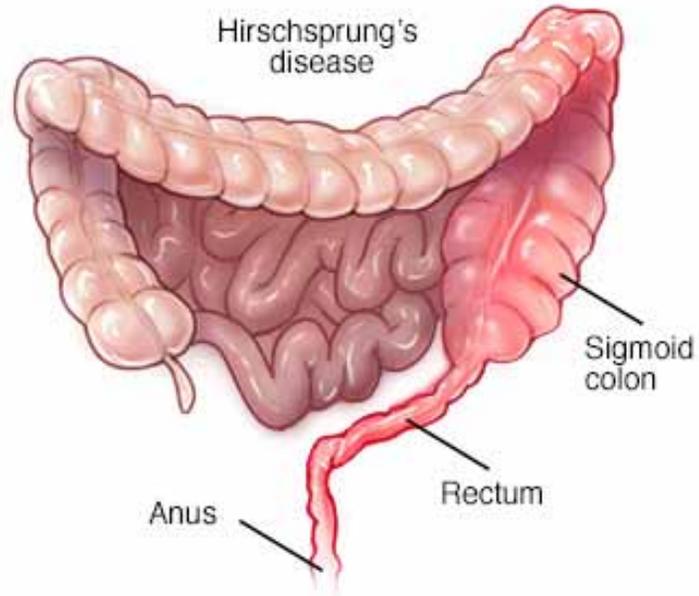


NEONATAL REPAIR
OF HIRSCHPRUNG'S DISEASE
WITHOUT LAPAROTOMY

DR KEVIN TEEROVENGADUM
PAEDIATRIC SURGEON

MEDICAL UPDATE UOM 07/09/2016



RECENT CASE REPORT

- **Malleck, 3 weeks old baby boy, sent from Comoros for severe constipation from birth**



PLAN OF CARE

- **Seen by pediatrician who asked for investigations, including:**
 - **1) Karyotyping to confirm Down's syndrome**
 - **2) Cardiac ultrasound**

- **Barium enema -> recto-sigmoid disease**
- **Rectal biopsy -> confirmed the aganglionosis**



- Barium enema -> recto-sigmoid disease
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DISCUSSION

THE USUAL MANAGEMENT IS A 3 STAGE REPAIR :

- 1) DEFUNCTIONING COLOSTOMY AT DIAGNOSIS
- 2) SURGICAL REPAIR (SWENSON/SOAVE/DUHAMEL)
NORMALLY AROUND THE AGE OF 6 MONTHS
- 3) REVERSAL OF THE COLOSTOMY 1-2 MONTHS
LATER

DISCUSSION

AFTER CAREFUL ASSESSMENT OF THE **PERI-OPERATIVE RISKS** AND BECAUSE IT WOULD HAVE BEEN DIFFICULT FOR MALLECK TO BE BROUGHT BACK TO MAURITIUS 2 MORE TIMES, IT WAS DECIDED TO CARRY OUT THE **REPAIR OF THE HIRSCHSPRUNG'S DISEASE IN 1 STAGE** CALLED SOAVE'S TRANS-ANAL PULL-THROUGH, **THE BABY DID NOT UNDERGO ANY LAPAROTOMY AND THE SICK RECTUM AND SIGMOID COLON WERE REMOVED ENTIRELY THROUGH THE ANUS.**

DECISION MAKING –
ANESTHETIST (DR SUBIR) /
PEDIATRICIAN (DR JAGATSINGH)

PROS

- **No colostomy and its associated morbidity**
- **Reduced hospital stay/ good parental acceptance**
- **Barium enema : a classic recto-sigmoid disease (ie not an extra-long segment disease)**

CONS

- **Increased risk of post-operative enterocolitis because dilated colon has not been left to decompress prior to surgery.**
- **Down's syndrome: increased risk of infection / prolonged surgery**

