Intestinal lengthening: an experimental and clinical review

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Summary: Small intestinal lengthening by the Bianchi procedure has now had successful clinical application in children and neonates with the short-bowel syndrome. This paper reviews the background experimental work and clinical cases so far treated. A personal case of intestinal lengthening in a 7-week-old baby with 35 cm jejunum is described in detail.

Intestinal lengthening appears to reduce dependence on parenteral nutrition, thus allowing earlier establishment of total enteral alimentation. The procedure may therefore have a useful place in the overall management of the short-bowel syndrome.

Introduction

Children surviving the neonatal phase and the subsequent fluid, electrolyte and nutritional problems following massive small bowel loss, have excellent prospects for relatively normal lives (Rickham *et al.* 1977). Increasing expertise with total parenteral nutrition in neonates has led to a steady reduction in the minimum length of small bowel that is compatible with survival, so that the 30 cm proposed by Benson *et al.* (1967) has now been reduced to 20 cm (Lister & Rickham 1978). Indeed, survival with extremely short lengths of small bowel is becoming more common (Postuma *et al.* 1983, Kurz & Sauer 1983). Of special relevance when assessing such reports, however, is the long-term quality of life for these children, which may prove far from ideal. Furthermore, their potential not just for survival, but especially for an acceptable growth, requires careful assessment.

Tilson (1980) states that surgical procedures for difficult patients with short-bowel syndrome cannot be advised at present, firstly because the time course for adaptation is in the order of one year, and secondly because such procedures may result in further loss of precious bowel surface area. The argument for prudence and careful assessment in such cases cannot but be upheld. Equally, however, procedures which can be shown to enhance intestinal adaptation, reduce dependence on parenteral alimentation and establish earlier total enteral nutrition, thereby improving the long-term outlook, should not lightly be discarded. Indeed, they may have a useful role in the overall management of the short-bowel syndrome.

Aetiology of short gut

The majority of underlying conditions that lead to major loss of intestine in neonates and infants have their origins in intrauterine life. Antenatal vascular accidents resulting in intestinal infarction present as intestinal atresias, and occur also in association with gastroschisis. Volvulus of the midgut secondary to malfixation and malrotation frequently results in catastrophic bowel loss, and may arise *in utero* or at any time postnatally. Large resections of bowel are a regular feature of extensive necrotizing enterocolitis, a condition found increasingly frequently in most neonatal units. Other factors of lesser frequency include Hirschsprung's disease involving the small bowel, mesenteric vascular embolism or thrombosis secondary to hypercoagulable states, or clots or vegetations associated with cardiac lesions or aortic catheters. Inflammatory conditions such as Crohn's disease or radiation enteritis usually affect an older age group.

Pathophysiology

The essential defects giving rise to the short-bowel syndrome following massive loss of small bowel are a severe shortage of absorptive mucosa and a marked reduction in intestinal length. As a consequence, transit time is rapid and mucosal contact time much reduced, with resultant massive fluid and electrolyte losses and insufficient absorption of nutrients. Marked dilatation of the obstructed proximal bowel in intestinal atresia renders it peristaltically ineffective. Even after operative relief of the mechanical obstruction, a relative failure of propulsion leads to stasis in the dilated segment with consequent bacterial colonization. The

