

The Child with Short Bowel : Lesson learnt over 30 years



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Agenda Slide

- 1981
- Definition
- Why
- Indication
- Results
- IRP
- A.G.I.R.
- Future



Manchester, UK
1981

-L.S. jejunal atresia
38 cm

LILT (Feb 1981)

Off PN (Sept 1981)

-21yrs BMI 20

Pregnant (B12, Folate, Iron
Zn)

Delivered 2.74 Kg

Male (7th %ile)

Successful pregnancy following maternal small bowel reconstruction for congenital short bowel syndrome

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Case report

Miss LS was born with jejunal atresia. She had lost most of her small bowel leaving her with 38 cm of massively dilated proximal jejunum distal to the duodeno-jejunal flexure. She retained the entire colon but lost her ileocaecal valve. Failure of conventional surgery to establish enteral function, initially with a jejunostomy followed by a jejunocolic anastomosis, led to a 'Blanchi' longitudinal intestinal lengthening and tailoring (LILT) procedure at seven weeks of age (see Fig. 1), with subsequent normal feeding.

Since then, Miss LS continued to grow and developed normally (body mass index of 20 at 21 years of age). Despite a greater number of liquid motions, she developed full faecal continence and was able to lead a normal quality life, requiring only vitamin B₁₂ supplements. She had her menarche at 13 years of age with a regular menstrual cycle thereafter.

Miss LS, a 21-year-old G3 P0 + 2, was referred to St Mary's hospital at 16 weeks of gestation for tertiary obstetric care because of potential concerns over her ability to cope with the nutritional demands of pregnancy. Her two previous pregnancies had resulted in first trimester miscarriages (vaginal bleeding and falling hCG levels). The index pregnancy was conceived spontaneously. Miss LS was managed by both an obstetrician and a gastroenterologist. She was receiving monthly injections of vitamin B₁₂ (hydroxocobalamin 1 mg im) and daily folate supplementation (5 mg), which continued throughout pregnancy. Regular assessment (every four weeks) of both haemoglobin levels and micronutrients was undertaken. Ferrous sulphate (200 mg tds) was commenced empirically at 20 weeks of gestation, and zinc supplementation (Soluzinc 135 mg bd) began at 28 weeks of gestation, following low serum levels (7.3, normal range 10–21).

An anomaly scan at 20 weeks of gestation indicated no gross fetal anomaly. Serial growth assessment showed that fetal growth was maintained on the 50th centile, with good growth velocity.

The pregnancy continued without any complication, and labour was induced at 39+ weeks of gestation, as Miss LS did not live locally. She delivered vaginally, after a 12-hour uncomplicated labour, a normal male infant weighing 2.74 kg, corresponding to the 7th centile for gestational age.

Discussion

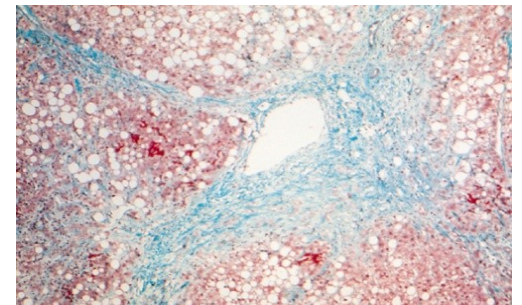
Bowel atresia is recognised to be secondary to an antenatal vascular insult.^{1,2} This may be segmental, with loss of an insignificant amount of bowel, but is more frequently the result of midgut volvulus because of malrotation or gastroschisis, with extensive loss of small and large bowel. Dead

bowel is reabsorbed and a high jejunal atresia, obstructed proximal to normal small bowel length, absorptive bowel is curtailed and is no longer available. Intestinal and nutrient loss, followed by macro and micronutrient deficiencies leads to an inability to absorb nutrients. The end result is excessive loss of bile salts and more than 75% of the infants survive is only possible with term survival dependent on the use of parenteral nutrition to achieve enteral feedings. Procedures such as bowel



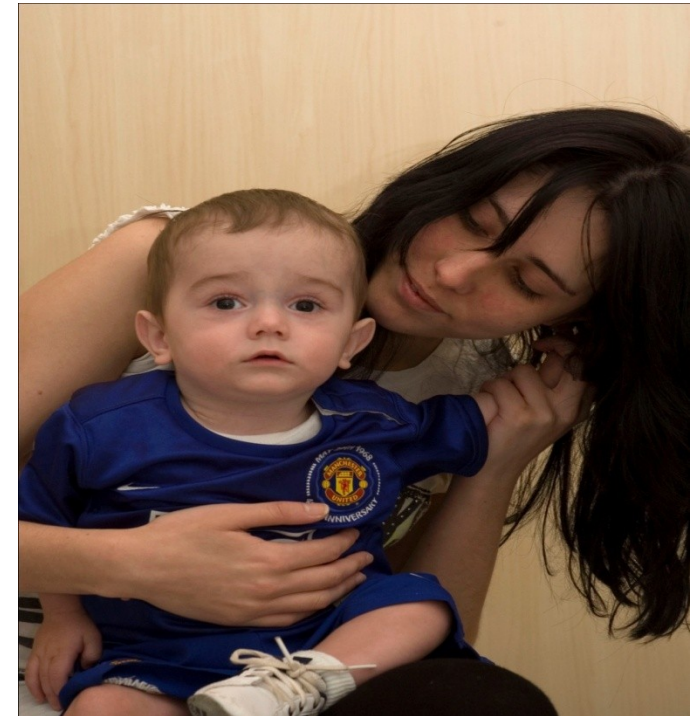
Initial Results 1982 - 1997

- 45 % Survival
- All 40cm and greater
- IFALD
- Loss of venous access



Definition

multisystem disorder caused by malabsorption of nutrients as a result of inadequate intestinal length



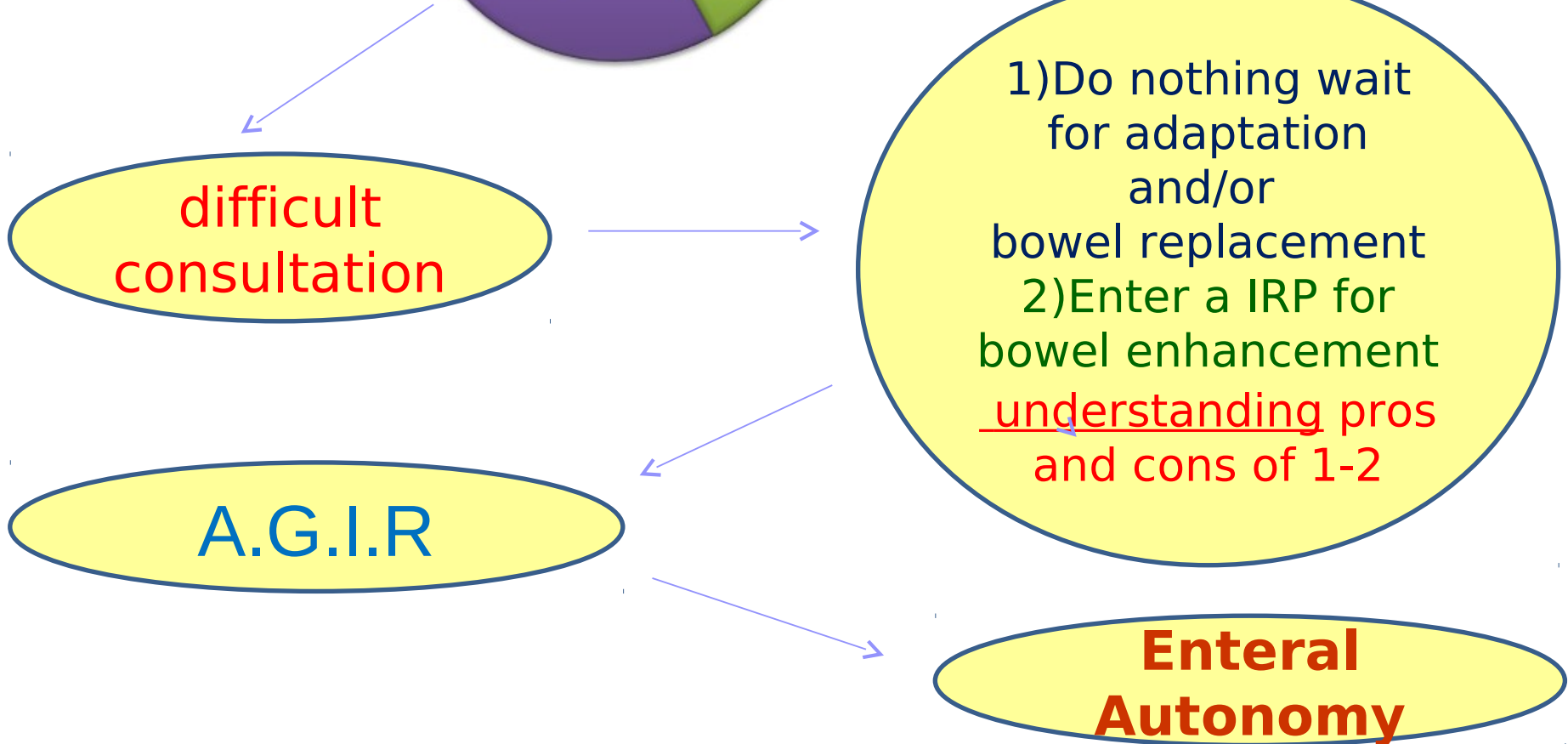
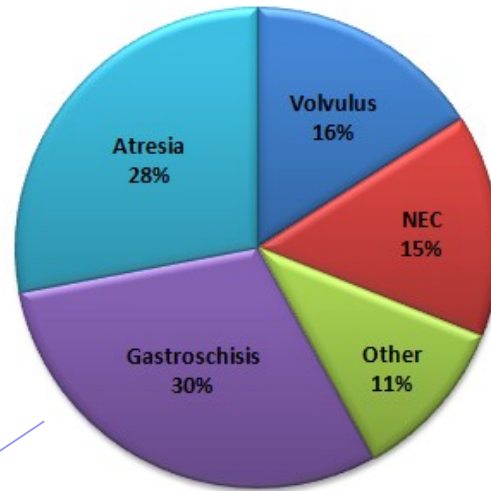
Present results

- Survival: 92%
- Off TPN: 96%
- Median length: 25cm

- Median time off PN:
 - 6 months post reconstruction

- Median Weight centile:
 - Pre-surgery: 0.4th centile
 - Post surgery: 9th centile





WHY A. G. I. R. ?

- Improve absorption
- Prevent liver failure
- Treat bacterial overgrowth
- Prevent sepsis
- Autologous Bowel





Care Giver Evaluation and Satisfaction With Autologous Bowel Reconstruction In Children With Short Bowel Syndrome: A Pilot Study.

Edge H, Hurrell R, Bianchi A, Carlson G, Zaidi T, Gozzini S, Khalil BA, Morabito A.
J Pediatr Gastroenterol Nutr. 2011 Sep 14.

INDICATIONS for AGIR

- Severe Short Bowel State
- Neonatal / Paediatric mucosa sparing
- Bacterial overgrowth when on 100% PN
- Failure to progress to Enteral Autonomy
- Clinically Significant Intestinal Dilatation

further INDICATIONS AGIR

- Bowel dilatation (following AGIR)
- Bacterial overgrowth (following AGIR)
- Lack of progress to Enteral Autonomy
(12-18m following first lengthening)

we realised.....

Bowel Elongation / Lengthening

are “**procedures**” alongside others

&

NOT a solution to short bowel state !!!

Therefore ...

A.G.I.R.

AUTOLOGOUS **G**ASTRO

INTESTINAL **R**ECONSTRUCTION

In the context of an
Intestinal Rehabilitation Programme

Aims in A.G.I.R.

Optimize bowel & patients
E.A. / healthy patient

Increase absorptive
surface area

Slower Transit Time

Improve
peristalsis

S U R V I V A L - Liver-sparing T P N

- **Veins/CVC preservation**
- **Early Oral Feeding** (Brain-Bowel)
- **Play therapy**
- **Social Integration**

**NON TRANSPLANT
SURGERY
(A.G.I.R)**

Transplant

Programme will **not work** if....

- **Multi-systemic** nature of disease is ignored
- **Little/NO** interest on CVC preservation
- **Gastro Team** focused on calories
- **Surgeons** focused on bowel length

I F.....

- **Ineffective** initial surgical procedure
- **?? Dilated loop / collapsed loop ?? IFABD**
(The “Dysmotility Syndrome”)
- **Stasis = Translocation = Sepsis =**
& more Problems.....



I F...

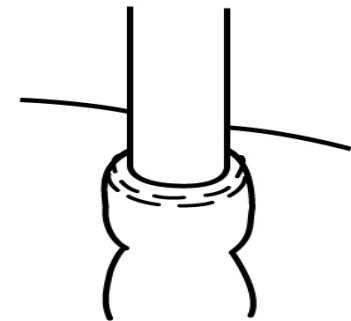
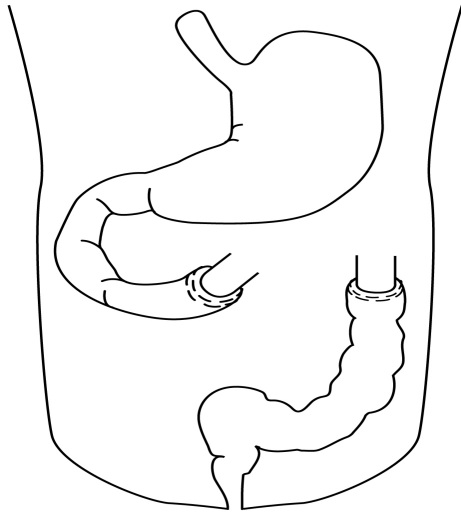


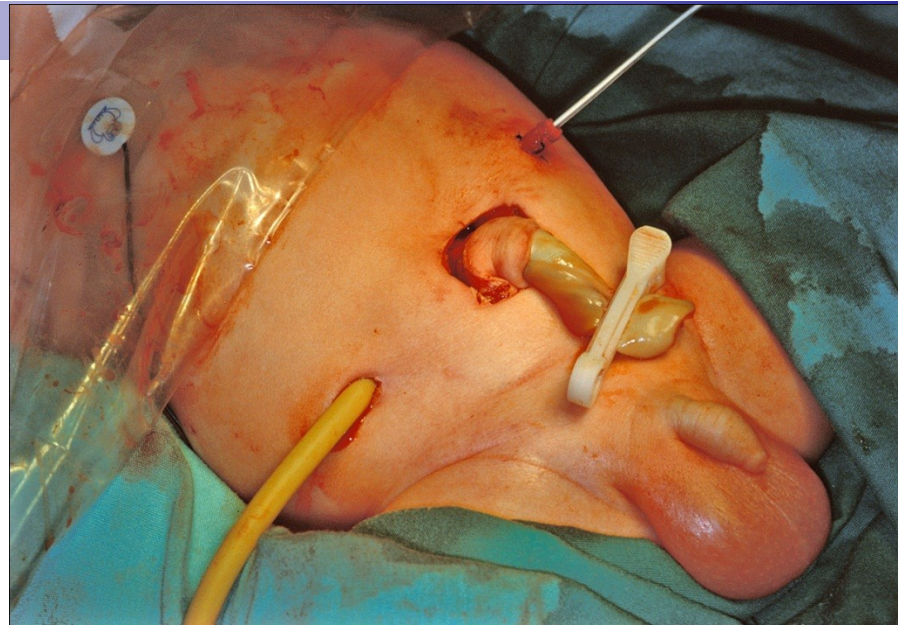
- Bad **Timing**
- Patient **NOT** ready
- Surgery done as an “**emergency**”
- By the **occasional surgeon** ! ! !