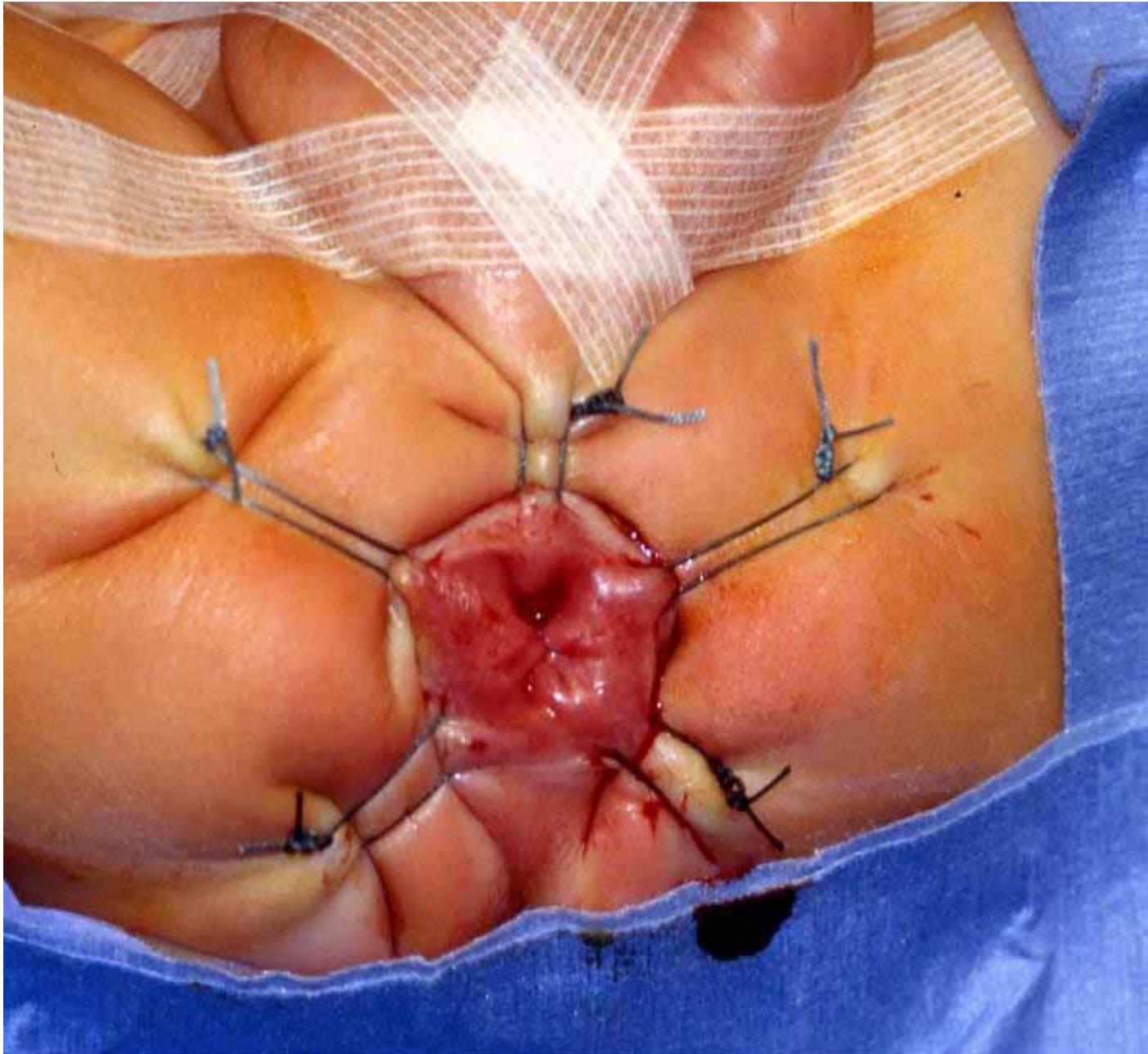


