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Horizon Scanning Technology Prioritising Summary

Serial transverse enteroplasty

**June 2008
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**Australian
Safety
and Efficacy
Register
of New
Interventional
Procedures -
Surgical**



**Royal Australasian
College of Surgeons**

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Enquiries about the content of the report should be directed to:

HealthPACT Secretariat
Department of Health and Ageing
MDP 106
GPO Box 9848
Canberra ACT 2606
AUSTRALIA

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This Horizon scanning prioritising summary was prepared by Mr Luis Zamora from the Australian Safety and Efficacy Register of New Interventional Procedures – Surgical (ASERNIP-S).

BACKGROUND

Short bowel syndrome (SBS) is a malabsorptive disorder caused by the surgical removal or dysfunction of a segment of the small intestine (DeLegge et al. 2007). The condition can be acquired or congenital in nature (DeLegge et al. 2007).

When SBS is acquired, the condition is usually a result of surgery in adults to treat conditions such as Crohn's disease, intestinal volvulus, small intestine tumours, injury/trauma to the small intestine, necrotising enterocolitis, or other surgeries performed to remove diseased or damaged portion(s) of the small intestine (Jackson and Buchman 2005). The congenital form of SBS presents in paediatric patients and may be a result of congenital intestinal atresia or conditions for which massive enterectomy is indicated (e.g. gastroschisis, necrotising enterocolitis) (Jackson and Buchman 2005).

Patients with SBS suffer from complications related to malabsorption and the subsequent deficiency of various minerals and vitamins (Jackson and Buchman 2005). As a result, many patients experience diarrhoea, steatorrhoea, abdominal pain, fluid retention, weight loss, fatigue, anaemia, hyperkeratosis, muscle spasms, poor blood clotting, and bone pain (Sawyer 2006). SBS is also associated with serious long-term complications such as liver and biliary complications, nutrient deficiencies, fluid and electrolyte disturbances, bacterial overgrowth, hyperoxaluria and D-lactic acidosis (DeLegge 2007).

In response to its compromised absorptive ability, the small intestine of the affected patient undergoes intestinal adaptation, an endogenous mechanism to compensate for its reduced length (Sawyer 2006). During intestinal adaptation, the remaining portion of the small intestine undergoes physiological changes to adapt to the reduced length and increase its absorptive ability including enlargement and lengthening of the villi, increase in the diameter of the small intestine and slow down in peristalsis or movement of food through the small intestine (Jackson and Buchman 2005). The process of intestinal adaptation may take weeks or months to occur (Cuffari 2006). Unfortunately, despite intestinal adaptation many patients still experience the symptoms of SBS.

Currently there is no cure for SBS (Sawyer 2006). Because the underlying causes of morbidity in SBS are the compromised digestive and absorptive processes of the small intestine management of the condition is focused on providing adequate nutrition (Cuffari 2006). Current non-surgical treatment of SBS includes using pharmacological agents to improve nutrition and relieve symptoms associated the condition, such as anti-diarrhoeal medicine, vitamin and mineral supplements, H2 blockers, proton pump inhibitors and lactase supplements. In patients in whom proper nutrition cannot be restored via pharmacological means, parenteral nutrition may also be administered (DeLegge et al. 2007). However, the use of parenteral nutrition is associated with substantial risk of liver and biliary complications, catheter related infections and catheter occlusion among others (Vanderhoof 1997). In patients in whom serious life-threatening complications arise, surgical treatment (specifically, intestinal transplantation) may be required (DeLegge et al. 2007).

Surgical management of SBS falls into two categories; intestinal or combined liver-intestinal transplantation and non-transplant operations. Intestinal or combined liver-intestinal transplantations are normally reserved for patients with serious potentially life threatening complications. Unfortunately transplantation has been associated with mixed results and is associated with a high risk for infection, rejection, and other complications related to immunosuppression (Sudan et al. 2007). Transplantation has been reported to have early postoperative mortality rates of up to 30% (Sawyer 2006).

Non-transplant operations include the creation of artificial enteric valves, stricturoplasty and intestinal tapering procedures (Sawyer 2006). Procedures to lengthen the dilated bowel resulting from intestinal adaptation such as the Bianchi procedure (a procedure where the bowel is cut in half and one end is attached to the other) and serial transverse enteroplasty (STEP) are also included in this category (Sawyer 2006).