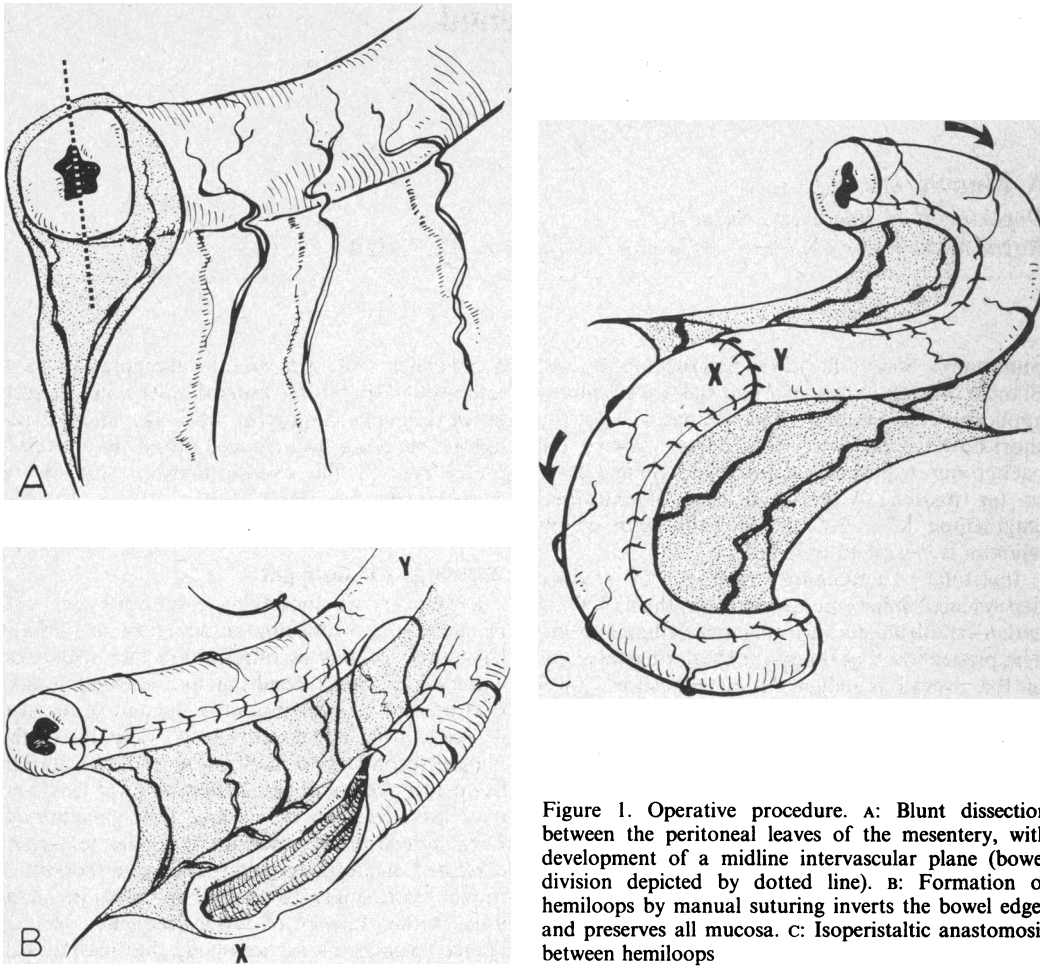

Figure 1. Operative procedure. A: Blunt dissection between the peritoneal leaves of the mesentery, with development of a midline intervascular plane (bowel division depicted by dotted line). B: Formation of hemiloops by manual suturing inverts the bowel edges and preserves all mucosa. C: Isoperistaltic anastomosis between hemiloops

development of the blind-loop syndrome is accompanied by atrophy of valuable potentially absorptive mucosa. Authoritative detailed descriptions of the pathophysiological changes and catastrophic effects on nutrition following massive small bowel loss are available in the literature (Wright & Tilson 1971, Tilson 1980) and are not dealt with further here.

Management

The mainstay of management for children with the short-bowel syndrome consists of a prolonged course of parenteral nutrition and dietary adjustments until a degree of intestinal adaptation compatible with life and sustained growth has been achieved. Surgical procedures have, for the most part, attempted to increase absorption by prolonging transit time. These have included vagotomy and pyloroplasty (Frederick & Craig 1964), recirculating small bowel loops (Budding & Smith 1967), reversed small bowel segments – single and paired (Hammer *et al.* 1959, Keller *et al.* 1965, Venables *et al.* 1966), formation of a pouch (Poth 1969) and prejejunal or preileal colon transposition (Hutcher & Salzberg 1971, Hutcher *et al.* 1973, Lloyd 1978, Garcia *et al.* 1981). Other investigators have attempted to grow small intestinal mucosa on colonic serosal patches (Binnington *et al.* 1973, 1974, 1975), or have used vascularized abdominal wall pedicle flaps (Lillemoe *et al.* 1982), mucosally denuded tubular colonic segments (Norton *et al.* 1975, Watson *et al.* 1980) and prosthetic materials (Watson *et al.* 1980, Harmon *et al.* 1979). Experimental models for small bowel transplantation have also been developed (Lillehei *et al.* 1967, Preston *et al.* 1966, Stauffer 1975, Stauffer *et al.* 1978), though clinical application is presently limited.