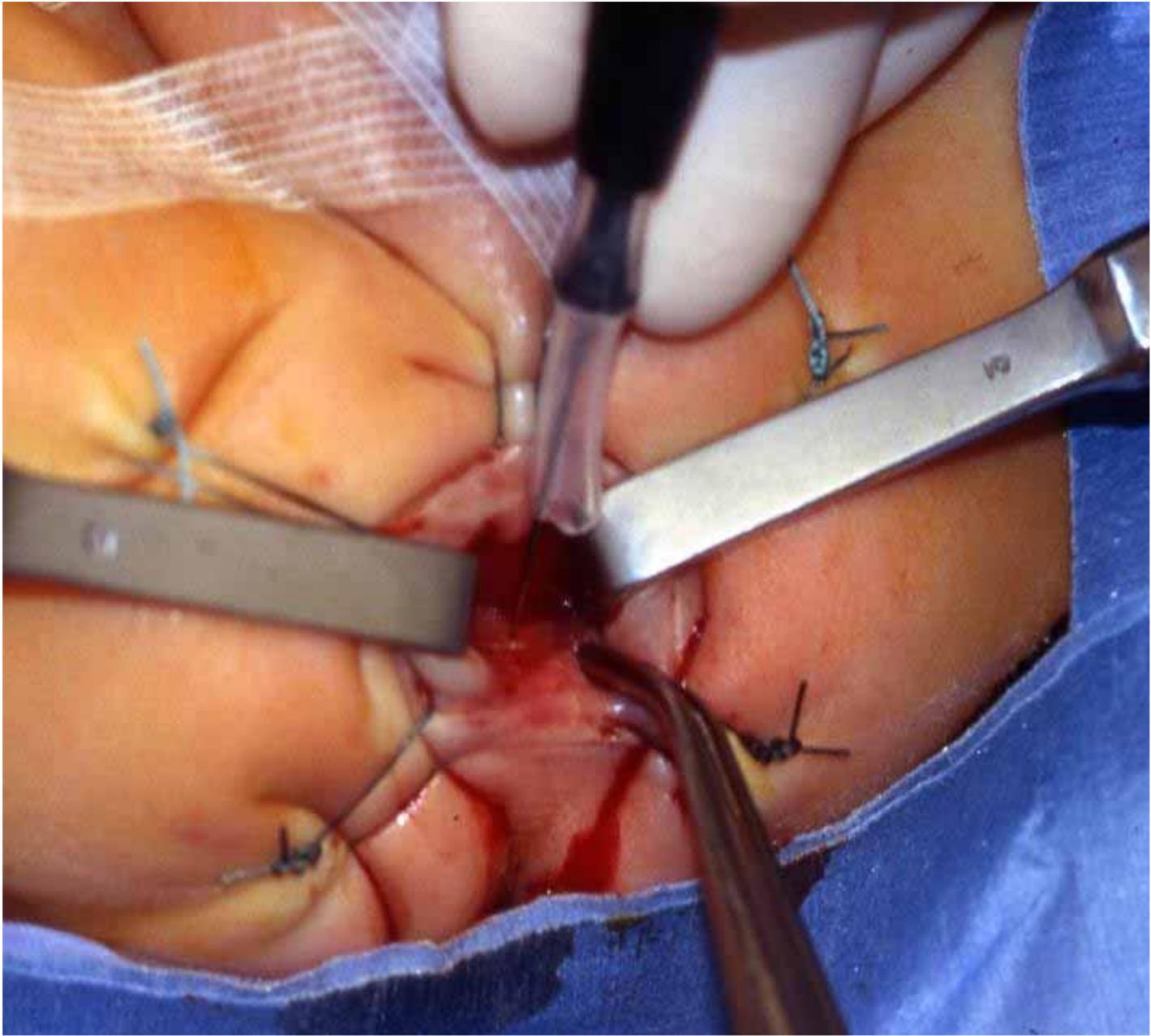
OPERATIVE
VIEW:

