

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

## 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 the Institute's information for patients ('Understanding NICE guidance') is recommended (available from [www.nice.org.uk/IPG232publicinfo](http://www.nice.org.uk/IPG232publicinfo)).
- 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 International STEP Data Registry at Children's Hospital Boston, USA (available from [www.childrenshospital.org/cfapps/step/index.cfm](http://www.childrenshospital.org/cfapps/step/index.cfm)).

## 2 The procedure

### 2.1 Indications

- 2.1.1 SBS is a rare but serious condition which may either be associated with congenital conditions or follow bowel resection. Patients with 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 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

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### This guidance is written in the following context

This guidance represents the view of the Institute, which was arrived at after careful consideration of the available evidence. Healthcare professionals are expected to take it fully into account when exercising their clinical judgement. This guidance does not, however, override the individual responsibility of healthcare professionals to make appropriate decisions in the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

Interventional procedures guidance is for healthcare professionals and people using the NHS in England, Wales, Scotland and Northern Ireland.

This guidance is endorsed by NHS QIS for implementation by NHSScotland.

simultaneously stapled and dissected on alternating sides in a direction perpendicular to its long axis. The small bowel is left with a zigzag appearance.

Most of the Specialist Advisers stated that the prognosis of SBS is uncertain and that some patients improve spontaneously without treatment.

## 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% (3/38) 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  $68 \pm 44$  cm at baseline to  $115 \pm 87$  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, refer to the 'Sources of evidence' section.
- 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.

## 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, refer to the 'Sources of evidence' section.
- 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.

Andrew Dillon  
Chief Executive  
September 2007

## Information for patients

NICE has produced information describing its guidance on this procedure for patients and their carers ('Understanding NICE guidance'). It explains the nature of the procedure and the decision made, and has been written with patient consent in mind. This information is available from [www.nice.org.uk/IPG232publicinfo](http://www.nice.org.uk/IPG232publicinfo)

## Sources of evidence

The evidence considered by the Interventional Procedures Advisory Committee is described in the following document.

'Interventional procedure overview of serial transverse enteroplasty procedure (STEP) for bowel lengthening in parenteral nutrition-dependent children', March 2007.

Available from: [www.nice.org.uk/ip385overview](http://www.nice.org.uk/ip385overview)

## Ordering information

Copies of this guidance can be obtained from the NHS Response Line by telephoning 0870 1555 455 and quoting reference number N1339. 'Understanding NICE guidance' can be obtained by quoting reference number N1340.

The distribution list for this guidance is available at [www.nice.org.uk/IPG232distributionlist](http://www.nice.org.uk/IPG232distributionlist)

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Interventional procedures guidance makes recommendations on the safety and efficacy of a procedure. The guidance does not cover whether or not the NHS should fund a procedure. Decisions about funding are taken by local NHS bodies (primary care trusts and hospital trusts) after considering the clinical effectiveness of the procedure and whether it represents value for money for the NHS.

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