A reproducible technique for isoperistaltically increasing the length of the residual small bowel was first described in 1980 (Bianchi 1980). The procedure has the advantage of simultaneously tailoring the bowel and preserving all available mucosa for intestinal adaptation. Intestinal lengthening has since had successful clinical application in neonates and children with the short-bowel syndrome (Boeckman & Traylor 1981; also personal communications from H Bieman Othersen & W E Bomar 1981, and R J Brereton 1983).

This paper reports on the experimental background and describes further clinical experience with the intestinal lengthening procedure.

Operative procedure

A segment of small intestine is isolated on its vascular pedicle. Blunt dissection between the peritoneal leaves of the mesentry (Figure 1A) gives access to the mesenteric vessels, which are allocated alternately to one or other side of the bowel loop. Since the vessels do not enter the bowel wall directly in the midline but rather to one or other side, a relatively avascular space can be developed along the mesenteric border. Using bipolar diathermy the bowel is divided longitudinally, as depicted by the dotted line in Figure 1A, along its mesenteric and antemesenteric borders leaving equal amounts on either side. Formation of hemiloops (Figure 1B) by inverting Lembert sutures, allows inversion of the bowel edges and reduces the risk of damage to the blood supply and interloop fistulae. Alternatively, bowel division and stapling (Bianchi 1980) can be performed with the GIA Autosuture Stapling Instrument (United States Surgical Corporation, Stanford Conn 06902). The hemiloops are anastomosed (point X to point Y in Figure 1C) so that a vascularized isoperistaltic loop, which is double the length and half the diameter of the original segment, is constructed without loss of absorptive mucosa.

Experimental data

Large white Landrace-cross pigs were used for the study. In all cases intestinal lengthening was performed with the GIA Autosuture Stapling Instrument. The first phase of the study was designed to develop a reproducible operative technique for doubling the length of a segment of small bowel and simultaneously reducing its diameter without loss of absorptive mucosa. Of the four animals (weighing 15–20 kg) in the first phase of the study, two had 10 cm jejunal segments increased to 20 cm, one had a 20 cm ileal loop increased to 40 cm, and a fourth had a 30 cm jejunoileal segment increased to 60 cm. The lengthened segments were converted into self-emptying blind loops, the distal end opening as an ostomy on the anterior abdominal wall (Figure 2A). Two animals died at 48 hours and 8 days from sepsis and intestinal adhesions, and two survived to termination of the experiment 16 weeks later. Macroscopic and histological examination confirmed viability and luminal patency of all four lengthened segments.

The object of the second phase was to assess the response of lengthened small bowel segments to a short gut situation (Figure 2B). 20 cm jejunal loops of 1.5 cm diameter were increased in length to 40 cm by the Bianchi technique. A 75% mid small bowel resection was performed simultaneously to provoke intestinal adaptation. The lengthened bowel was anastomosed isoperistaltically between the duodenojejunal flexure at the

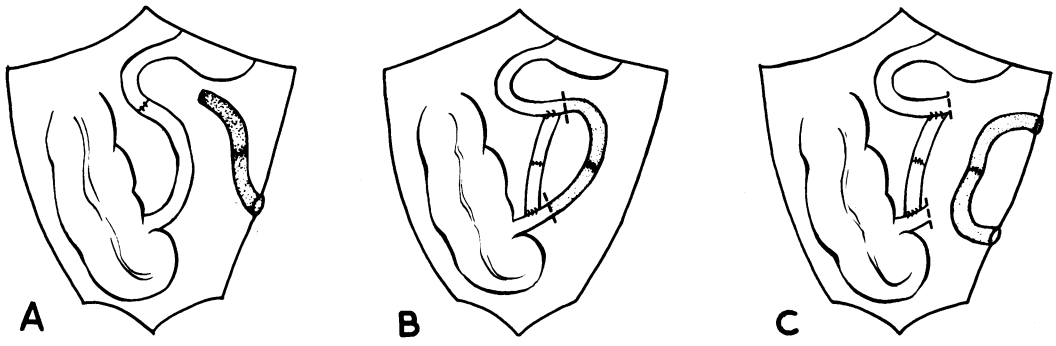

Figure 2. Experimental study. A: First phase. Lengthened small bowel segments converted to self-emptying blind loops with ostomy opening on abdominal wall. B: Second phase, operation 1. Jejunal loop lengthening and anastomosis in parallel with 25% small bowel after 75% mid small bowel resection. C: Second phase, operation 2. Total dependence on lengthened segment. Resection of residual 25% small bowel (initially preserved as Thiry-Vella fistula but later resected)

