

## STEP Procedure (Serial Transverse Enteroplasty Procedure) in Uruguay

Berazategui Bernardo\*

*Hospital Pereira Rossell, Montevideo, Uruguay*

\***Corresponding Author:** Berazategui Bernardo, Hospital Pereira Rossell, Montevideo, Uruguay.

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### Abstract

**Introduction:** The treatment of Short Bowel Syndrome (SIC) in pediatric patients is a great challenge due to its complexity and individual variability. The treatment of SIC requires a multidisciplinary team, which is based on 3 basic pillars: nutritional support, intestinal rehabilitation and transplantation. The concept of intestinal rehabilitation refers to those measures designed to promote the adaptation of the residual intestine, achieving the suspension or reduction of PN needs. Among the therapeutic alternatives, we highlight the STEP procedure as an alternative for intestinal lengthening.

**Objective:** To disseminate the STEP procedure as a proven alternative for the treatment of SBS and as a strategy to take into account for any situation that determines a pathological dilation of the small intestine. In addition, this publication is interested in the dissemination of the disease as an increasing entity due to advances in the field of neonatology and surgical alternatives for its resolution.

**Clinical Case:** This is a preterm NB with a prenatal diagnosis of Gastroschisis that at the time of reception, an almost total ischemic involvement of the small intestine and colon is verified, determining an intestinal insufficiency due to a Short Bowel Syndrome. The evolution of intestinal pathology is described from resection, through intestinal adaptation within which the STEP technique stands out as one of the treatment alternatives. We highlight the procedure and the results obtained with it.

**Theoretical Framework:** We mention the transcendental theoretical concepts for the understanding of the intestinal adaptation process and the treatment alternatives.

**Discussion:** This section compares the different surgical techniques for the treatment of Short Bowel Syndrome. Among the surgical alternatives for intestinal elongation, the Bianchi procedure and the STEP stand out. We carried out a bibliographic search to analyze each particular technique with its results and finally the comparative studies between the different elongation techniques.

**Conclusion:** SIC is a pathology with a tendency to increase the incidence due to the improvement of neonatal units and nutritional supports, therefore, in addition to knowing the disease, we must acquire therapeutics for its management. It is our desire to communicate the technique that we have carried out, to inform about its benefits and the therapeutic alternatives in the surgical arsenal of short bowel syndrome.

**Keywords:** *Short Bowel Syndrome (SIC); STEP (Serial Transverse Entero-plasty); Uruguay*

### Introduction

The treatment of Short Bowel Syndrome (SIC) in pediatric patients is a great challenge due to its complexity and individual variability. Require the integration of medical, nutritional, surgical and psychological disciplines, and treatment should be based on the patient's age, remaining intestinal anatomy, nutritional status, and underlying disease, as well as the many complications that may appear. The

fundamental pillar is undoubtedly nutritional therapy and it has been shown over the last few years that it is one of the factors that has drastically modified the survival of these patients.

The treatment of SIC requires a multidisciplinary team, which is based on 3 basic pillars: nutritional support, intestinal rehabilitation and transplantation. The concept of intestinal rehabilitation refers to those measures designed to promote the adaptation of the residual intestine, achieving the suspension or reduction of PN needs. It consists of the combination of medical-surgical therapeutics in order to preserve the functionality with autologous intestine.

Within surgical therapeutics, the intestinal lengthening technique called STEP (Serial transverse Entero-plasty), described by Kim in 2003 [1], is developed as an alternative to other lengthening techniques and especially to the one developed by Bianchi in 1980 [2], which was until then. moment the reference technique for pathology.

### Goals of the Study

We analyze the case of a patient with SIC in which the first STEP procedure was performed in our country. Of interest in this publication is the dissemination of pathology as an increasing entity due to advances in the field of neonatology and surgical alternatives for its resolution.

### Clinical Case

Mother 16 years old, from Salto, primipar. Complicated pregnancy with untreated lower genital infection.

Prenatal ultrasound diagnosis of Gastroschisis at 18 SEC. Enter a high-risk unit. 24 weeks ultrasound control, non-dilated loops. Normal echokaryogram.

She was admitted for painful uterine contractions in January/2014, normal infectious paraclinic. RPM, clear liquid. Echo 32 weeks breech anhydramnios. Complete lung maturation. Cesarean section was decided based on MPR, CUD, and position.

RN, male, EG 32 - 33, weight 1680 AEG, Apgar 8/9, joint reception with surgeon. A small parietal gap with most of the intestinal loops and necrotic sigmoids stands out. Silo is made.