Staged Reconstruction

- Controlled Tissue Expansion (Bowel Dilatation)
- AUTOLOGOUS BOWEL RECONSTRUCTION
 - Single/Multiple procedures
 - +/- Slowing transit time

Controlled Tissue Expansion





- tube-jejunostomy
- tube-colostomy
- jejunal effluent is drip-fed down the distal stoma to develop the available distal bowel
- progressive clamping of the tube-jejunostomy increases mucosal contact, create tissue expansion & absorption often sufficient to reduce PN



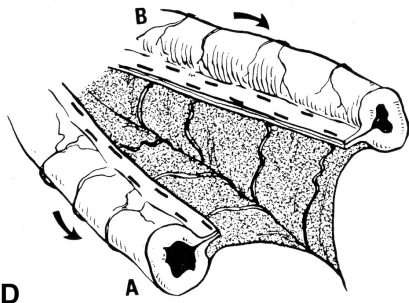
controlled obstruction
results in **dilatation, elongation**
of the intestine & **no stasis**
with **growth** of all layers of the bowel

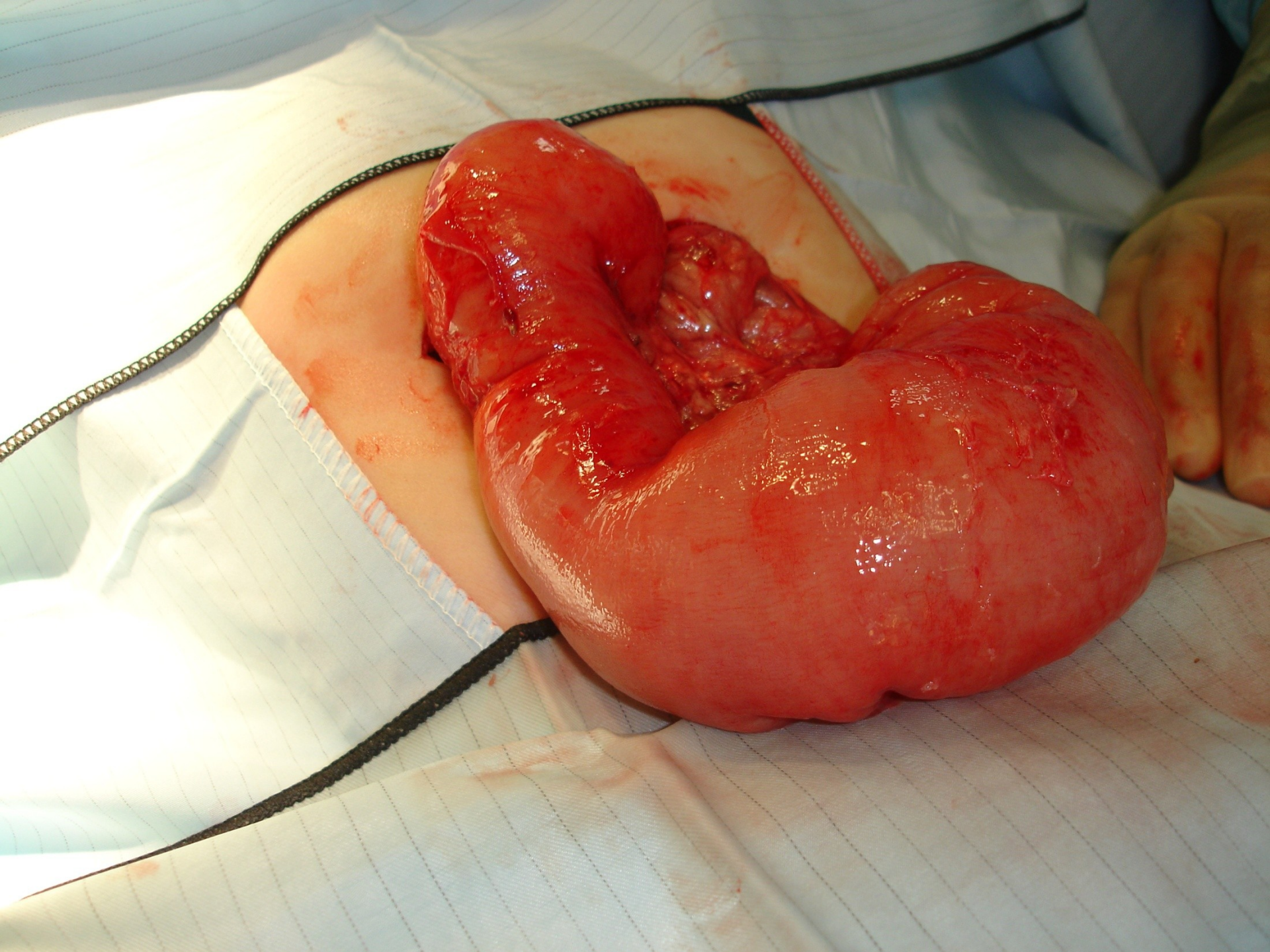
**Mucosal hyperplasia
not hypertrophy**

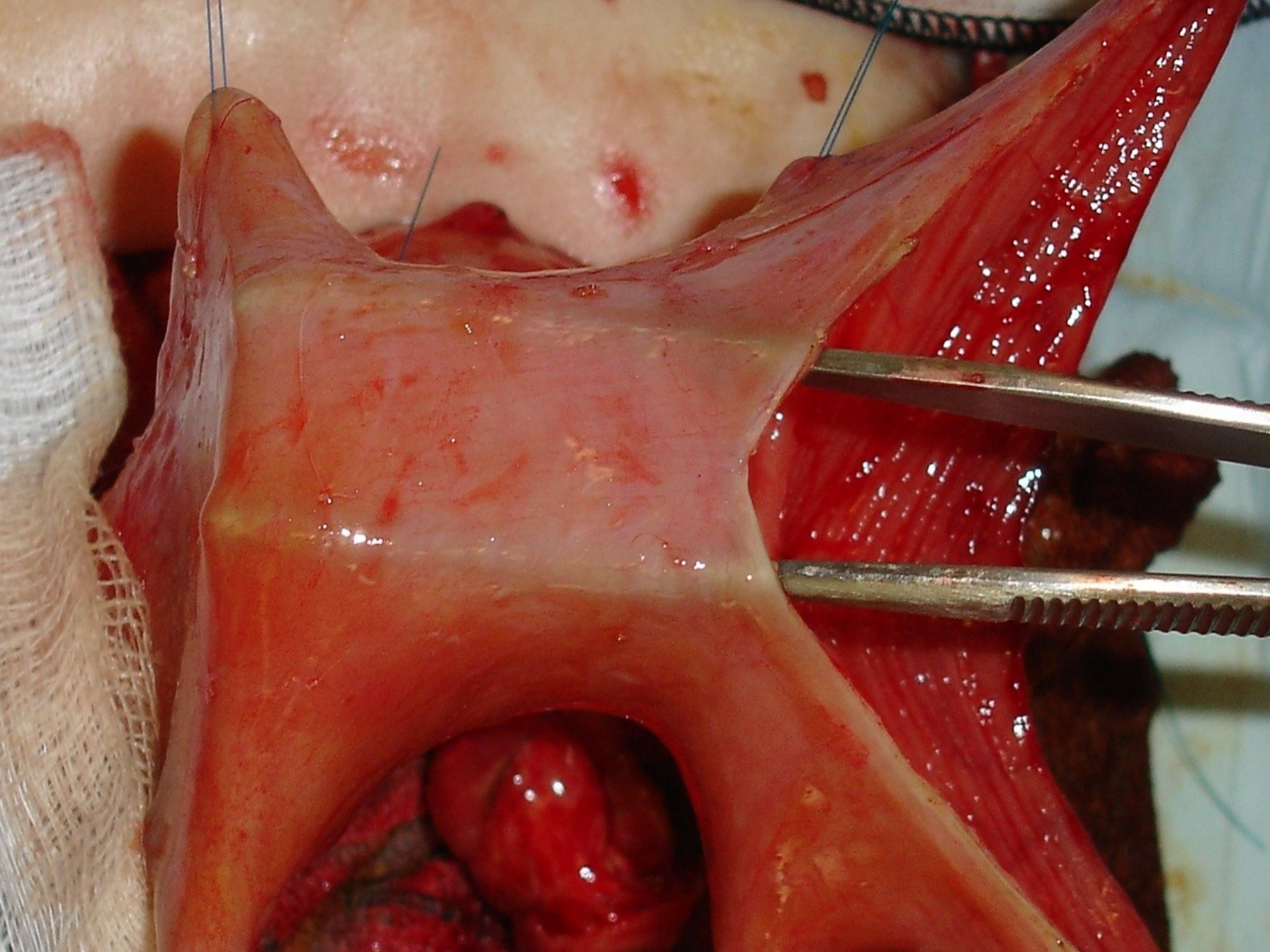
Longitudinal Intestinal Lengthening & Tailoring

Doubles (100%) the length of isoperistaltic bowel while **reducing** its diameter

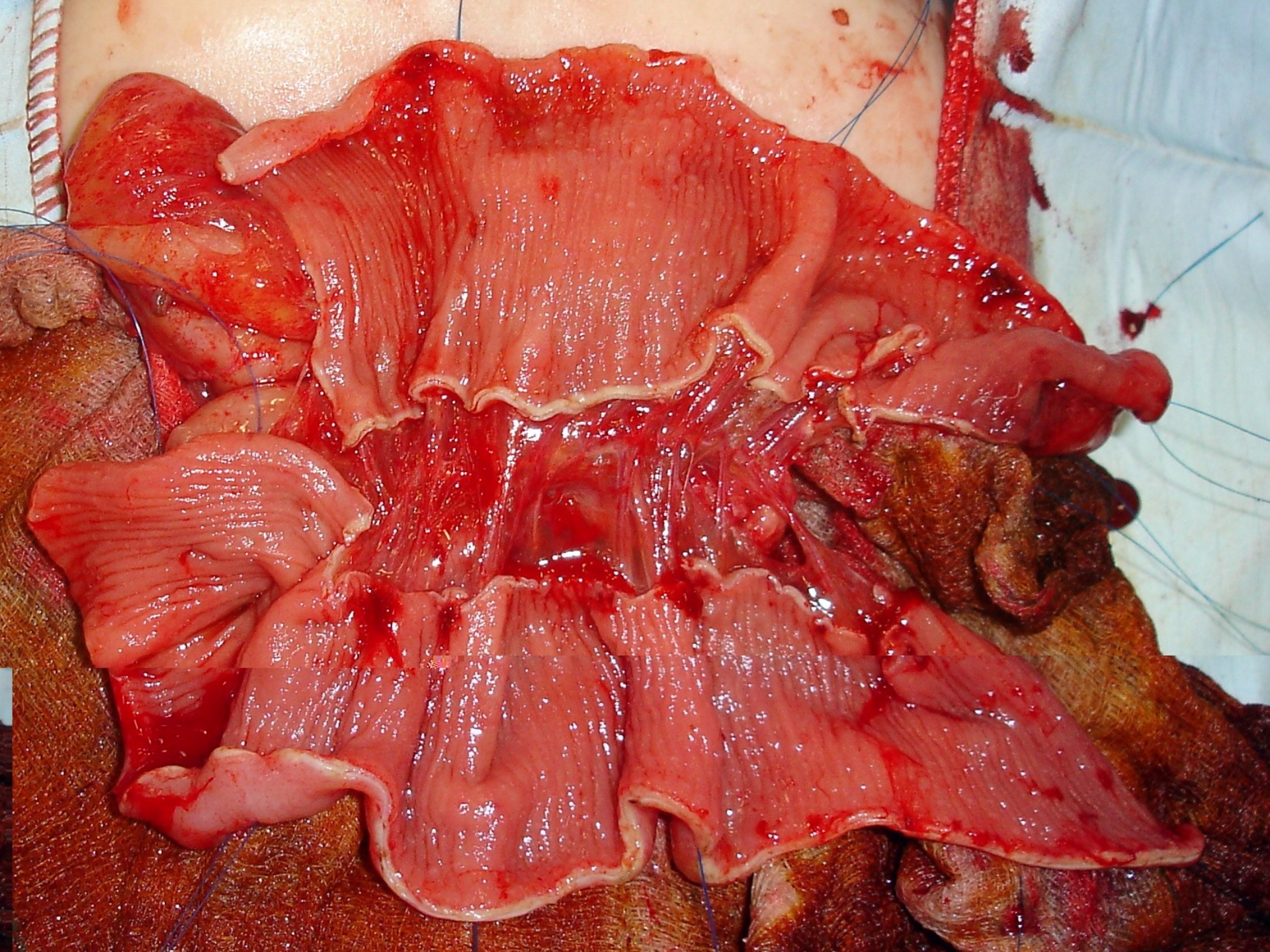
Better propulsion
No loss of mucosa
Avoids Stasis