Table 1. Phase 2 of experimental studies in pigs: 20–40 cm jejunal loop lengthening; 75% mid small bowel resection. Subsequent total dependence on lengthened segments

No.	Survival	Total loop dependence	Macroscopic analysis	Histology
5	29 wks	8 wks	Patent, dilated lumen Increased wall thickness Isoperistalsis	Villous hypertrophy
6	28 wks	7 wks	Patent, dilated lumen Increased wall thickness Isoperistalsis	Villous hypertrophy
7	25 wks	4 wks	Patent, dilated lumen Increased wall thickness Interloop fistula	Villous hypertrophy
8	30 wks	15 wks	Patent, dilated lumen Increased wall thickness Isoperistalsis	Villous hypertrophy
9	19 wks	4 wks	Patent dilated lumen Increased wall thickness Interloop fistula	Villous hypertrophy

ligament of Trietz, and the ileum just proximal to the ileocaecal valve, in parallel with the residual 25% small bowel. Five animals, 5–8 weeks old and weighing 7–15 kg, were allowed access to an unrestricted diet (pig weaner/grower meal, Oakes Millers, Congleton) postoperatively. The animals initially failed to thrive and developed diarrhoea. However, 3–5 weeks later the motions began to form and a sustained weight gain commenced. At a second laparotomy (Figure 2c), 21 weeks after lengthening, complete dependence on the lengthened segments was established by excision of the residual 25% of small bowel. At this procedure the lengthened segments were noted to be viable and patent, to have dilated, and to undergo effective isoperistaltic contractions. Postoperatively on a normal unrestricted diet, the animals developed intermittent diarrhoea and weight loss but were otherwise active. Fluoroscopic studies were performed 4–15 weeks after loop dependence. Under sedation with azaperone (Suicalm) 6 mg/kg, a peroral transpyloric tube was passed into the first part of the duodenum. Barium was observed to pass freely through the lengthened segment which demonstrated isoperistaltic activity. Considerable luminal dilatation had occurred and the mucosa demonstrated a rugose pattern. All 5 animals were alive and came to terminal laparotomy (Table 1). Viability, luminal patency and dilatation, and isoperistalsis in the lengthened segments were confirmed. The bowel wall was thickened and the mucosa markedly rugose. However, no increase in length had occurred and two specimens contained interloop fistulae. Histological examination demonstrated villous and muscle coat hypertrophy when compared to control specimens taken at the first procedure, features consistent with an adaptation response.

The experimental study thus confirmed the feasibility of safely constructing a vascularized loop of small bowel up to twice its original length, while simultaneously halving its diameter, preserving maximal mucosal surface for intestinal adaptation and maintaining isoperistalsis. Furthermore, such lengthened segments in a short gut model demonstrated an adaptation response.

Clinical application (Table 2)

Case 1: The first clinical application of the technique was reported by Boeckman & Traylor (1981). A male child born with gastroschisis and extensive midgut necrosis was left with 39 cm jejunum. For 4 years the child remained dependent on parenteral nutrition. At laparotomy for intestinal lengthening the bowel measured 50 cm in length from pylorus to jejunocolic anastomosis, and 11 cm in diameter. Thirty-two centimetres were divided to give a total small bowel length of 82 cm. Ten weeks postoperatively the child was able to maintain himself solely on enteral alimentation. Seven months later he had gained 2.1 kg, was on a regular diet and passing three semiformed motions daily. He now attends school.

Case 2: The second child to undergo intestinal lengthening was operated on by Bomar and Bieman Othersen in 1981 (personal communication). A female child born with gastroschisis and extensive gut necrosis had a residual 28 cm jejunum. At 9 months of age the child, still dependent on parenteral nutrition, underwent

Table 2. Clinical application: intestnal lengthening related to establishment of total enteral nutrition postoperatively

Case	Age	Aetiology	Bowel diameter (cm)	Bowel length	Total enteral nutrition
1	4 yrs	Gastroschisis	11	50 cm→82 cm	10 weeks
2	9 months	Gastroschisis	6	54 cm→101 cm	12 weeks
3	7 wks	Jejunal atresia	6	37 cm→58 cm	8½ weeks
4●	3 wks	Jejunal atresia	5	30 cm→42 cm	12 weeks

●Intestinal obstruction 19 weeks postoperatively; now recovering

intestinal lengthening. Twelve weeks later she established total enteral alimentation and 7 months postoperatively had gained 2.2 kg in weight and was passing 3–4 formed motions daily. She has remained well on a normal diet and continues to put on weight.

