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Endorsing organisation

This guidance has been endorsed by [Healthcare Improvement Scotland](#).