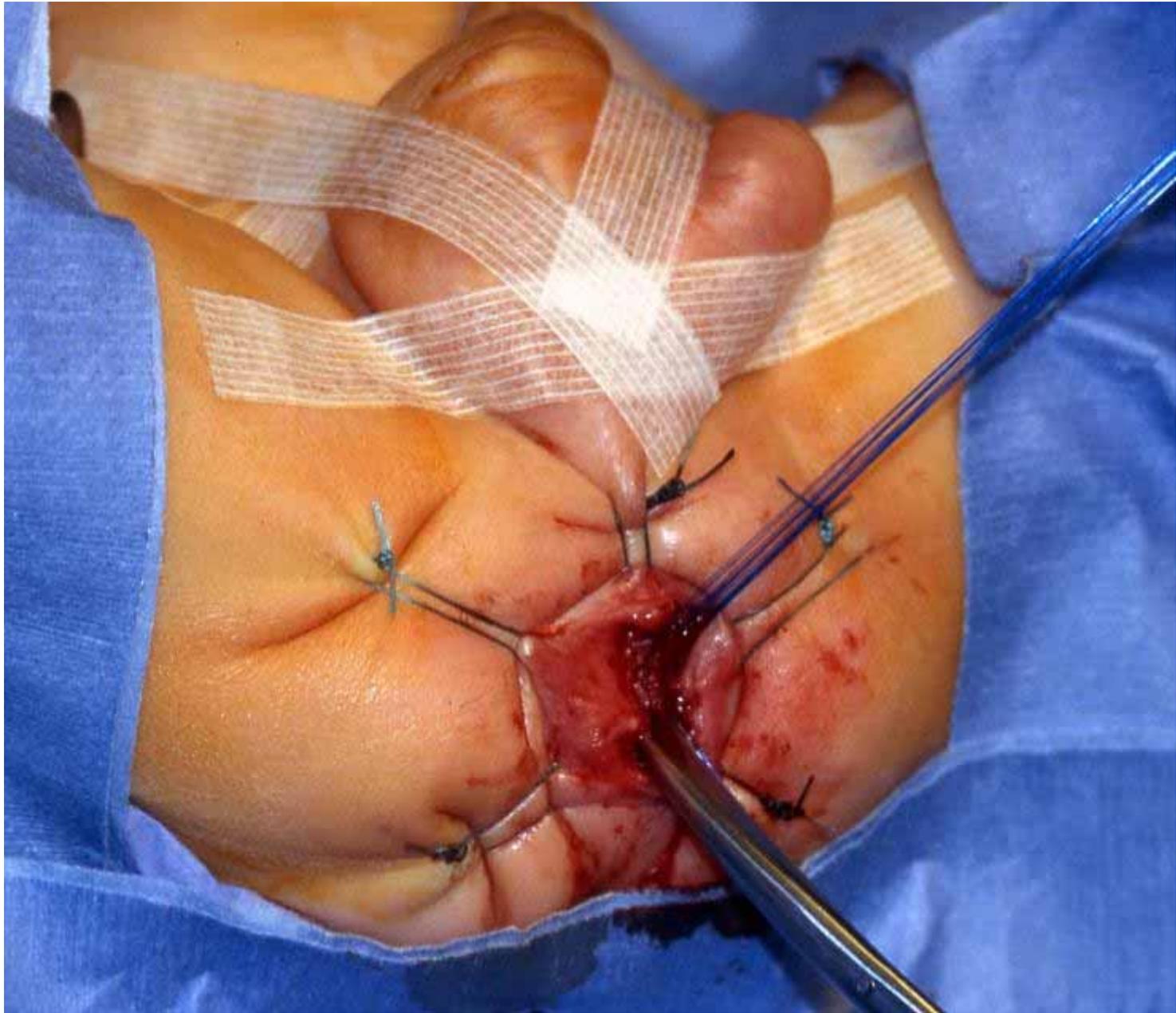
NORMAL COLON/

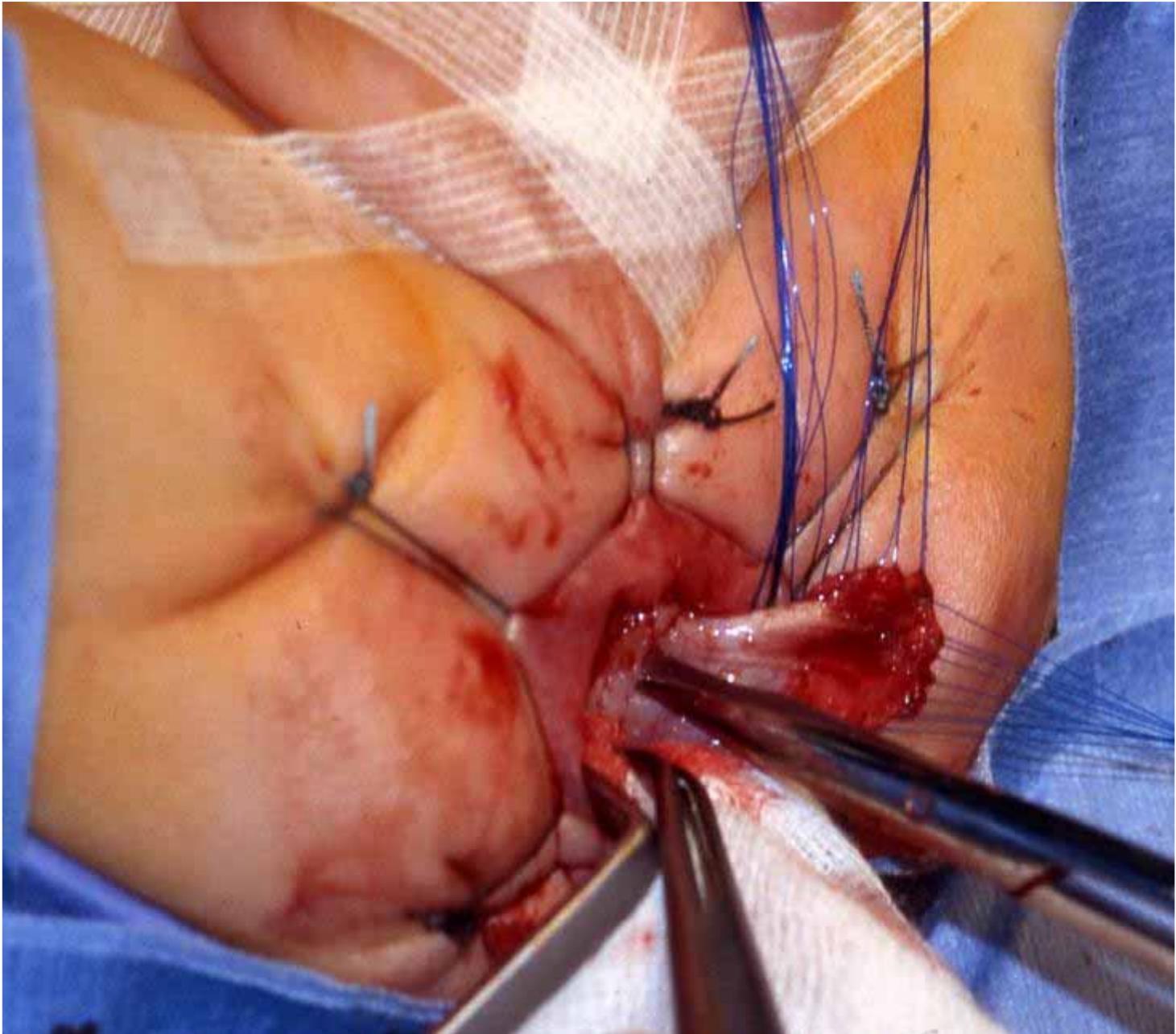
TRANSITIONAL
ZONE/

AGANGLIONIC