Case 3: Our first case, the third child to undergo intestinal lengthening, was operated on at 7 weeks of age in February 1982 at the Royal Manchester Children's Hospital. She was a 2.28 kg female, born with a high jejunal atresia at 35 weeks gestation following a normal pregnancy and delivery. At operation the intestine consisted of 37 cm jejunum, measured along the antemesenteric border with the bowel distended with gas from the ligament of Trietz to the bulbous tip of the dilated loop. The distal bowel consisted of 1 cm of ileum and the colon. An end-to-end jejunoileal anastomosis and a side jejunostomy were performed and the child maintained on parenteral nutrition. Because enteral alimentation led to increased ostomy losses, the side ostomy was closed at 5 weeks of age, with resulting

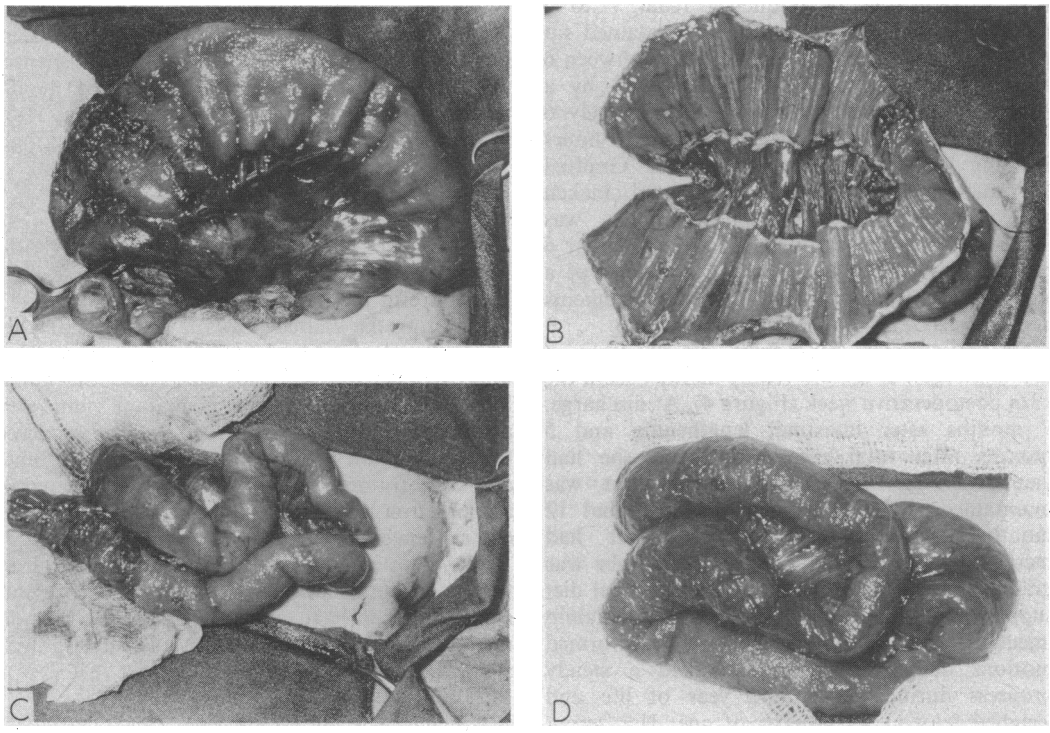

Figure 3. Case 3. A: Residual 37 cm jejunum. B: 21 cm jejunum divided by manual technique, maintaining vascular supply to each segment. C: Jejunum sutured to form two hemiloops of half original diameter. D: Isoperistaltic hemiloop anastomosis increasing total jejunal length to 58 cm

intestinal obstruction. Fluoroscopy revealed ineffective hyperperistalsis in the massively dilated jejunal loop and a patent jejunoileal anastomosis.