In surgery at 12 o'clock, extensive necrosis of the jejunum and ileum and the colon up to the middle transverse, resection of these sectors and proximal jejunal ostomy and transverse colon stands out. The first intestinal measurement refers to 16 cm from duodenum 2 to the stoma.

In a second surgery, 8 days after the first with the encouragement of intestinal reconstruction due to the unmanageable intestinal losses due to the proximity of the ostomy, a new measurement of the pylorus is made until the ostomy that refers 40 cm. We emphasize that both measurements have been made incorrectly, since they must be done from the duodenum jejunal angle, which is why the data previously provided lack veracity. In this surgery, given the great incongruity of ends due to the dilation of the jejunum and the defunctionalized small colon, the former is remodeled and the transit is reconstructed by means of an end-to-end anastomosis.

In the postoperative period, the patient presents a good evolution from the surgical point of view, without complications and persists with parenteral feeding, later the enteral supply begins to install, reaching 3 cc/hour. The result at the beginning is positive, Prokinetics must be added due to residuals greater than normal, but in the course of the evolution the patient deteriorates despite the efforts to enable the enteral route that force to suspend it and again spend to TPN Contrast Duodenal Gastro Study, highlights: stomach of usual morphology without signs of parietal lesion and normal evacuation. Duodenal arch with anomalous arrangement compatible with mal-rotation. Clear increase in the diameter of the duodenum and jejunum, with preserved peristalsis. Gastric duodenum reflux is observed several times during the study. At 20 min, minimal passage of contrast from jejunum to colon through the filiform canal is detected.

Below is a table 1 that summarizes the patient’s humoral assessment. The sustained increase in bilirubin and liver transaminases added to weight stagnation and the impossibility of introducing EN constitute the indication for surgical treatment. It shows the liver involvement associated with prolonged parenteral nutrition (complication of SIC). It was decided to perform surgical treatment of intestinal elongation. This aims to improve the absorption surface and thus try to decrease PN and progressively improve tolerance of NE. This mechanism called intestinal adaptation constitutes one of the means to reduce the complication associated with prolonged parenteral nutrition for which surgery was proposed (Figure 1).

Hepatogram	26/2	8/3	30/3	15/4
Total protections	3.95	4.70	4.59	4.26
Albumin	2.76	3.80	3.85	3.24
BT	3.12	4.69	4.60	4.24
BD	2.86	4.28	3.69	3.73
BI	0.26	0.41	0.90	0.51
TGO	47	58	136	283
TGP	45	70	125	441
GLOB	1.19	0.90	2.14	1.02
FA	518	811	733	882
GGT	304	256	256	147
Colinest	3883	3501	4800	4132
LDH	787	590	708	637
Col total	120	123		125

Table 1

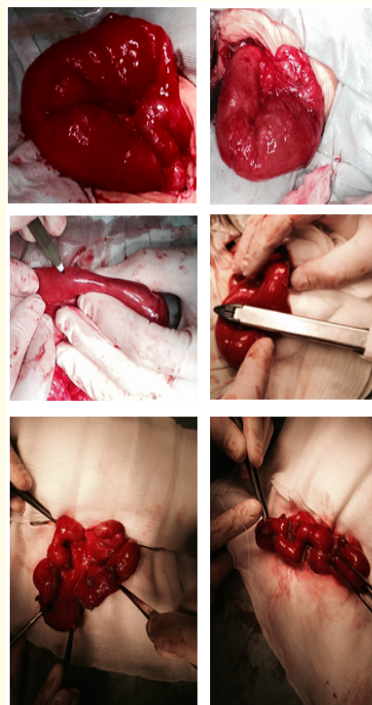


Figure 1: Paraclinical humoral.

### Of the surgery stands out (STEP)

Distended handles 4.5 cm in transverse diameter. Jejunum/colon caliber difference 4 cm/1.5 cm, good passage. Pre STEP intestinal length 31 cm.

8 shots with MLC alternating at 90 and 270 degrees from the meso, each separated by 2 cm.

Small intestine tube preparation of 1.5 cm in diameter.

Post STEP intestinal length 67 cm.

### Theoretical framework

SIC is defined as intestinal resection that affects intestinal autonomy and requires supplementation to meet nutritional demand. This determines an intestinal failure, which can be temporary or permanent. The incidence is unknown, some series estimate 24.5/100,000 live births in Canada or 2 - 3/1,000,000 in the United Kingdom, half of which are children.

The intestinal length in the full-term new born is generally between 2.5 - 3 meters. It has not been possible to establish exactly what is the length of the intestinal remnant that defines the SIC, however as a general rule it is accepted that it is the one smaller than 80 - 100 cm and the ultrashort smaller than 40 cm.