SEGMENT

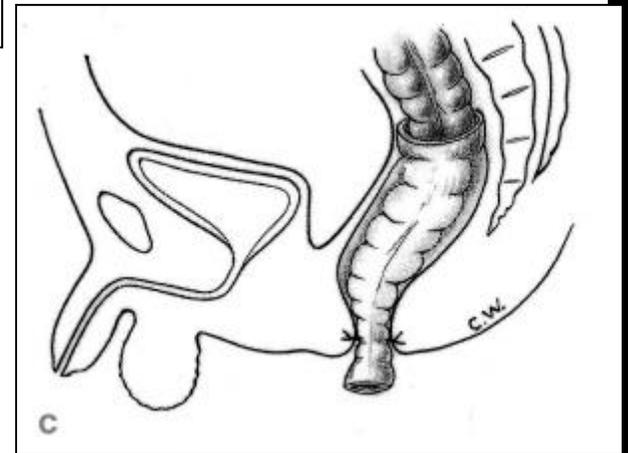
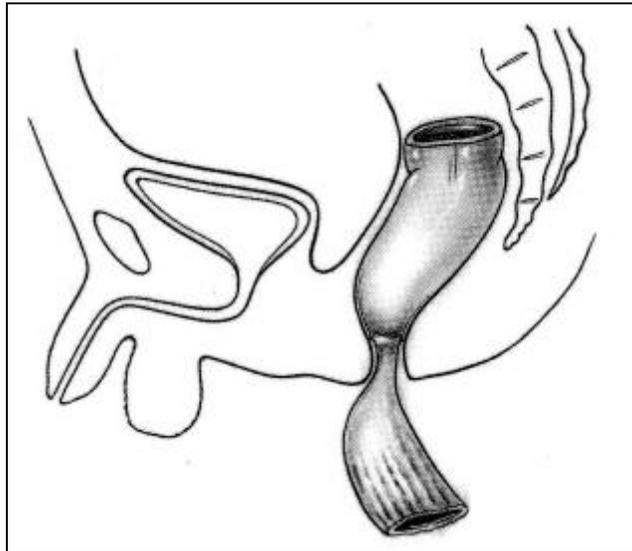
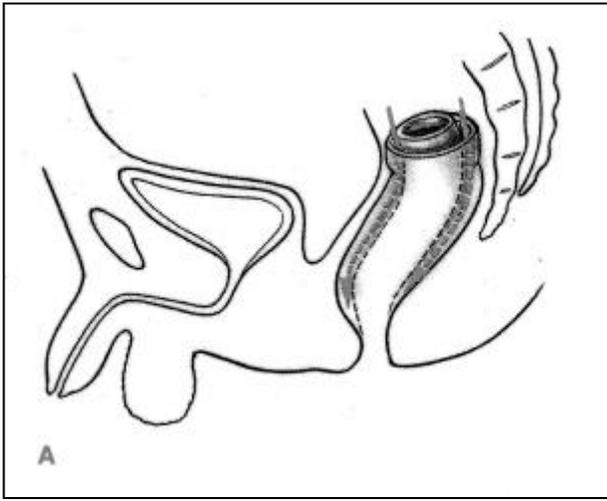