The STEP procedure is performed to eliminate adaptive dilation by elongating the shortened bowel and preserving the mucosal surface area taking advantage of the natural adaptive dilation of the small intestine during SBS (Modi et al. 2007). The procedure is performed in patients who are dependent on parenteral nutrition and are experiencing SBS complications or as an alternative to tapering enteroplasty. During STEP, a surgical stapler is applied in a serial transverse manner to incompletely staple and divide the intestine (Modi et al. 2007). The stapler is applied from alternating sides of the small intestine, resulting in a new intestinal channel able to direct enteric contents through a longer and narrower lumen (Modi et al. 2007). It has been suggested that the procedure is less technically demanding than other lengthening procedures such as the Bianchi procedure (Sudan et al. 2007).

CLINICAL NEED AND BURDEN OF DISEASE

According to the literature, it is difficult to obtain the true incidence and prevalence of SBS (DeLegge et al. 2007). A report published in 1990 estimated the incidence of SBS requiring parenteral nutrition in the United Kingdom to be two patients per million (Lennard-Jones 1990). Results from a European survey published in 1999 have reported the incidence of home parenteral nutrition for which SBS was the most prevalent indication to be approximately four patients per million (Bakker 1999).

Unfortunately the number of patients reported in the studies above does not include patients who may have SBS and who did not require parenteral nutrition or in whom parenteral nutrition could not be successfully initiated thus underestimating the number of SBS patients (DeLegge et al. 2007).

The incidence of SBS in the paediatric population ranges from 24.5 per 100,000 live births to 353.7 per 100,000 live births in premature neonates (Wales et al. 2004).

DIFFUSION

Currently STEP is in the investigational stage. Due to the relatively small numbers of SBS sufferers, published reports of the STEP procedure remain limited. According to

Modi et al. (2007) it is unlikely that a prospective, randomised, controlled trial of the STEP procedure will ever be performed, due to the limited and variable nature of the SBS patient population.

COMPARATORS

The comparator to the STEP procedure in patients suffering from SBS is the Bianchi procedure.

SAFETY AND EFFECTIVENESS ISSUES

The literature on the STEP procedure was limited. Three studies, one comparative retrospective review and two case series studies were selected for inclusion in this summary.

Modi and colleagues (2007) presented the initial results from the International STEP Data Registry, which enrolled 38 patients (median age 1.3 years, range: 0 to 19.9 years) from 19 centres in three countries. The indication of the patients enrolled included SBS with dependence on parenteral nutrition (n = 29), bacterial overgrowth in the setting of SBS (n = 6) and neonatal atresia with marginal residual bowel length (n = 3). The primary diagnosis in these patients was varied and mostly consisted of intestinal atresia (n = 13), gastroschisis ± volvulus (n = 11) and necrotising enterocolitis (n = 7).

A retrospective review comparing the outcome of the STEP procedure and the Bianchi procedure was reported by Sudan et al. (2007). Forty-three Bianchi and 34 STEP procedures were performed in 64 patients (including 14 adults and three patients with prior isolated liver transplants). The patients selected for study included those with dilated small bowel loops on endoscopy, or radiologic imaging studies (preferably ≥ 4 cm in diameter) and remained dependent on parenteral nutrition with poor enteral progression or adaptation. The patients also included those with indications for intestinal transplantation (number not stated) and two patients not dependent on parenteral nutrition (these two underwent Bianchi lengthening). Of the 64 patients included, 43 patients underwent the Bianchi procedure while 21 underwent the STEP procedure.

Wales and colleagues conducted a case series study of 14 patients who underwent STEP and assessed both clinical and biochemical outcomes at one, six and 12 months. The patients included 11 males and 3 females with a mean age of 24.8 months. Necrotising enterocolitis (n = 4) and atresia (n = 7) were the most common diagnosis while gastroschisis (n = 1), Hirschsprung disease (n = 1) and volvulus (n = 1) were also reported. The indications for STEP in this study were parenteral nutrition associated cholestasis (n = 6) and bacterial overgrowth (n = 7).

a) Safety

In the results obtained from the International STEP Data Registry, intraoperative complications resulting from the STEP procedure included a leak at the apex of a staple

line, which was successfully repaired (n =2) and aspiration of the gastric contents on induction of anaesthesia (n =1) (Modi et al. 2007). The patient who suffered aspiration of gastric contents developed respiratory insufficiency and required a prolonged course of intensive care. The patient subsequently developed progressive liver failure and underwent a multivisceral transplant.