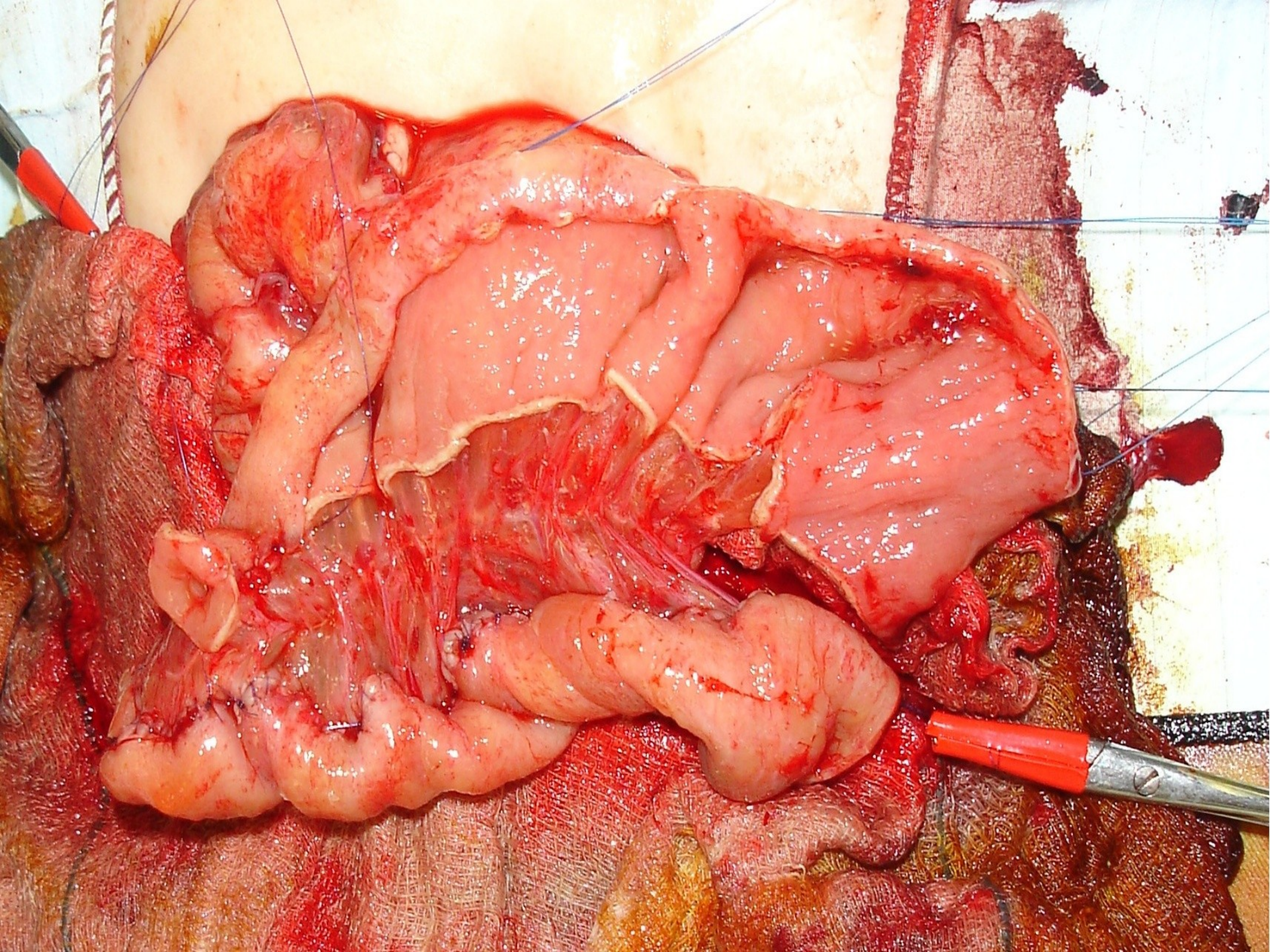






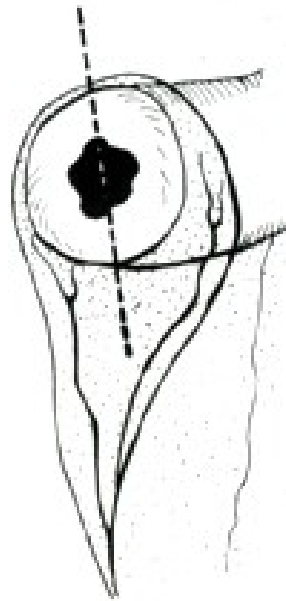




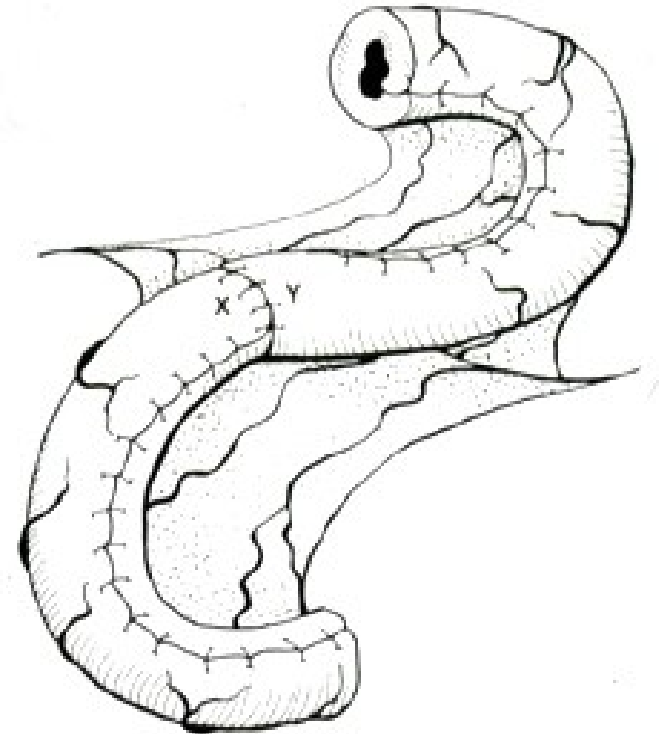
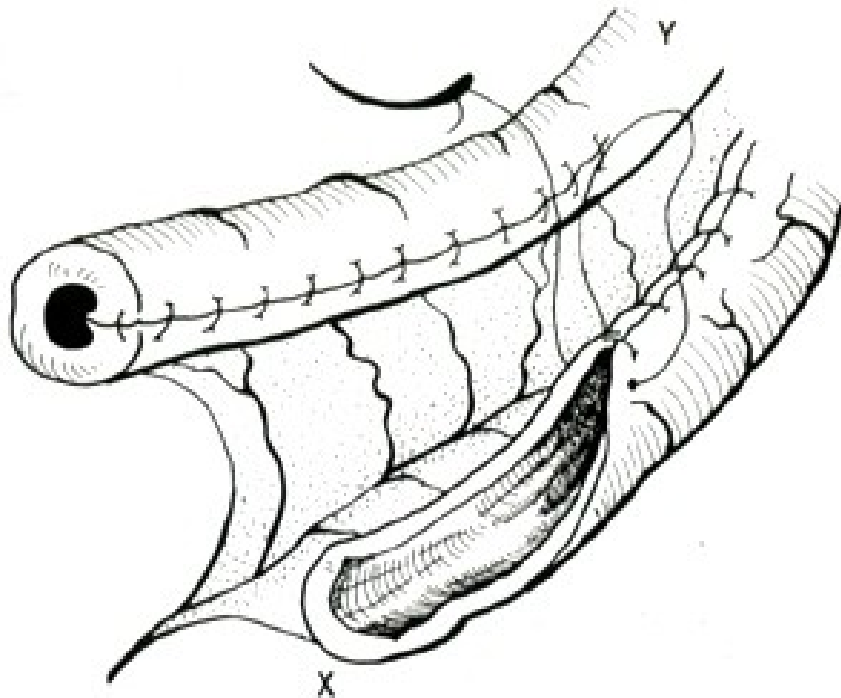


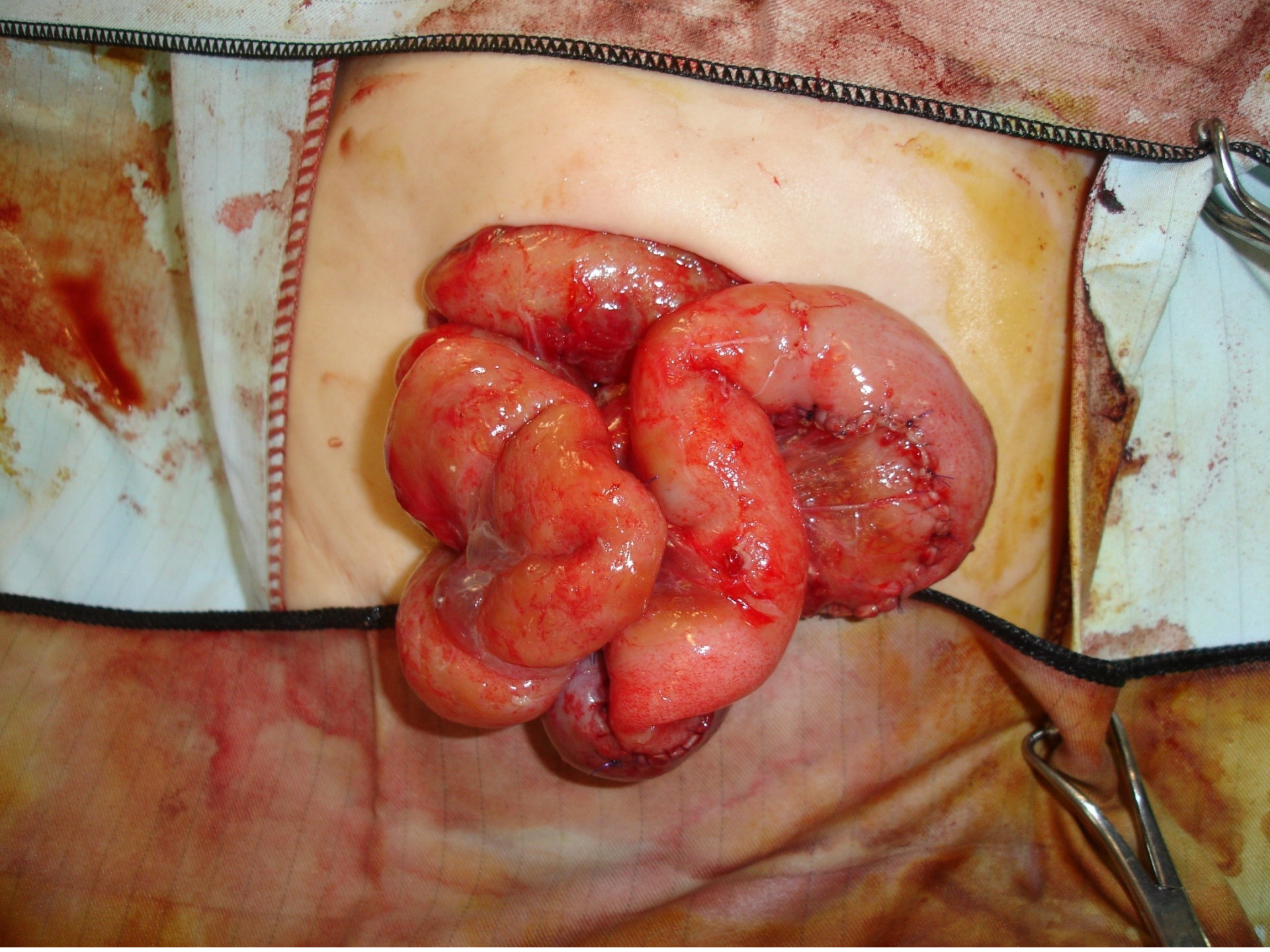
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1980

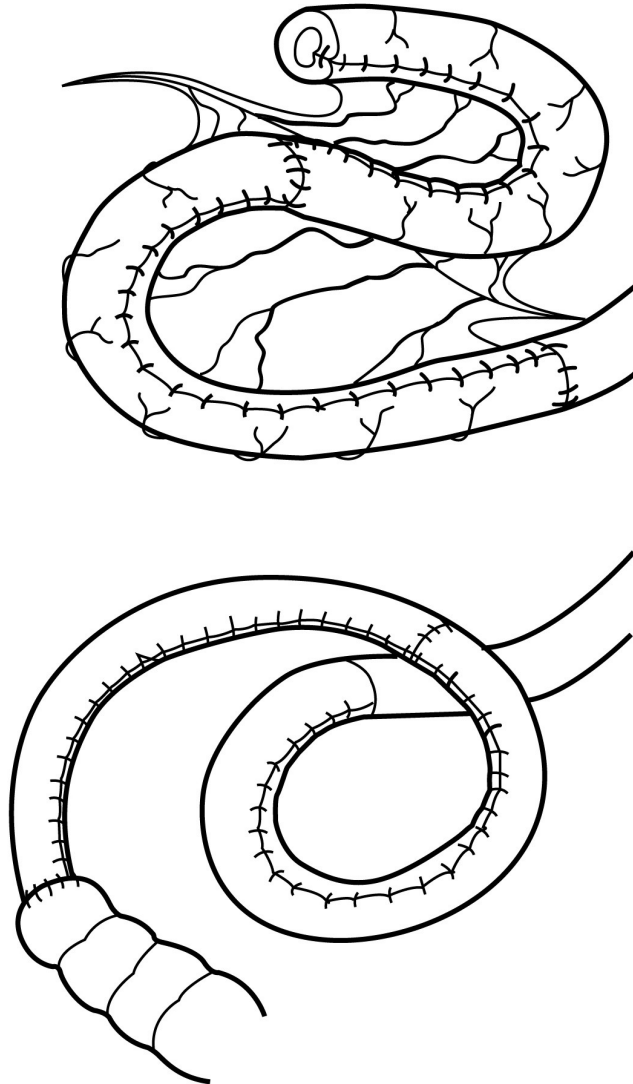


rabru





LILT Isoperistaltic anastomosis between hemiloops



Serial Transverse Enteroplasty STEP

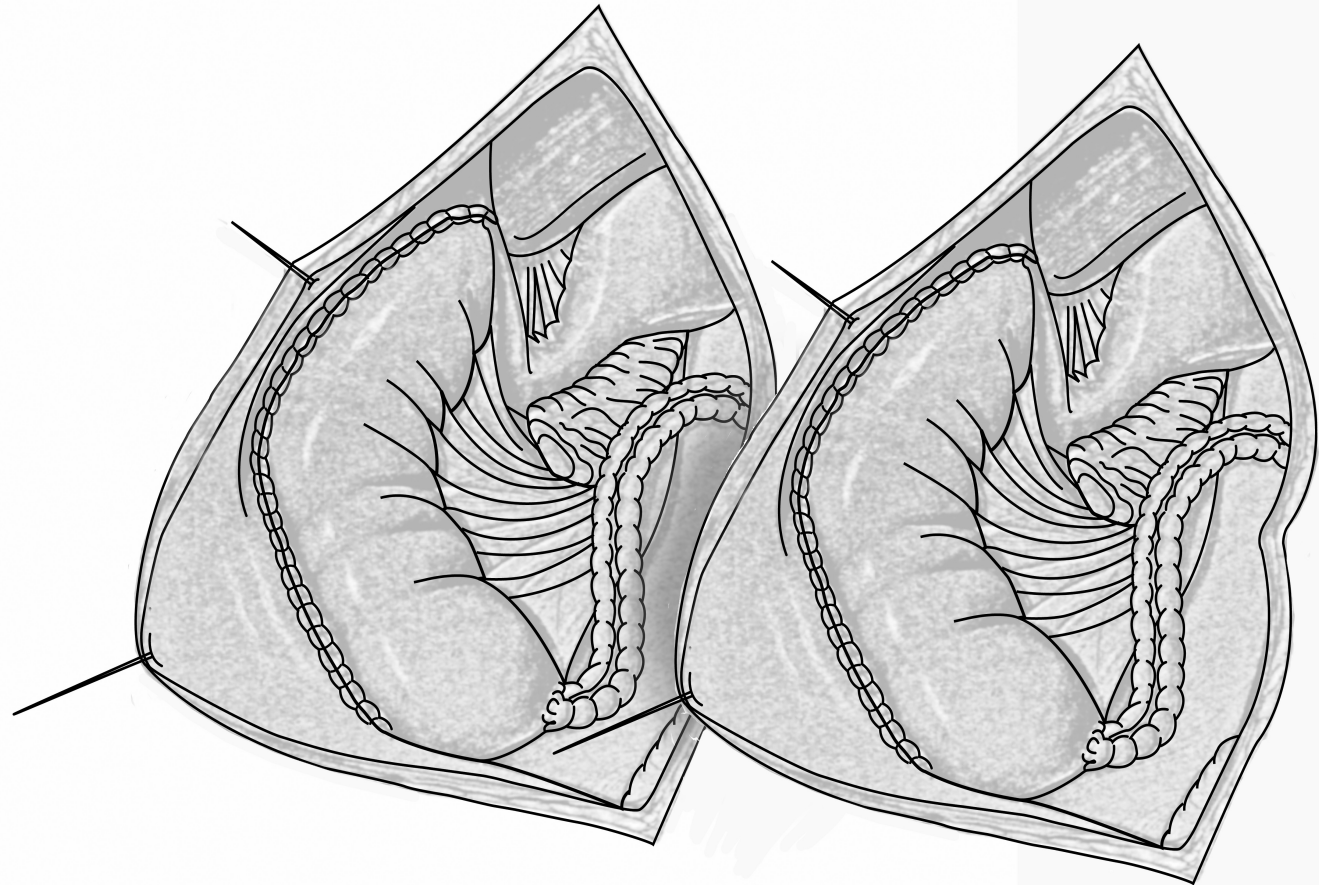


blood supply comes from the mesenteric border & traverses the bowel remaining perpendicular to the long axis of the bowel. Increases length by 68%

Can be performed primarily or after Bianchi-LILT

(Serial transverse enteroplasty for sbs. Kim HB, JPS,38 No 6 2003)