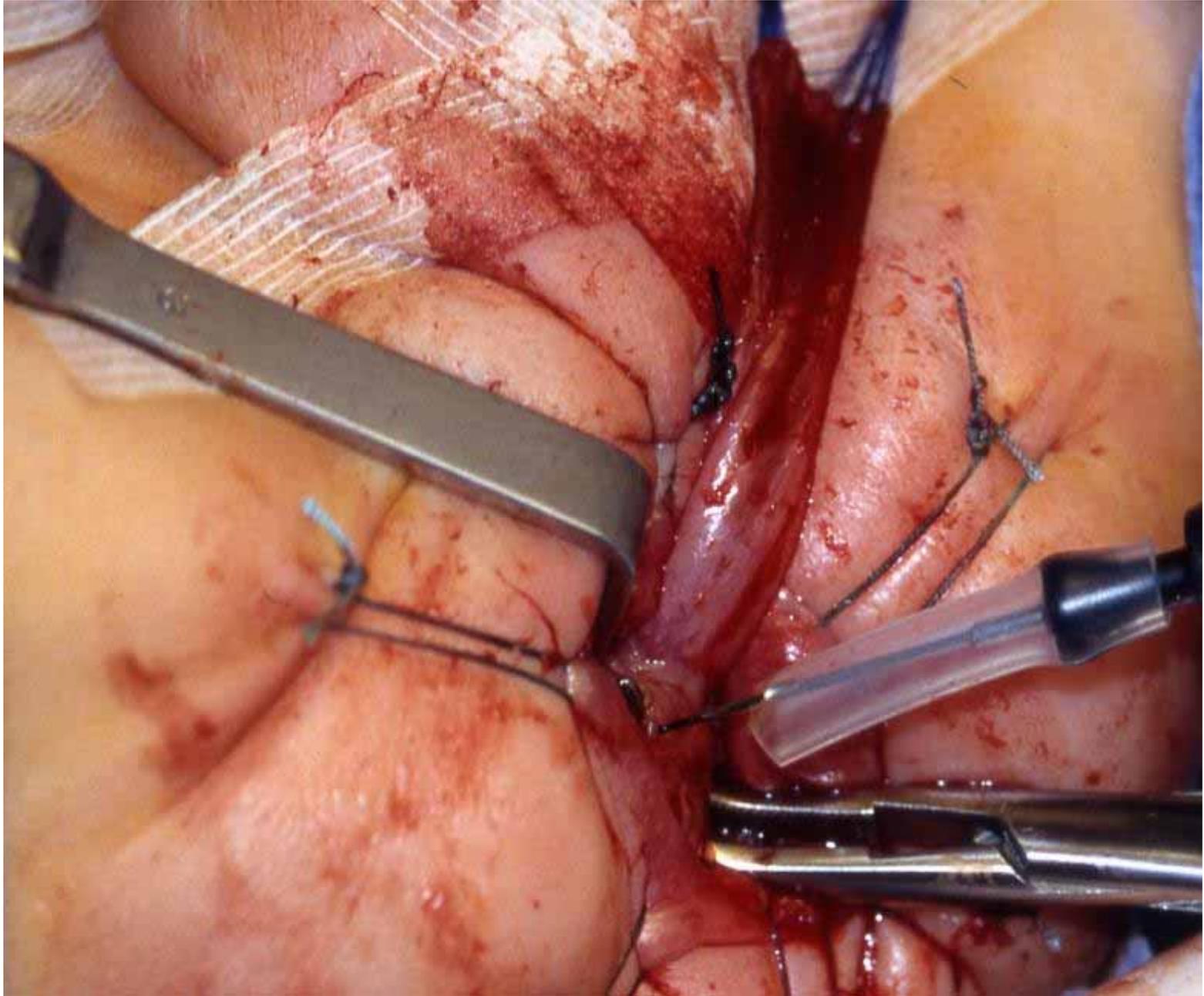