Postoperatively (within 30 days), five complications were reported as a result of the STEP procedure. These included bowel obstruction in two patients both of whom recovered successfully from the complication. Hypertension of unknown aetiology was reported in one patient who was being maintained pharmacologically. In the remaining patients, the postoperative complications included the development of intraabdominal abscess (n = 1), intraabdominal hematoma (n = 1) and serious pleural effusion (n =1). Each of these three complications was successfully managed using temporary, radiologically placed drainage catheters.

Late STEP complications (more than 30 days postoperatively) included the requirement for intestinal or multivisceral transplantation (n = 3) and death (n = 3). Multivisceral transplantation was performed in one patient while the remaining two received combined liver and intestine transplantation. The three transplantation patients all experienced progressive liver failure. An additional two patients, one experiencing ongoing feeding intolerance and the other experiencing progressive liver failure were referred for combined liver and intestine transplantation. Progressive liver failure and sepsis were reported as the cause of death in three patients (one neonate and two SBS patients with dependence on parenteral nutrition).

Sudan et al. (2007) reported six deaths, with an overall patient survival of 91% and no significant difference between the Bianchi and STEP groups. Five deaths occurred as a result of sepsis in children, two before the availability of intestinal transplantation, two in infants and one in a child with severe rejection three years after isolated intestinal transplant (4.3 years after lengthening). The sixth death occurred in an adult who experienced liver failure and sepsis after refusing intestinal transplantation.

Ten percent of patients experienced early major postoperative complications. There was no significant difference between the two types of lengthening procedures. Anastomotic leak (n = 1), intestinal obstruction (n = 1) necrosis of one of the two loops of bowel due to vascular injury (n = 1) and pneumonia (n = 1) were the early major postoperative complications following the Bianchi procedure. The first three patients underwent early reoperation while the pneumonia patient died two months after surgery. Following the STEP procedure, three patients developed high-grade obstruction. Two of these resolved after a period of observation without surgical repair. The third patient required surgical repair three months after the procedure but discontinued parenteral nutrition. Most patients (number not reported) experienced at least one episode of infection following surgery and many had repeated episodes until the central venous catheter was removed. Although infections were generally not life threatening, many patients required hospitalisation.

Late complications after the Bianchi procedure included interloop fistula (n = 3) and anastomotic stricture (n = 2, in patients who had discontinued parenteral nutrition for more than one year), all of which were successfully closed. There were 14 cases of recurrent bowel dilation (without fistula formation) which was treated with repeat lengthening (re-STEP) in 13 patients (including eight who had initially undergone the Bianchi procedure).

In the case series by Wales and colleagues (2007) three deaths were reported. Two deaths occurred at three months following the STEP procedure. One patient died as a result of sepsis while the other died as a result of complex congenital heart disease and sepsis. The third death, in a newborn patient followed a significant leak from the staple line which required emergency repeat laparotomy on the fifth postoperative day. The patient died as a result of a complicated course, liver failure and sepsis at 7.5 months.

Significant gastrointestinal haemorrhage refractory to medical management was reported in a 14 year old patient eight months after surgery. The patient underwent repeat laparotomy, and seven ulcers were discovered along the staple line. Unfortunately, after improvement, at 22 months postoperatively the patient developed repeat intestinal bleeding which at the time of publishing of the report was under investigation. Two more patients required emergency laparotomy as a result of a leak along the staple line. Both of these patients however ultimately died (already reported) from a prolonged and complicated course.

b) Effectiveness

In the Modi et al. (2007) report, the median follow-up period, from operation to the most recent update, death or transplantation was 12.6 months (range: 0 to 66.9 months). The pre-STEP mean intestinal length in patients with operative measurements was 68 ± 44 cm (range: 12 cm to 190 cm). This improved significantly following the STEP procedure to 115 ± 87 cm (range: 18 cm to 325 cm) (n = 27, p < 0.0001).

The pre-STEP mean intestinal width also decreased following the STEP procedure from 6.3 ± 3.9 cm (range: 2 cm to 24 cm) to 2.1 ± 0.9 cm (range: 1 cm to 5 cm), a significant improvement (n = 30, p < 0.0001).