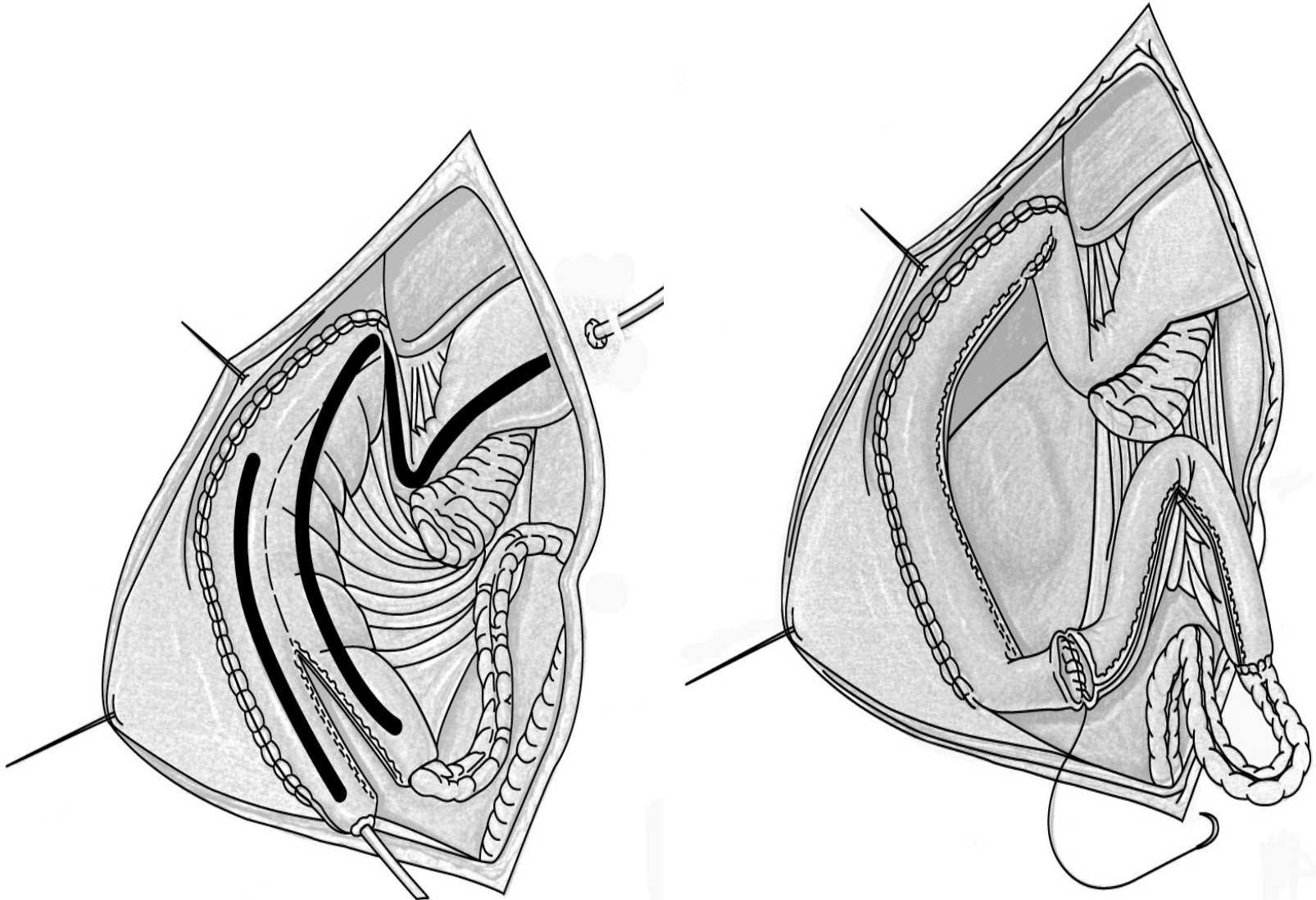
Isolated bowel segment lowa model



Suturing
of the SB
to a host
organ

Iowa model

once **new** blood supply has been achieved





Greater Manchester
POLICE

Protecting people

POLICE

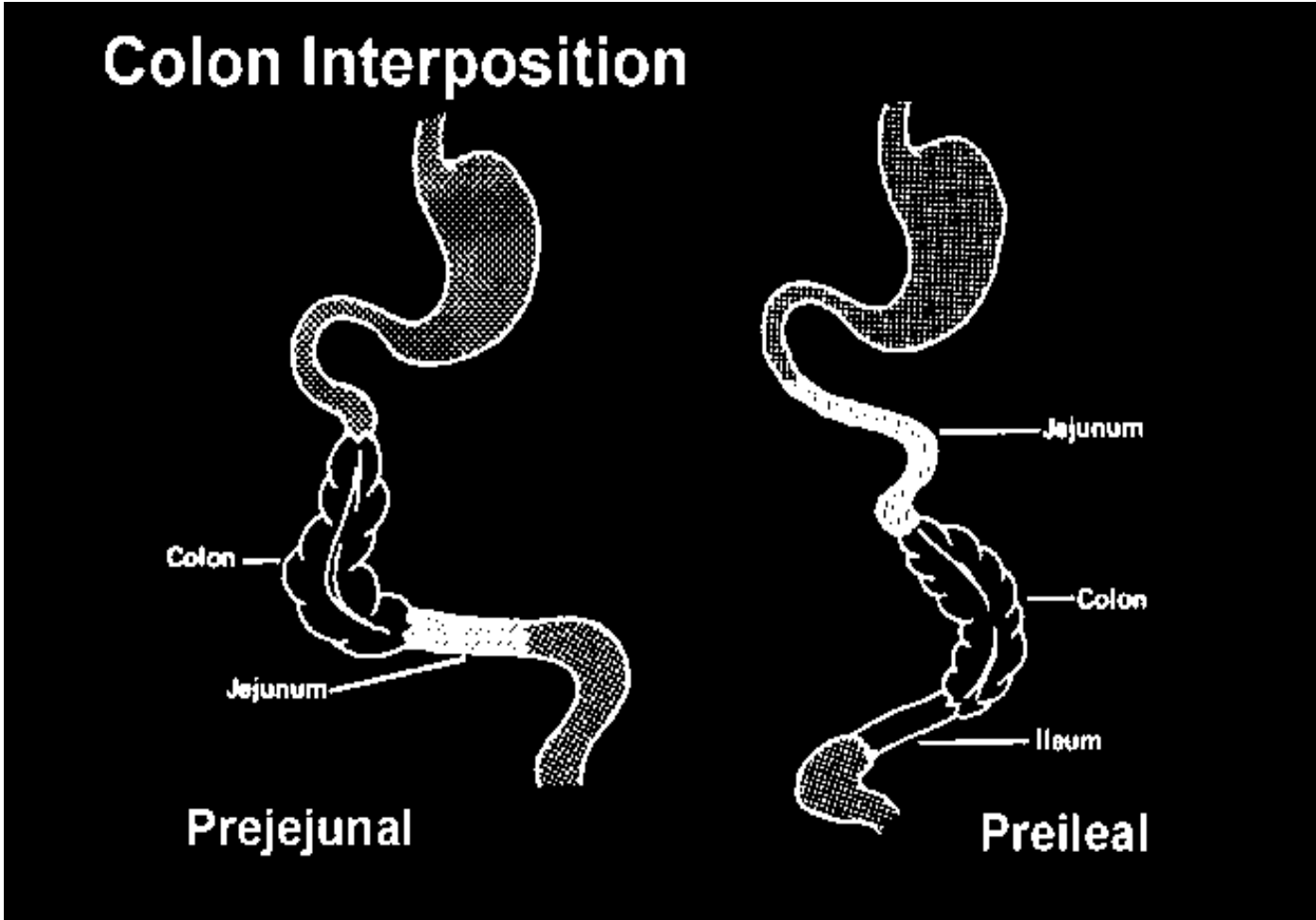
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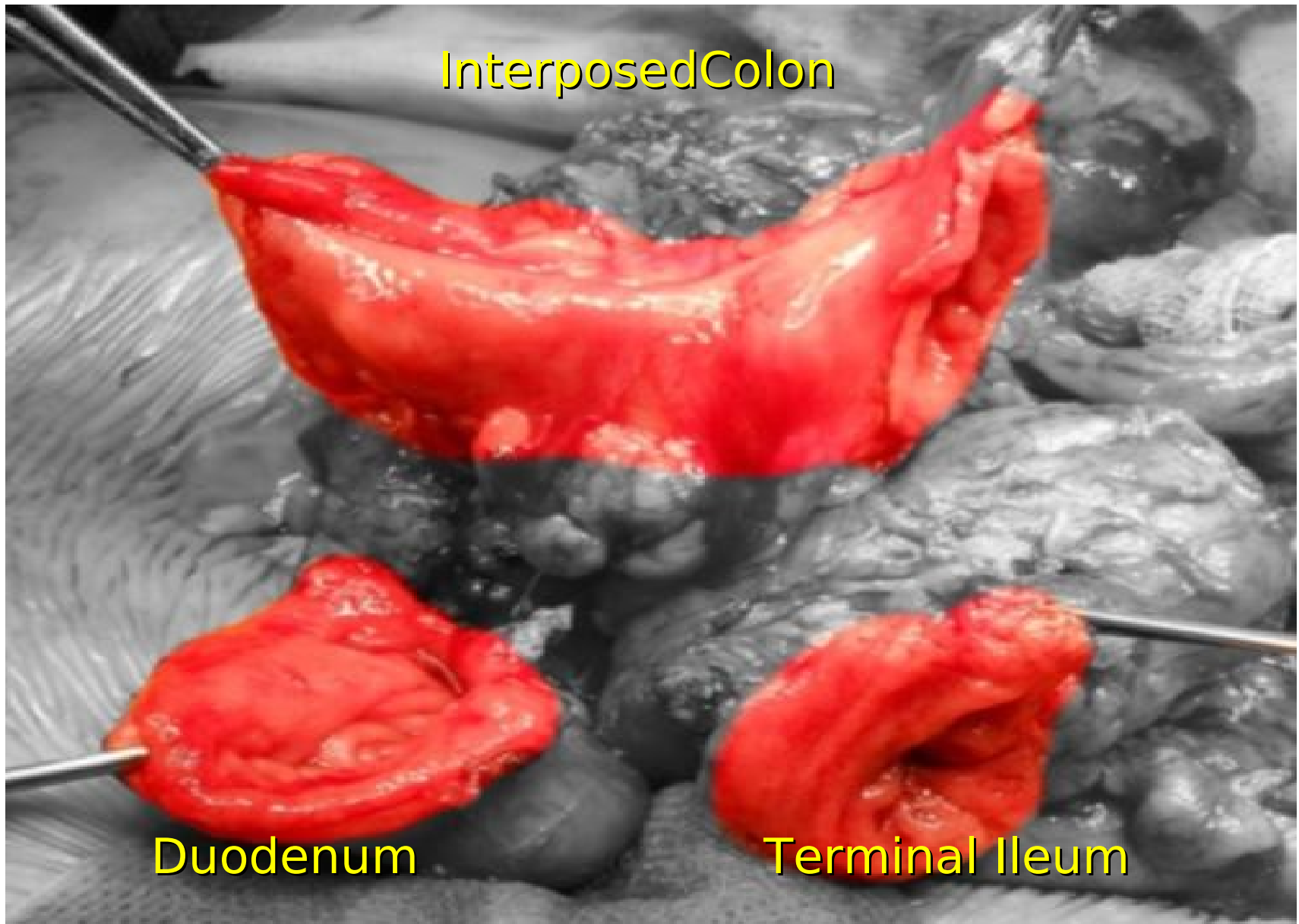
MX5

INDICATIONS for AGIR

- Short non dilated bowel BUT fast transit
(healthy patients on long term PN)

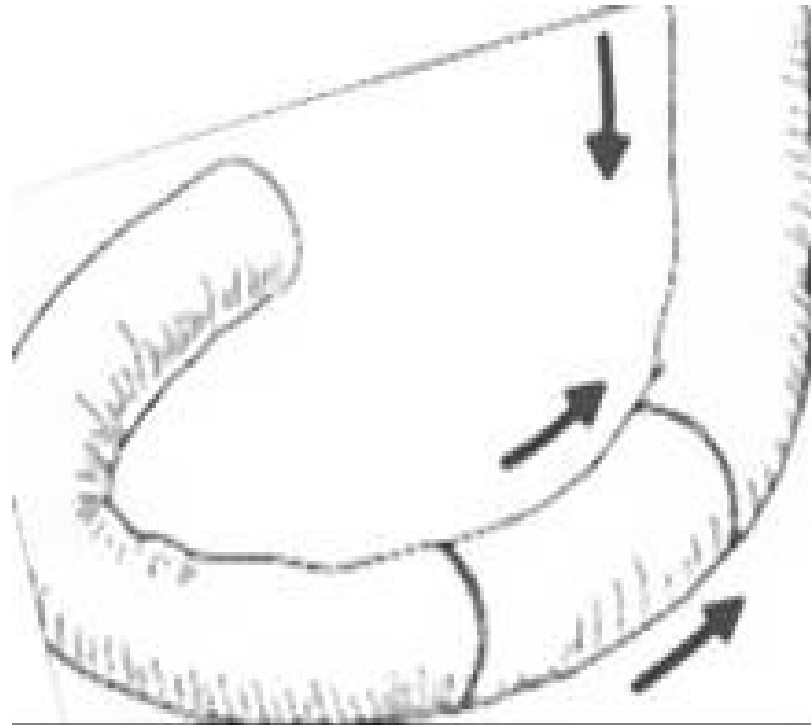
Slow transit Time: Colonic interposition

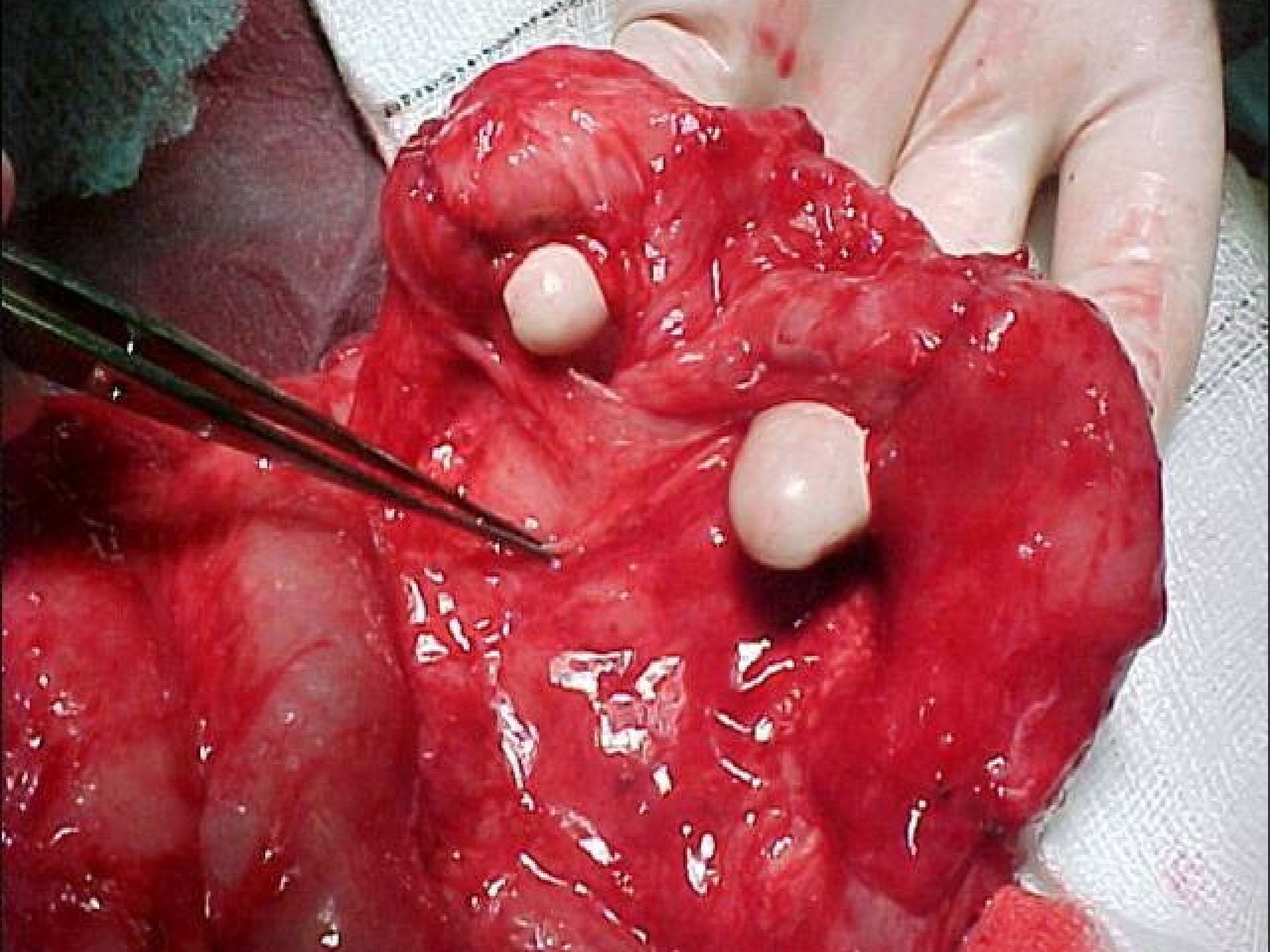




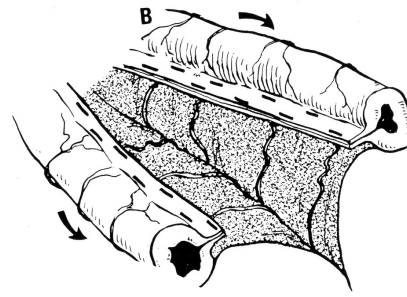
Slow transit time: Reversed segment

anti-peristaltic physiological delay
Usually between 3cm - 10cm

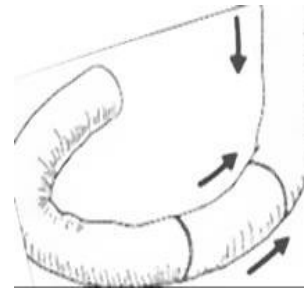
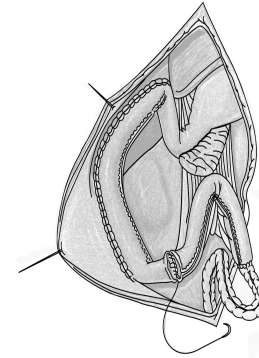
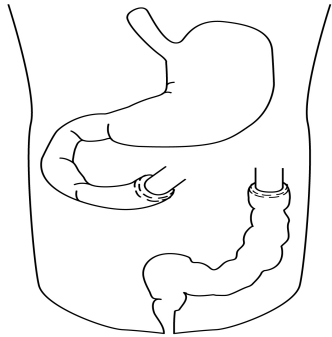
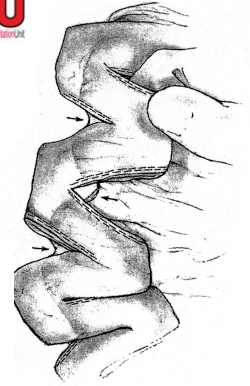