The main causes that determine SIC in the neonatal period (85% of the total occur in this stage) are: gastroschisis, intestinal atresia, midgut volvulus and necrotizing enterocolitis.

SIC passes from the initial noxa through 3 stages: the initial one, after intestinal resection, which is determined by the initial severity of the surgery and post-surgical management. Intestinal losses and their management are of great importance in this period. It is followed by the adaptation period, which can range from months to two years. It is a process that occurs as compensation after extensive intestinal resection, this determines structural and functional changes of the digestive system in order to improve the absorption of nutrients and fluids in the intestinal remnant. Finally, in case of intestinal failure despite the previous efforts, the only therapeutic alternative is intestinal transplantation. The determining factors that intervene in intestinal adaptation are in addition to the pathological history of the patient, nutritional status, anatomical factors of the resected sector, surgical treatments and complications of SIC. Among the prominent anatomical factors, the remaining intestinal length, the resected area of intestine (ileum replaces the functions of the jejunum but not the reverse), the presence of the ileus cecal valve (VIC), presence or absence of colon and if you have ostomies and their height. Other factors participating in intestinal adaptation are enteral nutrition, liver damage associated with PN and FI, bacterial overgrowth, and the management of venous access, all of which are considered complications of SIC.

### Discussion

As mentioned at the beginning of the article, rehabilitation is part of the therapeutics of SIC and includes those medical and surgical measures aimed at maintaining enteral autonomy, reducing the need for PN and avoiding complications from its prolonged use. Surgical alternatives for intestinal rehabilitation include: techniques for slowing down transit (not applicable in pediatric age), techniques that prevent bacterial overgrowth (enteroplasty, intestinal plication and correction of strictures, all aimed at avoiding major resections) and finally the techniques that seek intestinal lengthening.

Among the lengthening techniques, the Bianchi techniques or Longitudinal Intestinal Lengthening Technique (TAIL), Serial Transverse Enteroplasty (STEP) and Spiral Intestinal Lengthening and Tailoring (SILT) developed in 2014 as an alternative to the latter two stand out.

The point of discussion of this article focuses on 2 aspects of the treatment of the pathology, trying to resolve if the appropriate procedure has been carried out, for which our questions are: 1) if we choose the best method of intestinal lengthening and 2) if the results regarding intestinal length gain are comparable with the international registry, since it is the only variable that we can compare by the number of cases.

With regard to technique, of the three tested for the short bowel, SILT cannot yet be considered with respect to the results due to the lack of prospective and randomized data. The other two techniques, TAIL and STEP, are the subject of several scientific papers trying to find the differences in favor of an ideal technique.

One of the first comparative works on this topic was published in 2007 by the University of Nebraska. This is a retrospective study of a single center with a considerable number of 64 patients. They found no significant differences in survival between procedures or in the percentage of those who achieved PN independence. Patients operated with the STEP technique reach PN independence faster; this is a difference they find with respect to both techniques and they arrive at the explanation that this phenomenon is given because STEP can increase more than 100% of its initial longitudinal, while the Bianchi could only double its length at most.

A systematic review by Frongia, *et al.* in which 39 publications are reviewed, including the experience of a single center using the Bianchi technique for 20 years. It is concluded that there is no statistically significant difference between the two procedures regarding the gain in length of the intestine (approximately 70%), in the acquisition of EN and the reversal of the complications of PN. However, it ends by asserting that the STEP procedure reduces mortality and overall progression towards transplantation.

Today, the use of the STEP procedure is a recommendation for low-frequency centers, as it is a simpler and more reproducible technique than TAIL. Another widely accepted advantage of the former over the latter is the possibility of re-intervening the same intestinal segment in the event of a new symptomatic enteral dilation.

Regarding the results regarding the gain in cm of intestinal length in our patient it was 36% (31 cm pre-STEP, 67 cm post-STEP), which corresponds to a gain of 116%. Compared with the International Registry Center of STEP, it presents a pre-STEP measurement range of intestine that goes from 12 cm - 190 cm and post-STEP 18 cm - 325 cm, so the average gain goes from 68 + - 44 cm to 115 + - 87 cm. This ultimately results in an average increase of 69% for all STEP procedures. The rest of the variables analysed are not comparable in our case because it is a unique case [3-15].

### Conclusion

We are facing a pathology with a tendency to increase the incidence due to the improvement of neo-natal units and nutritional supports, so in addition to knowing the disease we must acquire therapeutics for its management. It is our desire to communicate the technique that we have carried out, to inform about its benefits and the therapeutic alternatives in the surgical arsenal of short bowel syndrome.

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