Intestinal lengthening was performed at 7 weeks of age when the child weighed 2.4 kg. The jejunum had a diameter of 6 cm and an unchanged length of 37 cm (Figure 3A). The jejunoileal anastomosis was patent. Using bipolar diathermy, 21 cm of jejunum was divided (Figure 3B) manually and sutured (Figure 3C) to give a total isoperistaltic small bowel length of 58 cm, with an average diameter of 2.5 cm (Figure 3D). The jejunoileal anastomosis and ileocaecal valve were resected and an end-to-end jejunocaecal anastomosis established. Total operating time was 2¼ hours, with a blood loss of 40 ml.

Her postoperative course was uneventful and bowel actions commenced on the fourth post-operative day. Nutrition consisted of a reducing course of parenteral alimentation (complicated by three episodes of catheter-related sepsis), combined with a nasogastric Pregestimil and cholestyramine infusion. Total enteral alimentation was established 8½ weeks after the intestinal lengthening procedure. During this period her motions, originally watery, became semiformed, with a reduction in frequency from 7 to 3 motions daily. The stool initially contained fat globules but no sugars and pH varied between 6 and 7. Intestinal transit time, measured by a marker, was 5 hours at 4 weeks postoperatively, 6 hours at 10 weeks postoperatively and 10 hours by the 16th postoperative week. Gradual introduction of Baby Rice, comminuted chicken and gluten-free/lactose-free meat dinners was accomplished without intestinal upset, so that 6 months postoperatively nutrition consisted of a weaning diet with vitamin and iron supplements and 4-hourly Pregestimil feeds (280 ml/kg).

On this regimen, her weight, initially static at 2.45 kg, began to show a steady increase from the 13th postoperative week (Figure 4). At discharge, 7 months after intestinal lengthening and 5 months after total enteral takeover, she had gained 1.6 kg. Subsequent weight gain was maintained at 100–150 g per week, so that 12 months after lengthening her weight had increased by 3.6 kg. During this period she was gradually weaned to an unrestricted normal diet supplemented by a bimonthly hydroxocobalamin injection, and was passing two semiformed motions daily. She has maintained a steady progress during her second year of life and weighed 9 kg at 25 months of age. Her serum protein levels, serum iron, serum calcium, trace elements, and vitamin and folate levels are within normal limits. Thyroid function studies, liver

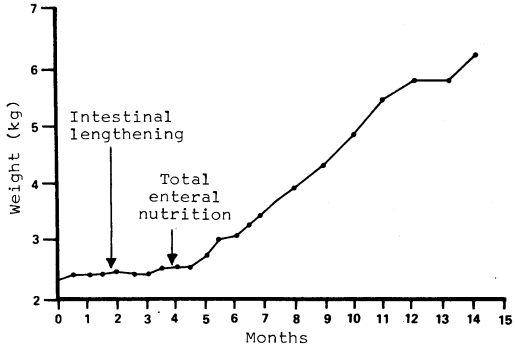

Figure 4. Monthly weight chart of Case 3

function tests, and a sweat test are normal; 24-hour urinary oxalate excretion is normal. Her bone age and physical development have improved rapidly and she now walks unaided. Her mental age and speech development have been consistent with her chronological age.

Case 4: The most recent child to undergo intestinal lengthening was operated on at 3 weeks of age by R J Brereton (1983, personal communication) and will be reported later. Following a high jejunal atresia, the child had 35 cm residual small bowel. Twelve weeks after intestinal lengthening she was able to dispense with intravenous alimentation. However, at 19 weeks she required a further laparotomy for intestinal obstruction and is presently recovering.

Conclusion

The present limited clinical experience with intestinal lengthening suggests that it is a safe procedure in the neonate as well as in the child. In tailoring the dilated bowel it preserves all the available mucosa for intestinal adaptation. The improved peristalsis avoids stasis and the blind loop syndrome with concomitant mucosal atrophy; while the increased length, reduced transit time and prolonged mucosal contact time aid absorption. Dependence on parenteral alimentation appears to be reduced and total enteral takeover facilitated (Table 2).

The intestinal lengthening procedure requires further careful evaluation, but, on present evidence, could have a useful place in the overall management of the child with the short-bowel syndrome.

Addendum: The author suggests a detailed index of all cases, and would be grateful to receive documentation of all applications of the procedure.

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