The present COMBINED TECHNIQUES



pabru
Papanicolaou Anastomosis and Bowel Resection Unit



Controlled Tissue Expansion

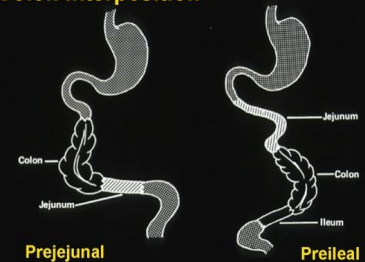
LILT - STEP - IOWA -

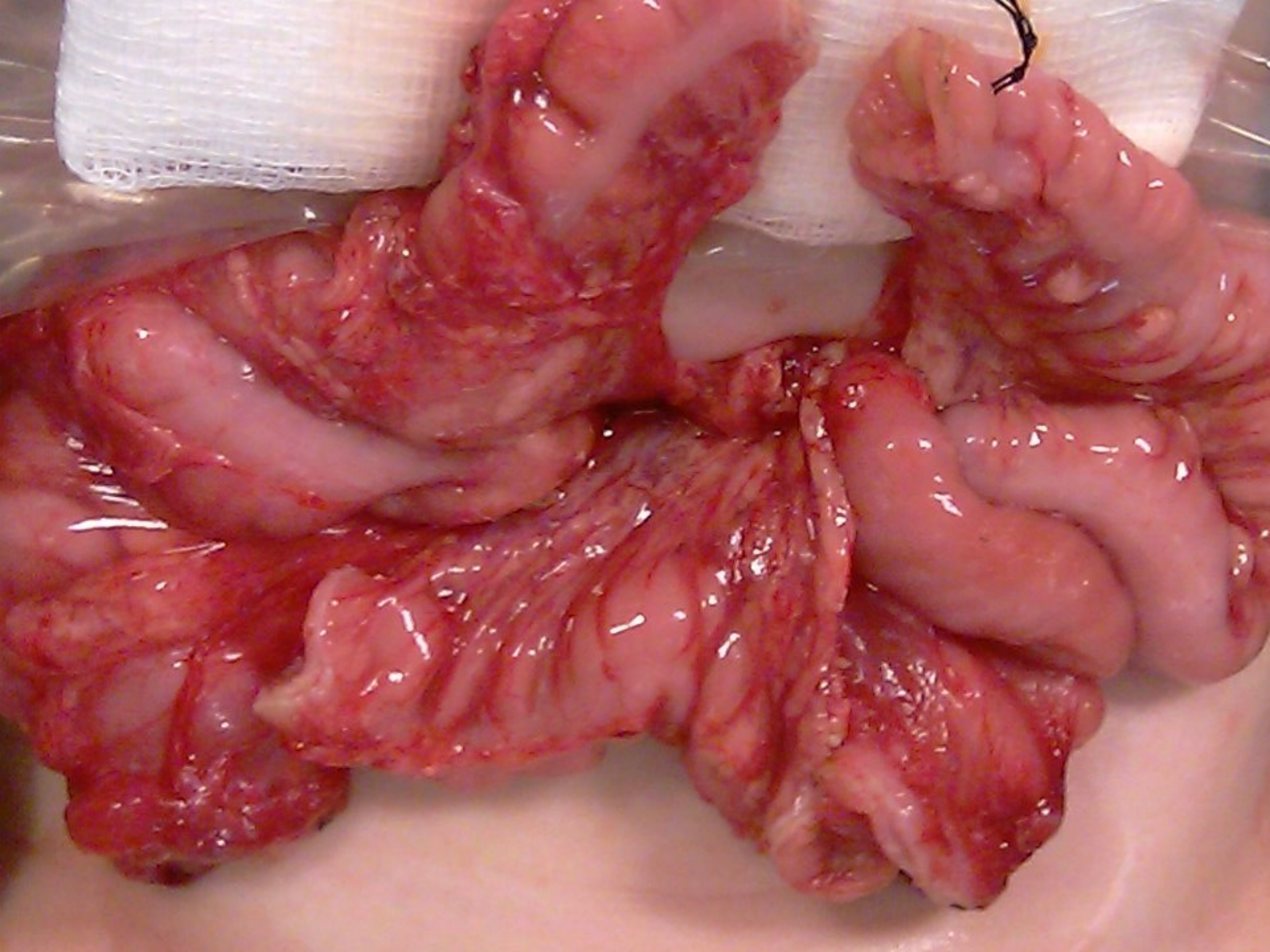
Single/Multiple Reversed Segment (s)

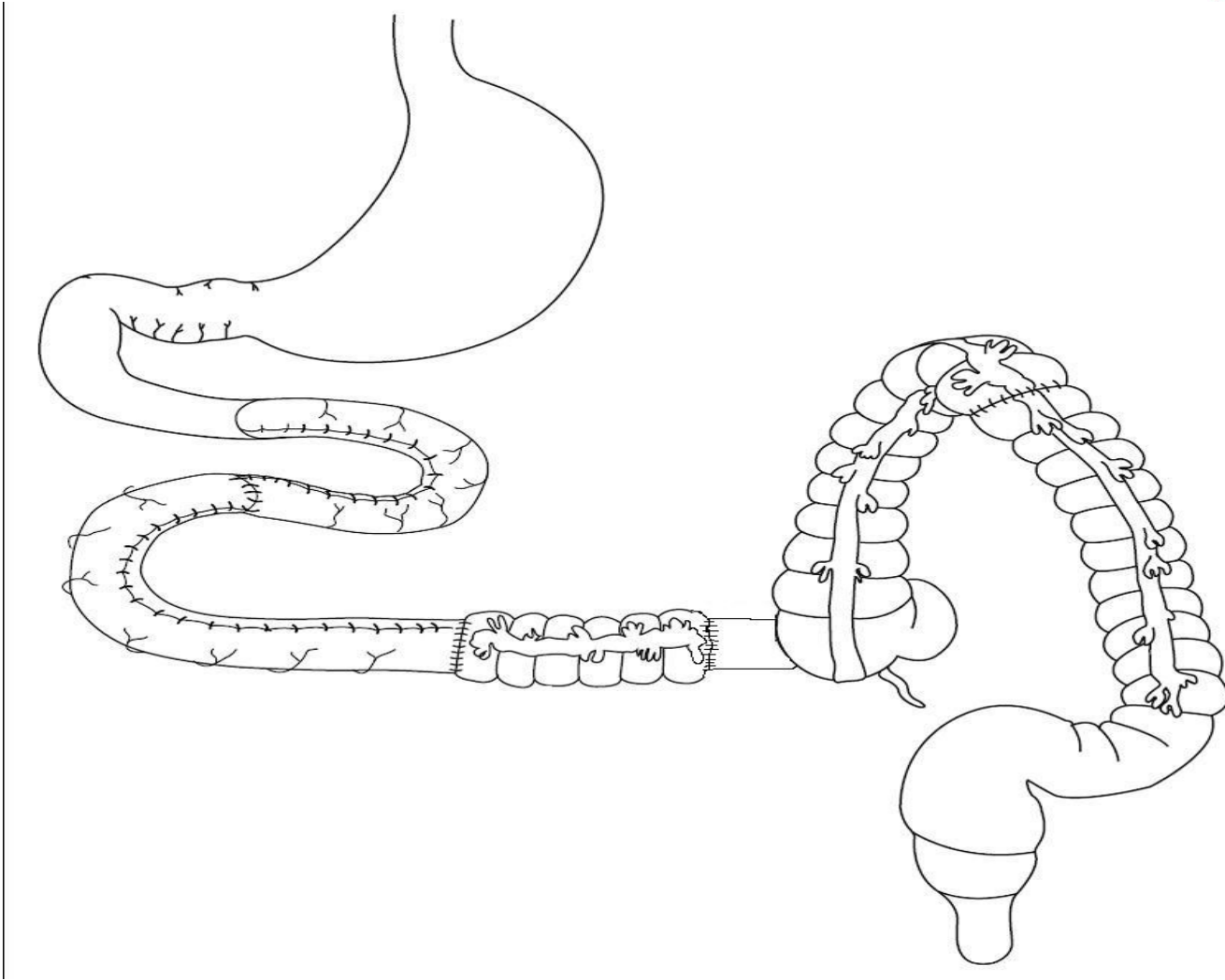
Colonic interposition

Re-lengthening

Colon Interposition







Managing short bowel patients

- It takes a lot of time
- It takes attention to details
- It takes personal experience & skill
- It takes knowledge of what has been done before by others
- It takes perseverance
- It takes the trust , motivation, strength of our patients & the families

PABRRU -TEAM

- Khalil BA
- Lynette Forsythe (dietician)
- Tracy Warburton (play therapy)
- Gastroenterologist
- Psychologist
- A. Shabani (Radiology)
- SALT
- Physiotherapist
- Nursing Staff- Specialist Nurse
- Social Services
- Sprs- Fellows- Medical Students

shortbowelsurvivor.co.uk



unfiloperlavita.it



Future: From bench to bedside !!!

- Improve tissue expansion's technique
- Lengthening procedures
- Tissue Engineered small intestine

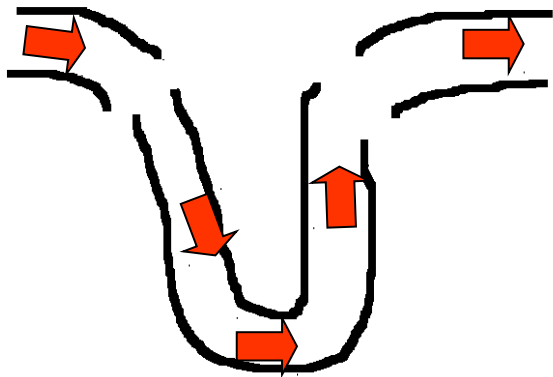


Spiral Intestinal Lengthening & Tailoring (SILT)

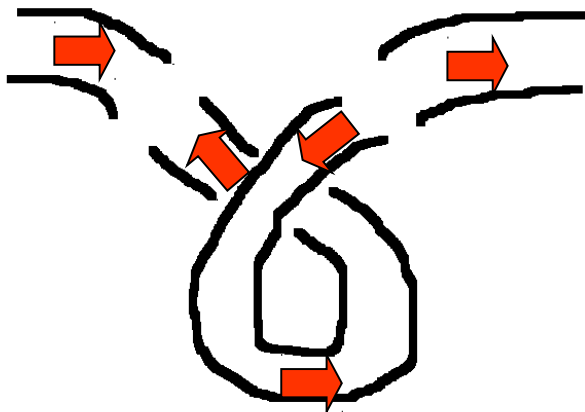


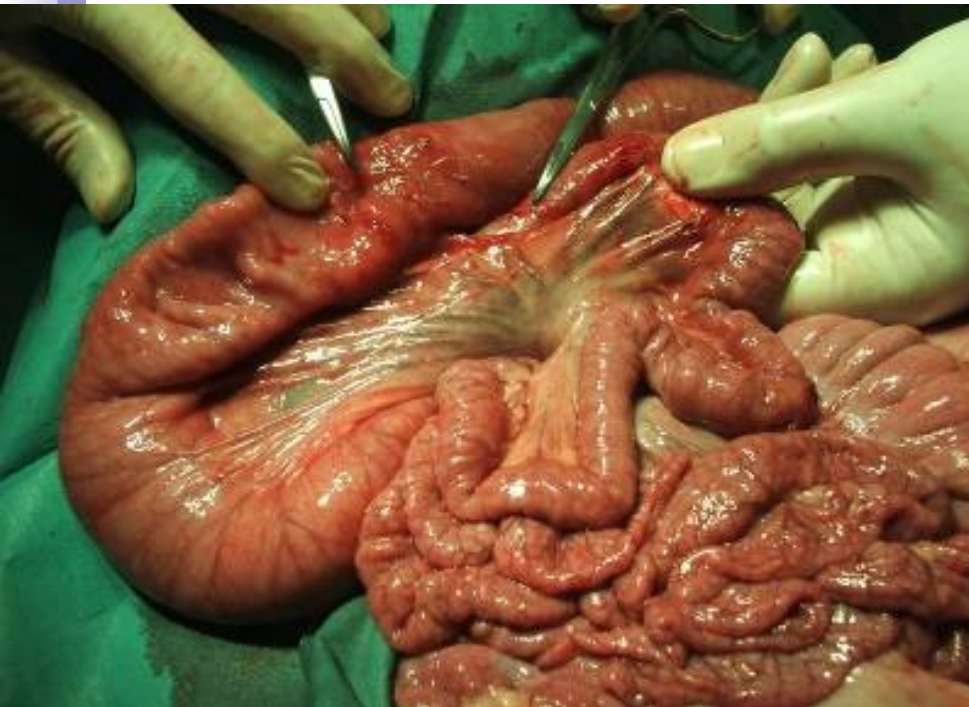
Creation of 15-cm-antiperistaltic reverse segment

1.



2.