Patients dependent on parenteral nutrition (excluding neonates and patients who progressed to transplantation or death) experienced an improvement in the percentage of total calories tolerated enterally from $31\% \pm 31\%$ to $67\% \pm 37\%$ (n = 21, p < 0.01). Of the 21 patients included, 10 were reported as completely weaned from parenteral nutrition at the time of publication of the report.

In the patients with bacterial overgrowth, five (out of six) experienced complete resolution of their symptoms. The patient who did not experience complete resolution had neurogenic anorectal incontinence and continued to experience diarrhoea at follow-up. Despite this, the patient had improved quality of life and improved bowel control. Of the three neonates that underwent STEP, two were reported to tolerate 100% and 80% of

their calories enterally. The third developed progressive liver failure, and was referred for liver and small intestine transplantation. However, the family elected to not pursue the transplantation and the patient died (reported in the safety section).

Three patients, previously listed for liver and intestine transplantation were removed from the transplant list as a result of steady improvement in their enteral tolerance following STEP.

Sudan et al. (2007) reported that the baseline median length of remnant small bowel in patients who underwent the Bianchi and STEP procedures was 44 cm and 45 cm respectively. Following the procedures, the median length of the small bowel increased in both the Bianchi and STEP recipients to 68 cm and 65 cm respectively ($p = \text{NS}$). When the percentage increase in length over the original length was compared between groups, it was revealed that the STEP procedure conferred a statistically significant greater increase ($p = 0.01$) than the Bianchi procedure (52% versus 48%).

At the time of surgery 62 out of 64 patients were dependent on parenteral nutrition (the number of patients dependent on parenteral nutrition in each group was not reported).

The median percentage of calories from parenteral nutrition was similar between the Bianchi and STEP groups before surgery (79% and 90% respectively, $p = 0.84$). At the three month follow-up, the median percentage of calories from parenteral nutrition had improved in both the Bianchi and STEP groups to 50% and 35% respectively. Patients who had received the STEP procedures experienced a statistically significant greater improvement in median parenteral nutrition compared to patients who underwent the Bianchi procedure ($p = 0.05$). Whilst the improvement in parenteral nutrition continued at the six month follow-up to 30% and 7% in the Bianchi and STEP recipients respectively, there was no difference between the groups ($p = 0.26$).

There were 62 parenteral nutrition dependent patients in the study, 58% of which were weaned following surgical lengthening. No statistically significant difference based on lengthening procedure was reported (Bianchi 55% versus STEP 60%). While there appeared to be faster weaning following STEP (4.8 months versus 8.4 months in the Bianchi patients) the difference was not statistically significant ($p = 0.07$).

Resolution of jaundice was achieved in 83% of patients (difference between the groups was not stated). There were two deaths and two intestinal transplantations among those who did not clear their jaundice.

Intestinal transplantation was required in nine patients at a median of 2.9 years after the lengthening procedures. Transplantation was significantly higher in patients who underwent the Bianchi procedure (18.6%) than patients who underwent STEP (5%) ($p = 0.03$). The indications for transplantation included loss of venous access ($n = 2$), jaundice ($n = 4$), and recurrent line-related septicaemia ($n = 3$).

Severe growth retardation was common among the 49 children in the study population, with the median baseline Z score for height and weight being -1.74 (range: -5.0 to 0.6)

and -1.25 (range: -7.29 to 1.52) respectively. Following the lengthening procedures, catch-up growth was reported in a large number of children (number of children and extent of catch-up growth not reported). No difference based on the type of lengthening procedure was reported.

Failure to wean from parenteral nutrition, age less than one year at the time of lengthening and extensive bridging fibrosis or cirrhosis were identified as significant factors associated with mortality.

Three variables were identified that differed between patients who were weaned from parenteral nutrition and those that were not. The final length of the small intestine after surgical lengthening was longer ($p = 0.03$) in patients who were weaned (75 cm, range: 23 cm - 150 cm versus 52 cm range: 20 cm – 160 cm). Secondly small intestine length gained was greater ($p = 0.005$) in patients who were weaned (25 cm: range: 7 cm – 92 cm versus 15 cm range: 5-35cm). Finally, the percentage of calories taken enterally at the time of surgical lengthening was higher in weaned patients (p value not reported).