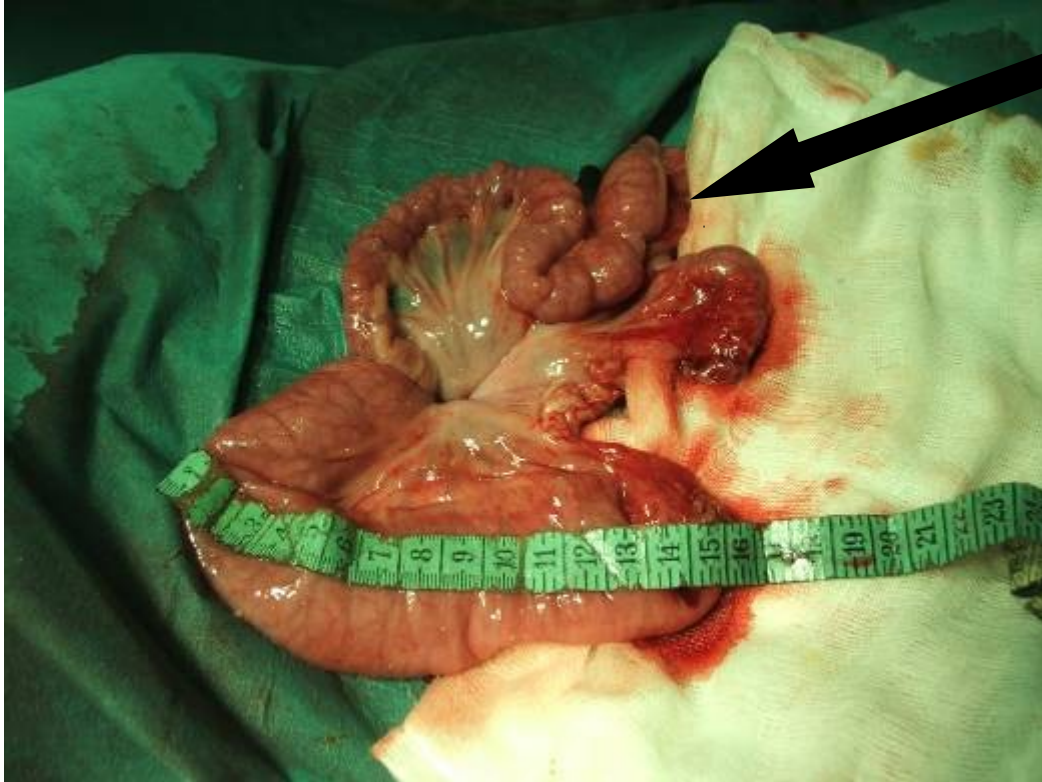




dilated small
bowel proximal
to the reversed
segment



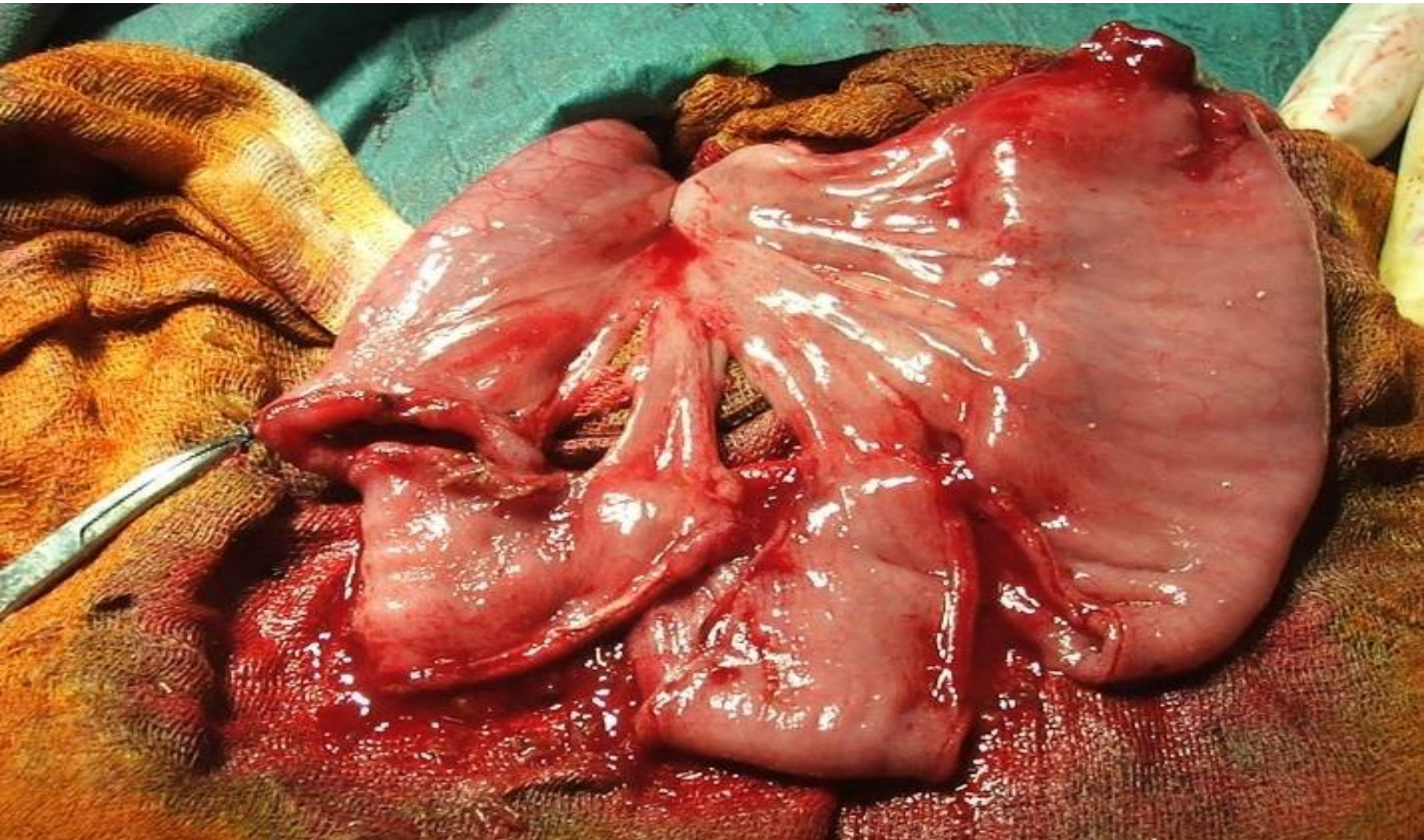
small bowel
dilated from 1 cm
to 4 cm



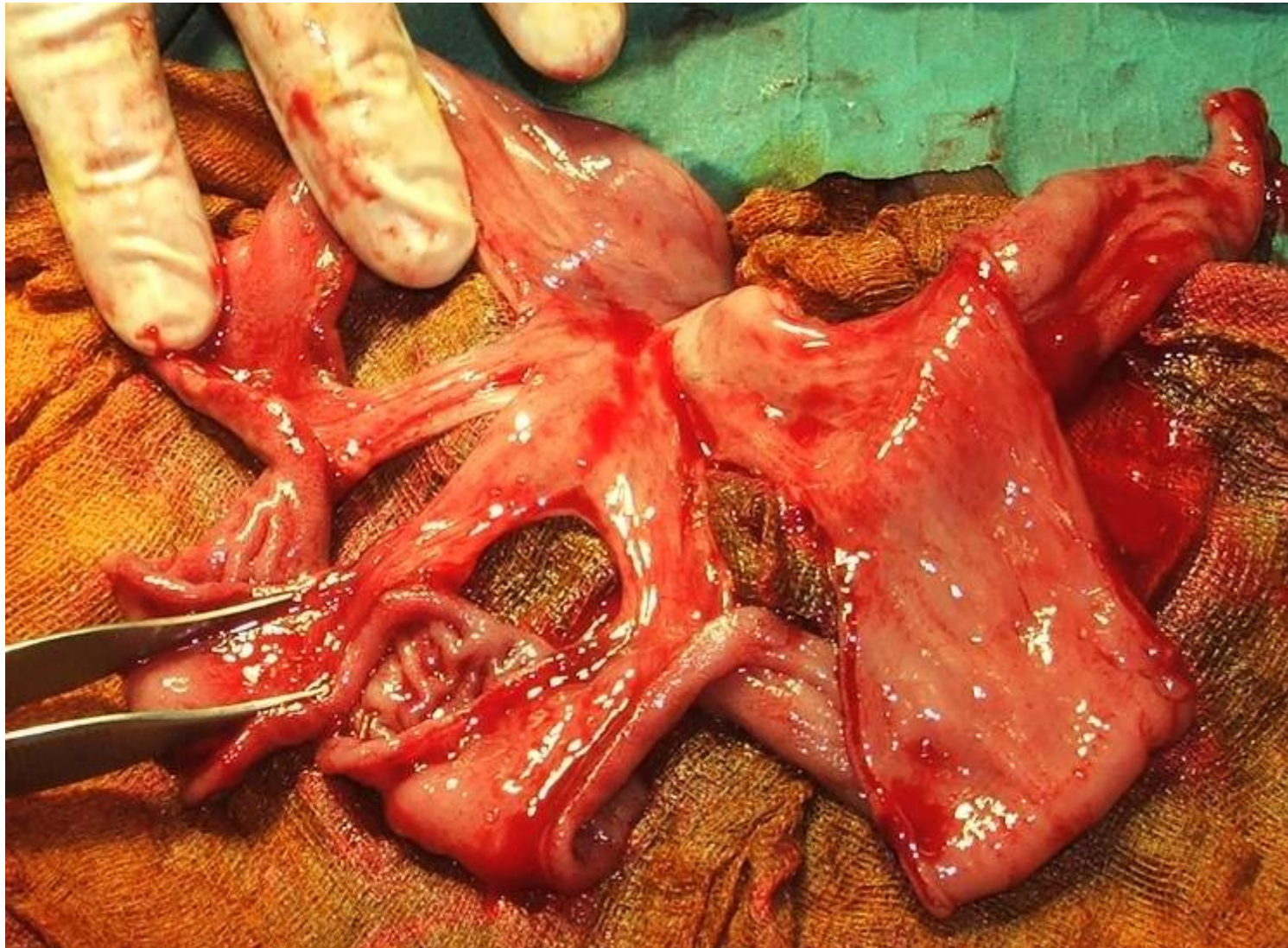
distal small bowel
with the original size

- 15 cm dialted small bowel was selected for SILT

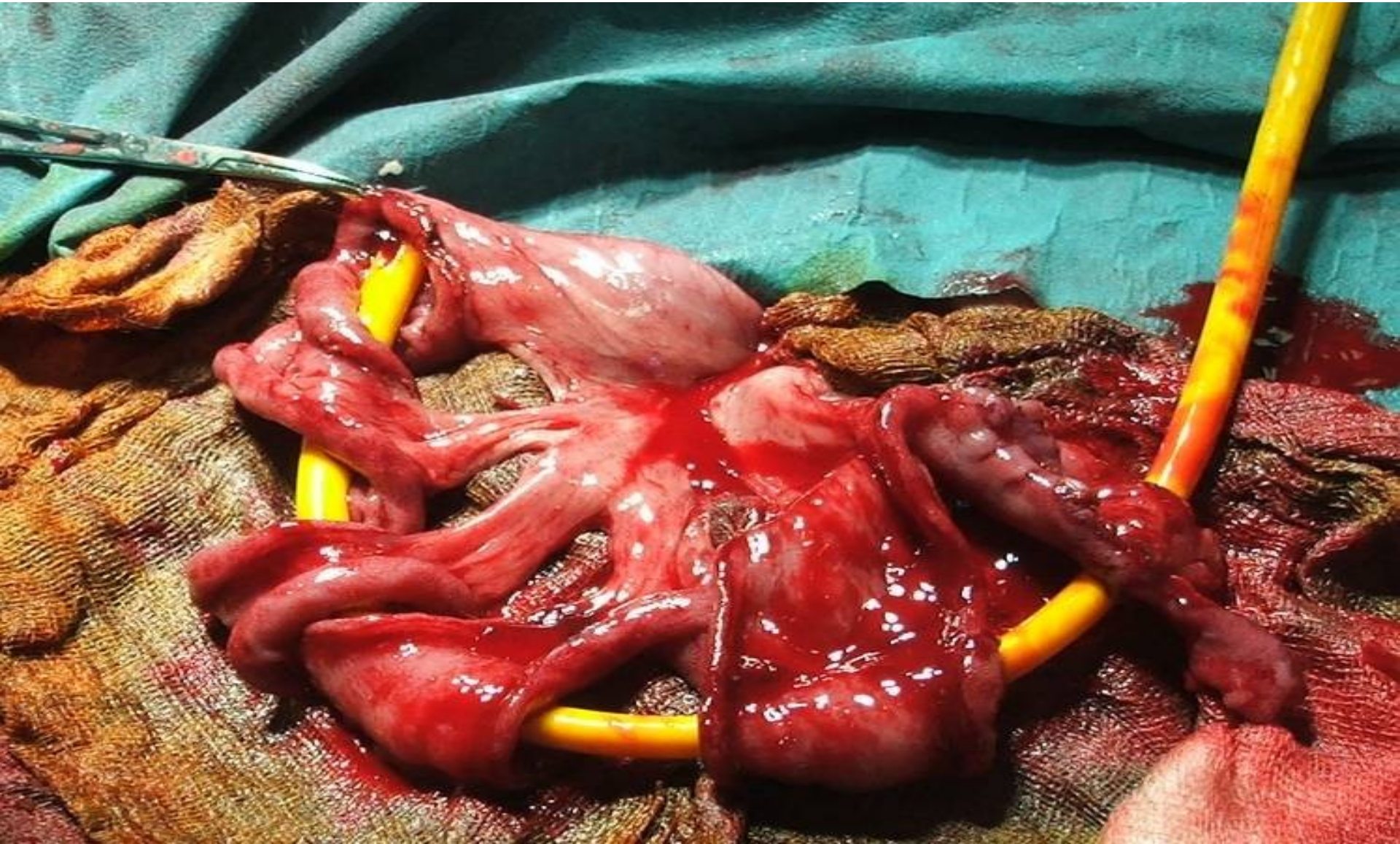
Spiral incision



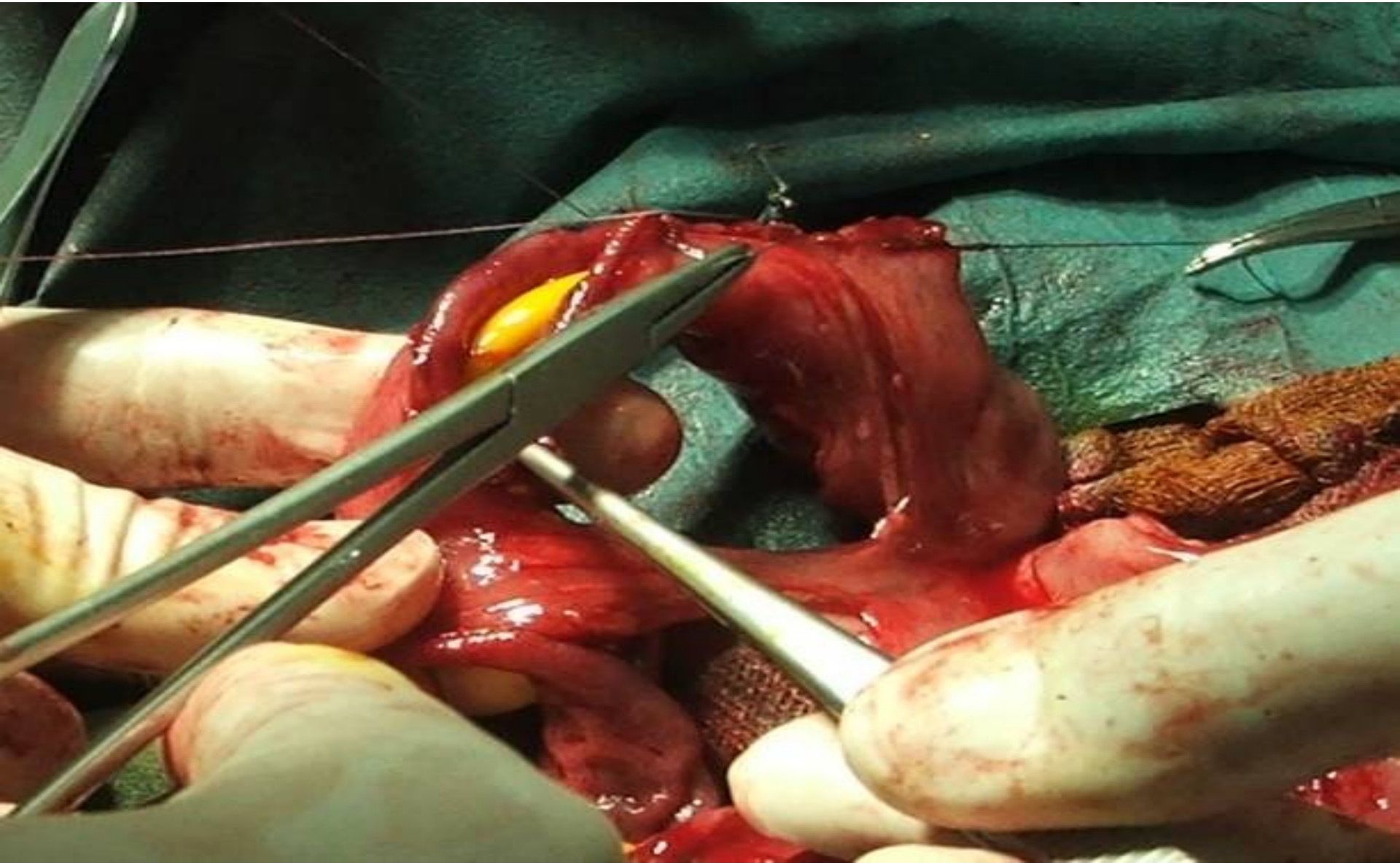
Spiral incision + incision on the mesentery



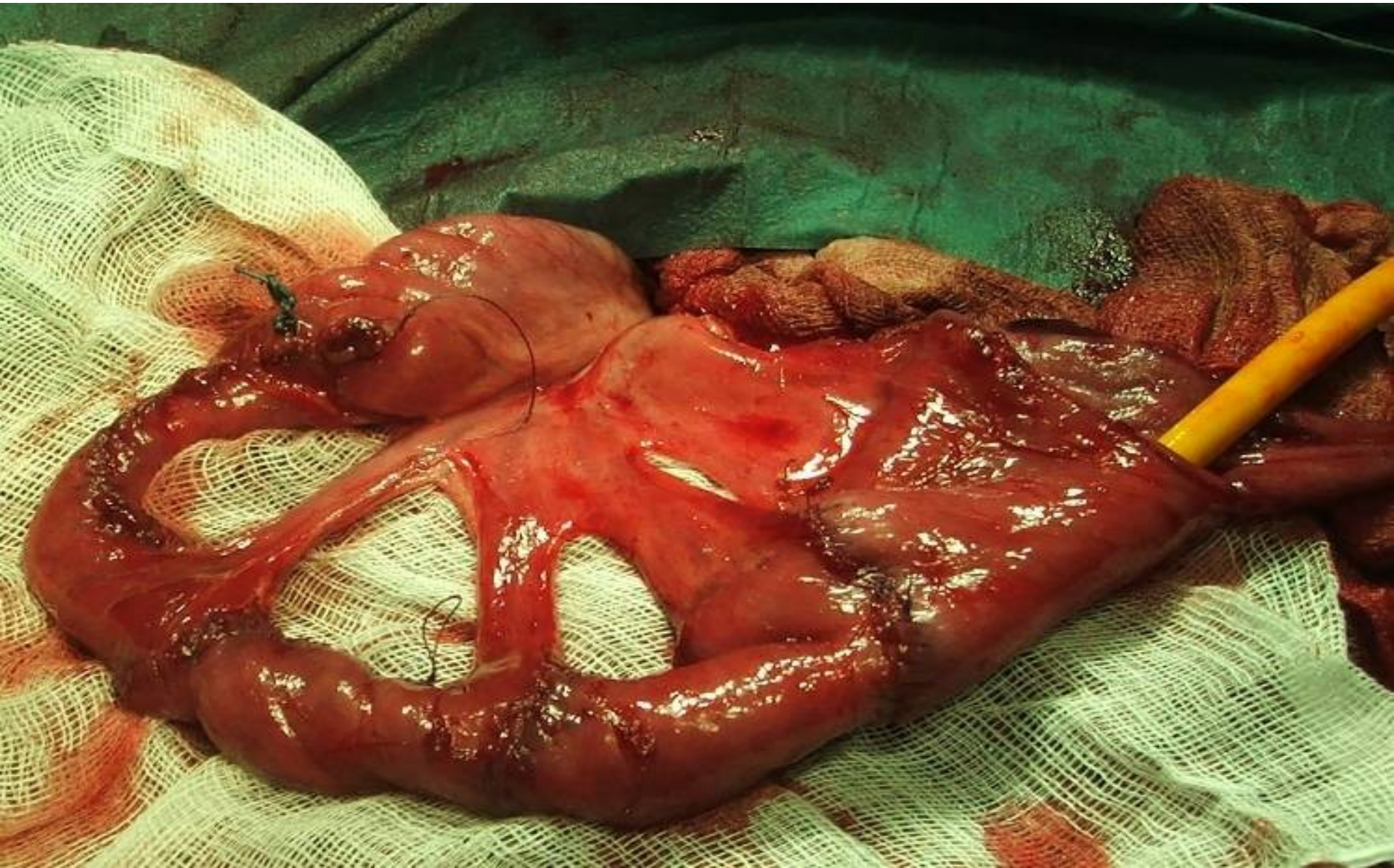
Retubularisation over a catheter



suture line in one layer

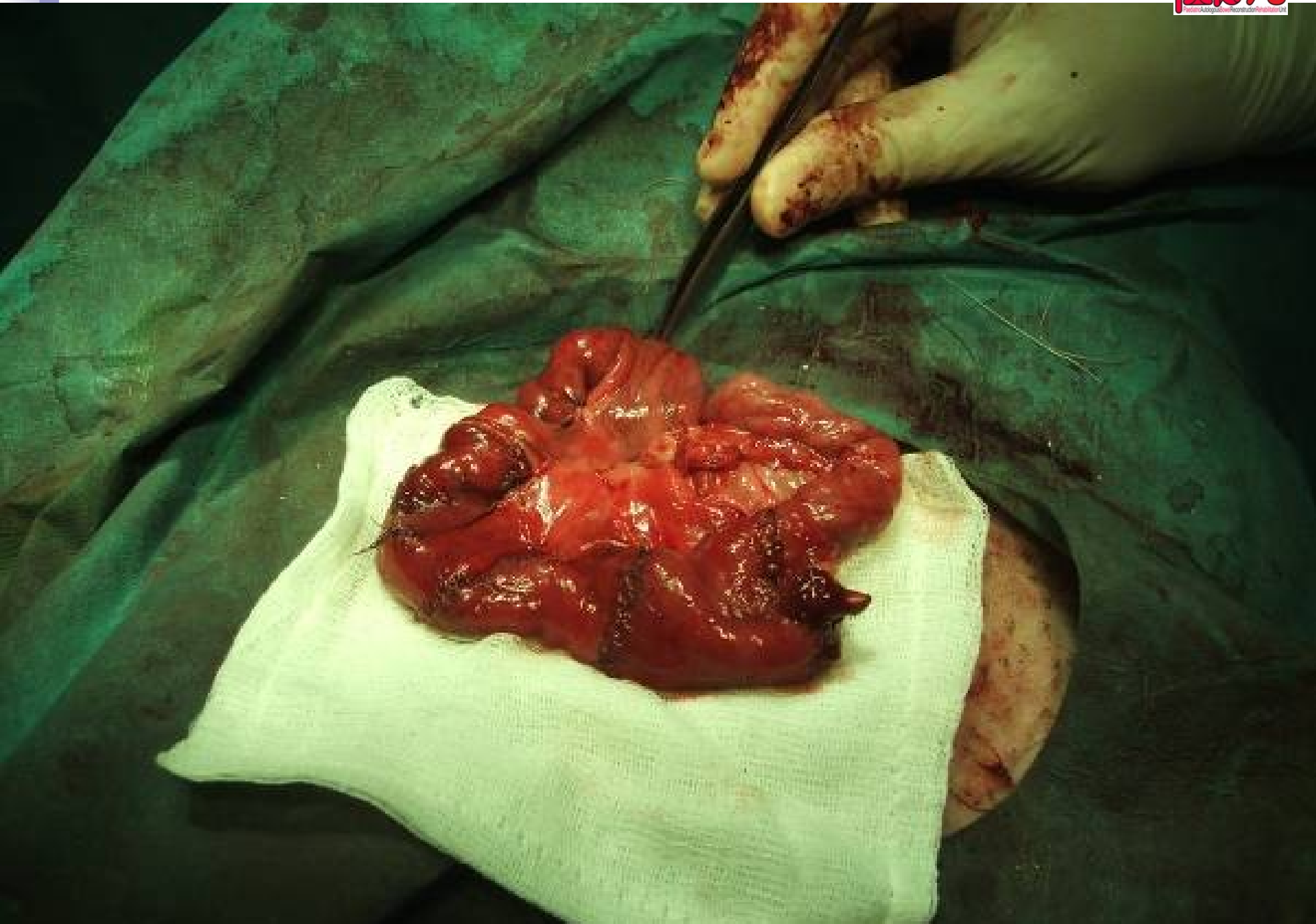


The tailored and lengthened segment





- The length increased from 15 cm to 30 cm
- The diameter reduced from 4 cm to 1 cm



Well on the 7-th postop day....

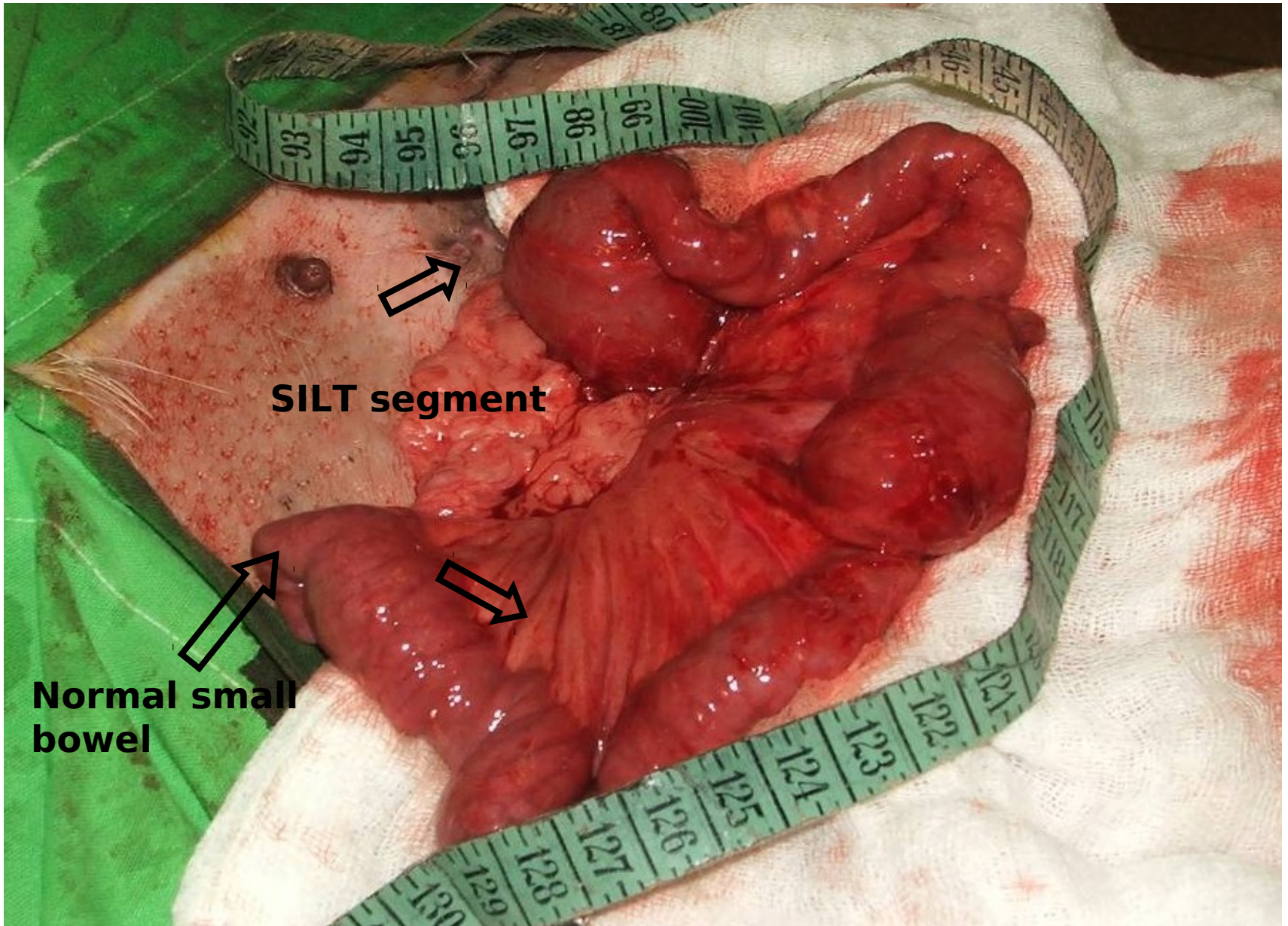


postop care:

- Painkillers, antibiotics
- drinking water with sugar on the 1st and 2nd postop day
- soup and diluted baby formula on the 3-4th day
- Turmixed normal food after the 5th postop day