The need for intestinal transplantation after surgical lengthening was associated with five variables. The first variable was the type of lengthening procedure (Bianchi 18.3% versus STEP 5%, $p = 0.001$). The second variable was length of remnant intestine before lengthening which was shorter in patients who required intestinal transplantation (29 cm versus 50 cm, $p = 0.04$). The third was the length of remnant intestine after surgery which was greater in those who did not require transplantation (70 cm versus 50 cm, $p = 0.03$). Fourth, the indication for undergoing lengthening was more frequently jaundice (6/9; 67%) in those that required transplantation ($p = 0.05$). Finally, the percentage of enteral calories at six months after lengthening was lower in patients who later required intestinal transplant (42% versus 89%, $p = 0.05$)

In the study reported by Wales et al. (2007), small intestine length increased from 107 ± 48 cm before STEP to 146 ± 58 cm after STEP, equating to a mean increase of the dilated bowel segment of $94\% \pm 30\%$ and increase in total small intestine length of $49 \pm 42\%$ (no p value reported). The mean diameter of the intestine also improved, from 6 ± 1 cm to 2 cm after STEP.

Data was also analysed as two separate groups, infants and older patients ($n = 11$) and newborn patients ($n = 3$). When the infants and older patients were analysed it was revealed that there was a significant increase in weight (from 14.5 ± 13.7 kg to 20.4 ± 14.9 kg) and decrease in the percentage of parenteral nutrition support (from $71\% \pm 21\%$ to $12\% \pm 24\%$) at 12 months compared to baseline ($p < 0.01$, $n = 6$). Stool frequency per day also improved in these patients from 8 ± 4 per day to 2 ± 1 per day at 12 months ($p < 0.05$, $n = 6$). Of the biochemical outcomes measured (citrulline, D-xylose, Alpha-1 AT and fecal fat percentage) only citrulline significantly changed from 17 ± 9 $\mu\text{mol/L}$ at baseline to 33 ± 7 $\mu\text{mol/L}$ at 12 months ($p < 0.05$, $n = 6$). It should be noted that only six patients were included in these analysis. The other five patients were excluded from

analysis because of intestinal liver transplantation in two, death of two other patients (already reported) and insufficient data in the fifth patient.

In the newborn patients (n = 3) the mean intestinal length improved from 115 ± 31 cm at baseline to 148 ± 44 cm post-STEP (no p-value reported).

COST IMPACT

The cost of the entire STEP procedure was not revealed in the searches conducted. However, Wales et al. (2007) reported that during their study the mean number of cartridges used in 2006 was 16 ± 9, resulting in total stapler costs of Can \$2878.51 ± 1406.22.

ETHICAL, CULTURAL OR RELIGIOUS CONSIDERATIONS

No issues were identified from the retrieved material.

OTHER ISSUES

No issues were identified from the retrieved material.

SUMMARY OF FINDINGS

Serial transverse enteroplasty is a recently developed method of intestinal lengthening for patients suffering symptoms of short bowel syndrome. Despite the limited evidence the procedure has demonstrated efficacy in improving the clinical status of recipients. Further studies involving larger patient numbers and using comparative methodology are required to more accurately determine its safety and effectiveness as a surgical lengthening procedure for patients with short bowel syndrome.

HEALTHPACT ACTION

Based on the limited evidence regarding the STEP procedure suggesting its effectiveness and the small population of patients with SBS, this technique will be monitored for 24 months.

NUMBER OF STUDIES INCLUDED

Total number of studies	3
Level III-3 intervention evidence	1
Level IV intervention evidence	2

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SEARCH CRITERIA TO BE USED

STEP

Serial transverse enteroplasty

Intestinal lengthening

Short bowel syndrome

SBS

PRIORITISING SUMMARY (2010 UPDATE)

NAME OF TECHNOLOGY

SERIAL TRANSVERSE ENTEROPLASTY

PURPOSE AND TARGET GROUP

TO LENGTHEN THE SMALL INTESTINE IN PATIENTS WITH SHORT BOWEL SYNDROME

2010 SAFETY AND EFFECTIVENESS

Study descriptions

An updated literature search identified four studies using the STEP procedure, three of which were case series studies and one of which was a case report. Two case series studies were eligible for inclusion in this update (Ching et al 2009; Andres et al 2008).