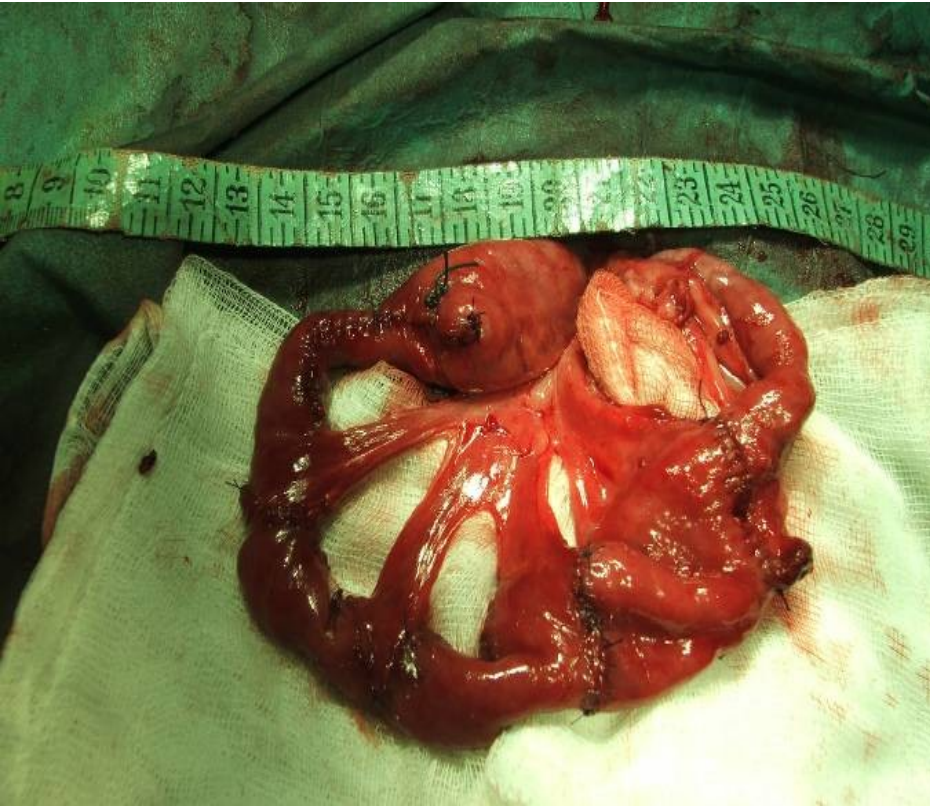
Exploration 6 weeks later



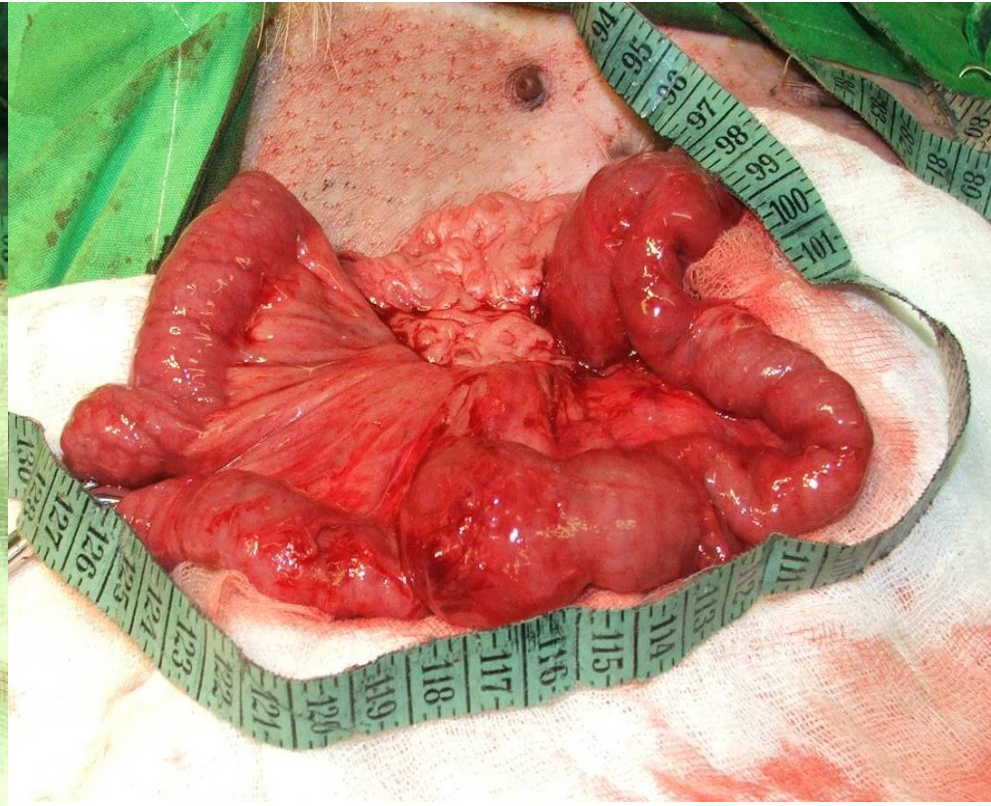


SILT segment

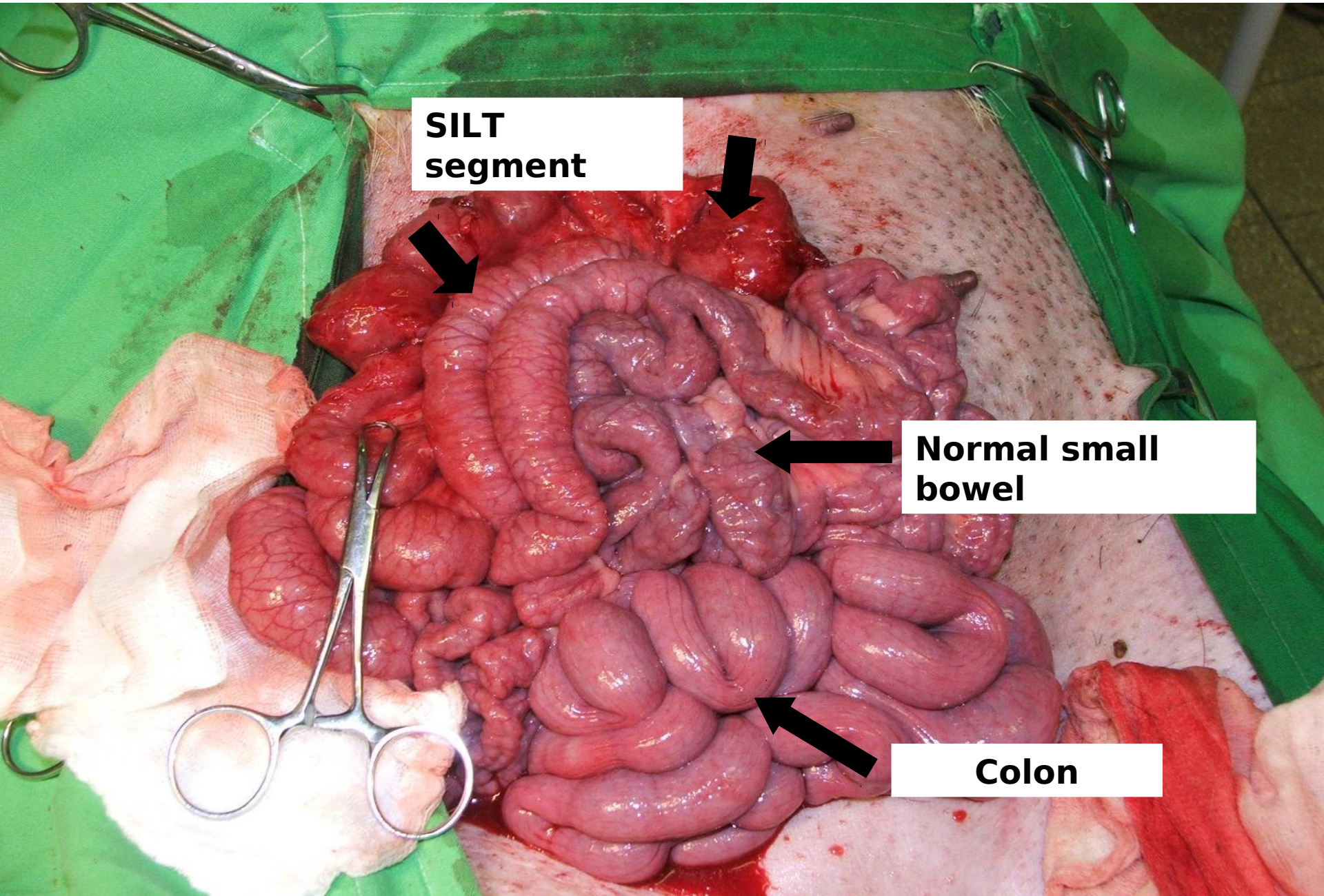
**Normal small
bowel**



SILT



SILT after 6 weeks



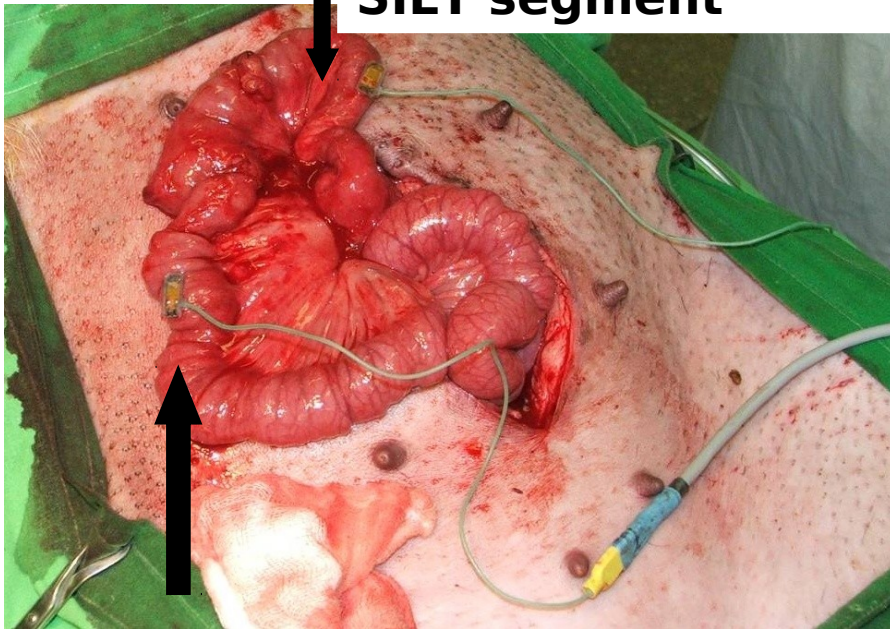
**SILT
segment**

**Normal small
bowel**

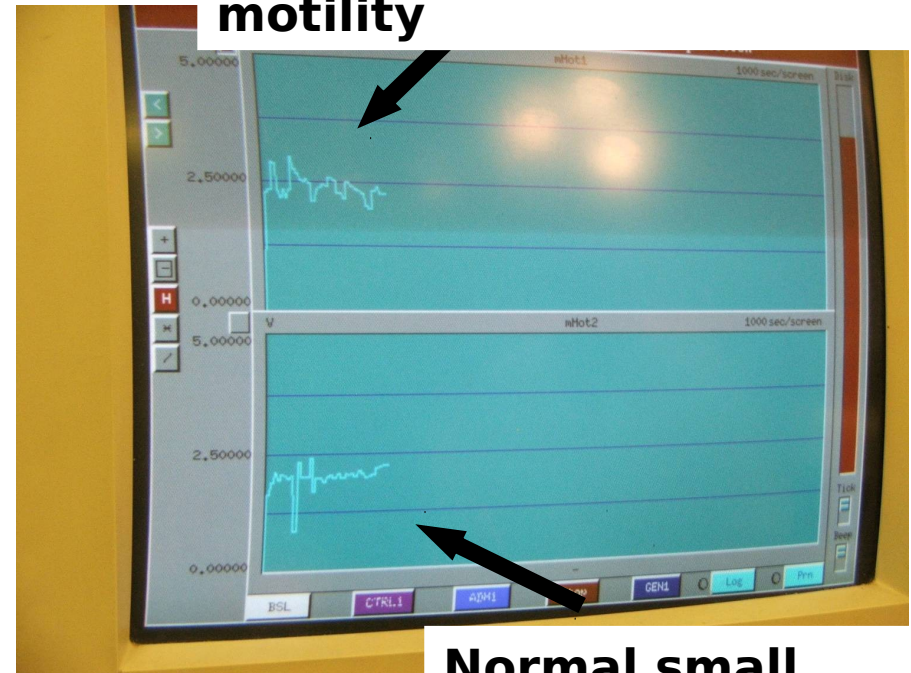
Colon

Motility of SILT segment compared to normal small bowel

Transducer on the SILT segment



SILT segment shows motility



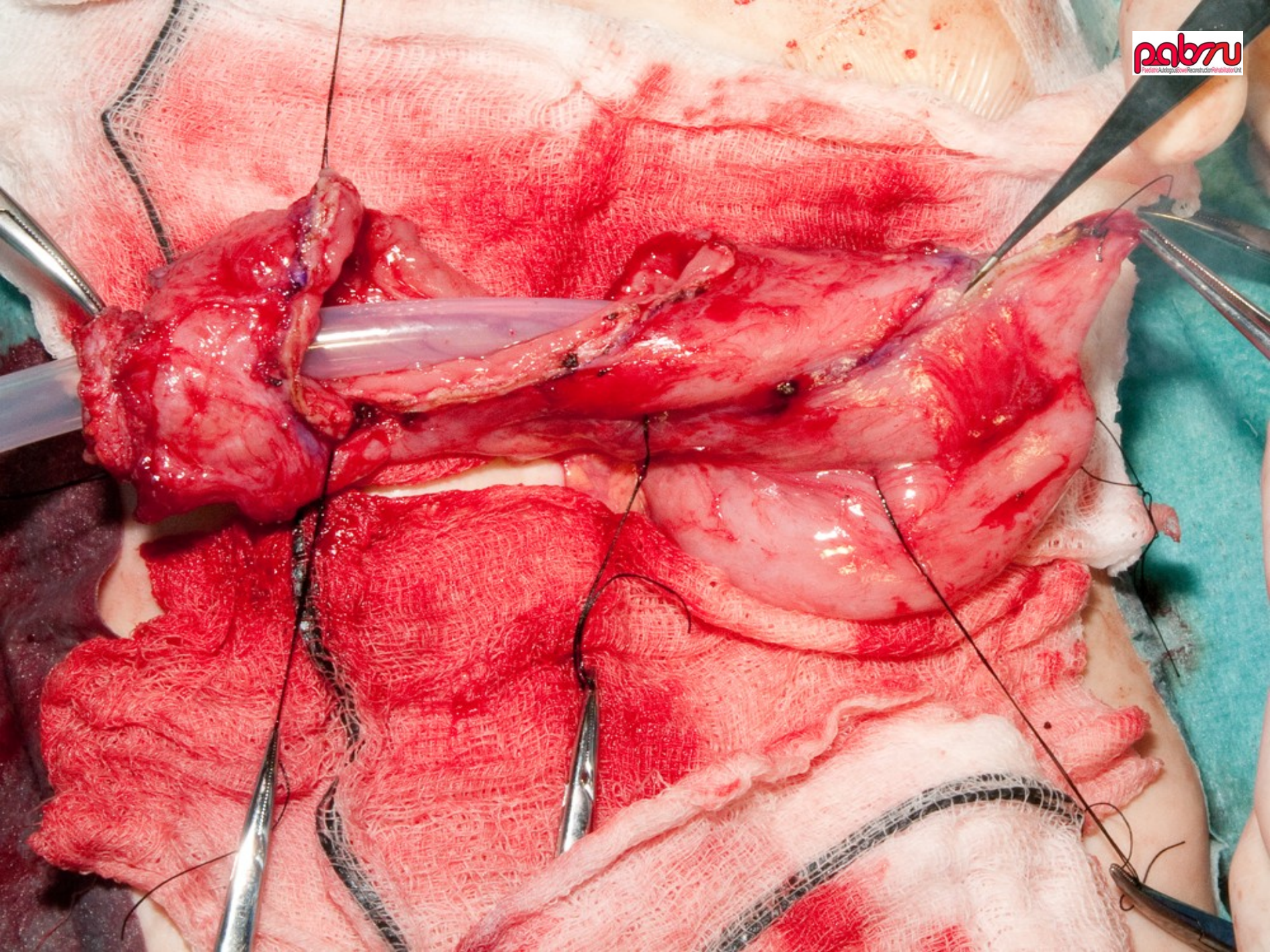
Normal small bowel

Transducer on the normal small bowel

Strain gauge transducers were sutured to the bowel wall, the signals were recorded by computerized data-acquisition system (Haemosys 1.17; Experimetria Ltd).

AUGUST
2012





The National Adult Intestinal Failure & Rehabilitation unit

Morabito A

Carlson GL

Stevens P

Lall S



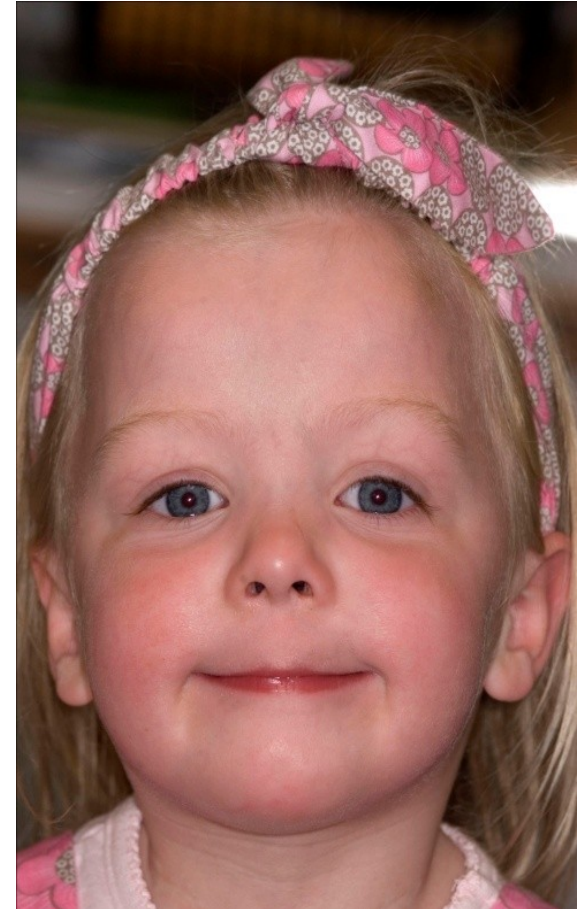
future

- Growing small bowel (MD Manchester University)
- Bowel expansion (MD Salford University)
- BSc Intestinal lengthening (Salford University)
- New drugs



The future is...

..autologous!



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