Ching et al (2009) aimed to assess clinical and nutritional outcomes in 16 patients (10 male, 6 female) undergoing the STEP procedure. These patients' chart and medical records were retrospectively reviewed with a mean follow-up period of 23 months (range, 1-71 months). The mean age of the patients at the time of the STEP procedure was 12 months (interquartile range, 1.5-65.0 months). Primary diagnoses included gastroschisis (n=6), intestinal atresia (n=4), midgut volvulus (n=3), necrotising enterocolitis (n=2), and Hirschprung's disease (n=1), and indications for STEP included failure to advance enteral feedings (n=11), bacterial overgrowth (n=3), or neonatal atresia (n=2). Using a linear mixed model with a random intercept the authors estimated mean postoperative trajectories (as a function of time) of the 16 patients' z scores for weight, height, weight-for-height, and percentage of enteral calories. These model-estimates were then compared with mean overall available measures obtained in specified intervals before starting the analysis. One patient who underwent STEP for bacterial overgrowth and who was receiving full enteral nutrition at the time of surgery was excluded from enteral tolerance analyses.

The study by Andres et al (2008) presented outcomes in 14 patients (7 male, 7 female) with intestinal failure due to short bowel syndrome who underwent a repeat STEP (re-STEP) procedure (16 procedures in total) following prior lengthening with either STEP (n=7) or Bianchi (n=7). Charts, operative records, and available clinical information were retrospectively reviewed in these patients. In patients who underwent more than one reSTEP procedure (n=2) results were taken from their last surgery. Patients were selected for the reSTEP procedure using the same principles as the initial procedure. The overall median time from the initial lengthening procedure to reSTEP was 12 months (range, 2 months-15 years).

Twelve patients were paediatric at the time of the initial procedure (median 3.5 months, range 2-35 months) and subsequent procedure(s) (median 19.5 months, range 10-71 months). In these patients, underlying disease was proximal intestinal atresia (n=8), gastroschisis (n=4), or omphalocele (n=1). Only one patient had the ileocecal valve at the first presentation. Two paediatric patients underwent three lengthening procedures each

(one Bianchi, two STEP and three STEP, respectively). One of the two adult patients undergoing reSTEP had short bowel syndrome due to gastroschisis and volvulus with intestinal failure and total parenteral nutrition dependence since birth and the other had malrotation and volvulus at the age of 5 years. Both adults underwent previous Bianchi at the ages of 4 and 11 years, and their reSTEP procedures at the age of 18 and 30 years, respectively. Ileocecal valve was not present in either adult patient. Additional interventions performed at the time of reSTEP surgery included stricturoplasties (n=3), simple bowel taperings (n=2), liver biopsies (n=4), and cholecystectomy (n=1). Median follow-up duration was 14.5 months (range, 1 month-4.6 years).

Safety

Long-term complications in the patient population reported by Ching et al (2009) included catheter-related bacteremias in 31% (5/16) of patients, gastrointestinal bleeding in 13% (2/16) of patients, and small bowel obstruction in 7% (1/15) of patients. Two patients underwent subsequent transplantation (1 multivisceral and 1 liver small bowel) for complications of progressive liver disease.

In the study by Andres et al (2008) there were no intraoperative complications encountered. Late surgical complications occurred in 28% of patients and included stricture (n=2), recurrent bowel obstruction (n=1), and central line infection (n=6). The two cases of stricture were diagnosed by repeat contrast studies performed because of emesis; stricturoplasty was performed at 6 and 24 months following reSTEP. The case of recurrent small bowel obstruction was due to a floppy dilated duodenum and was reoperated on 5 months postoperative. Two of the patients with central line infection were progressively weaned off parenteral nutrition (which significantly reduced admissions) and the remaining patients underwent intestinal transplantation. Overall morbidity (including the need for surgery or hospital admission for medical complications) occurred in 64% of patients.

Effectiveness

In the study by Ching et al (2009) there was a mean increase in small bowel length of 91% (standard deviation, 38%) so that bowel length increased from a mean of 60cm to 117cm post-STEP procedure (P value not given). Small bowel width decreased by a mean 70% (standard deviation 6%) from 6.5cm to 2.0cm post-STEP procedure (P value not given). Both weight-for-age and height-for-age z scores improved significantly per year, by 0.36 units and 0.24 units respectively (P=0.0001; P=0.004), as did weight-for-height for age z scores (0.48 units per year; P=0.02). Enteral tolerance also experienced a significant improvement of 1.4% per month (P<0.0001); with a trend in improvement seen up to 48 months postoperative. Thirty-eight percent (6/16) of patients transitioned off parenteral nutrition, achieving complete enteral autonomy, and remained off throughout follow-up. The study also reported no patient deaths.

In the study by Andres et al (2008) there was a 2.6-fold increase in median remnant bowel length from before the initial lengthening procedure to after the final reSTEP procedure. Median bowel length increased from 35cm before initial lengthening to 56.5cm after the initial lengthening, to 90cm after the final lengthening procedure (P

value not given). Patient survival was 100% at the end of follow-up. Total discontinuation of parenteral nutrition was achieved in 43% (6/14) of patients; three of these patients had been weaned off parenteral nutrition prior to reSTEP but restarted due to dehydration, intestinal obstruction, or bacterial overgrowth. They then managed to achieve discontinuation again following the reSTEP procedure. Therefore, 29% (4/14) of patients remained on parenteral nutrition at the end of follow-up. Whether the patient had undergone Bianchi or STEP as their previous lengthening procedure did not appear to affect their weaning ability (however patient numbers may be too small to detect such a difference). However it may be that final bowel length < 70cm affects a patient's ability to discontinue parenteral nutrition, as only 1/6 patients able to do this had a bowel length <70cm.

Intestinal transplantation salvage took place in four patients at 7, 9, 13 months, and 4.6 years post-reSTEP; one patient underwent adjuvant liver transplantation. Indications for intestinal transplantation included the persistent need for parenteral nutrition and multiple life-threatening infections. Seventy-five percent (3/4) of transplant recipients were able to discontinue parenteral nutrition. The average percentage of total calories tolerated enterally increased by 67% (relative to the initial evaluation – excluding patients who progressed to transplantation) (P value not given).

2010 SUMMARY OF FINDINGS

This update found a relatively low rate of complication and no patient deaths were associated with the primary STEP procedure. These observations come from a study with up to 5 years follow-up; however, additional long-term studies are needed in order to support these findings. It was also noted that with proper patient selection and ongoing intensive intestinal rehabilitation primary STEP may provide patients with improved enteral tolerance and independence from parenteral nutrition. The evidence base for primary STEP in patients with short bowel syndrome is still lacking in high-quality comparative evidence.

Previously the repeat STEP procedure has only been reported in animals. Results from the included study found reSTEP to be technically feasible and able to assist in discontinuation from parenteral nutrition in considerable proportion of patients. In addition reSTEP did not preclude subsequent intestinal transplantation. Until further literature regarding STEP, both as a primary and secondary procedure, is available it will be difficult to determine its safety and efficacy.

2010 HEALTHPACT ACTION

Based on the paucity of literature available (particularly the lack of comparative evidence), HealthPACT has noted the development of this technology. However, additional updates are not required at this time.

2010 INCLUDED STUDIES

Total number of studies	2
Level IV intervention evidence	2

2010 REFERENCES

Andres AM, Thompson J, Grant W, Botha J, Sunderman B, Antonson D, Langnas A, Sudan D. Repeat surgical bowel lengthening with the STEP procedure. *Transplantation* 2008; **85**(9): 1294-1299.

Ching YA, Fitzgibbons S, Valim C, Zhou J, Duggan C, Jaksic T, Kim HB. Long-term nutritional and clinical outcomes after serial transverse enteroplasty at a single institution. *Journal of Pediatric Surgery* 2009; **44**(5): 939-943.

2010 SOURCES OF FURTHER INFORMATION

Morikawa N, Kuroda T, Kitano Y, Tanaka H, Takayasu H, Fujino A, Shibata Y, Tanemura H, Muto M, Honna T. Repeat STEP procedure to establish enteral nutrition in an infant with short bowel syndrome. *Pediatric Surgery International* 2009; **25**(11): 1007-1011.

Sigalet D, Boctor D, Robertson M, Lam V, Brindle M, Sarkhosh K, Driedger L, Sajedi M. Improved outcomes in paediatric intestinal failure with aggressive prevention of liver disease. *European Journal of Pediatric Surgery* 2009; **19**(6): 348-353