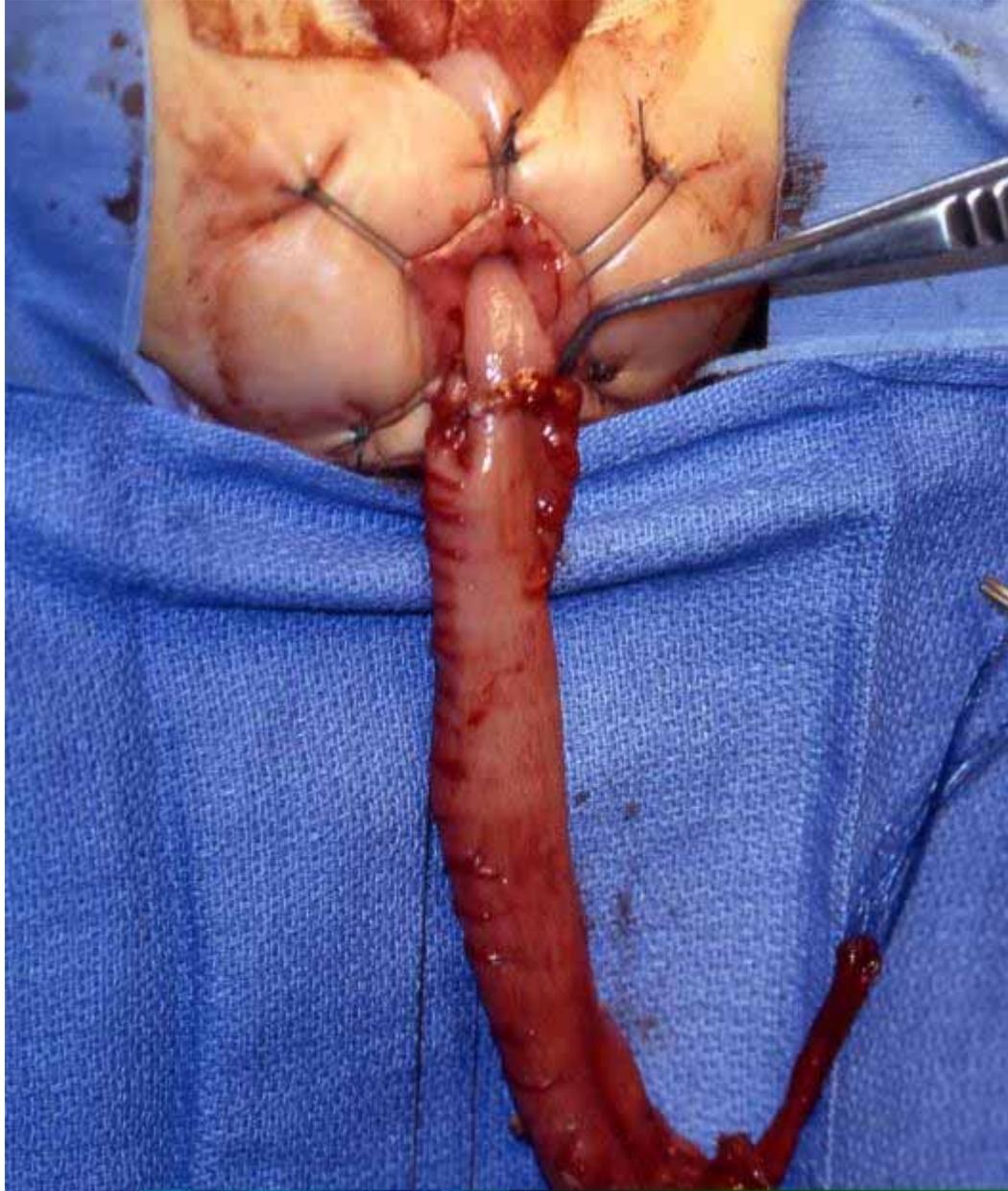


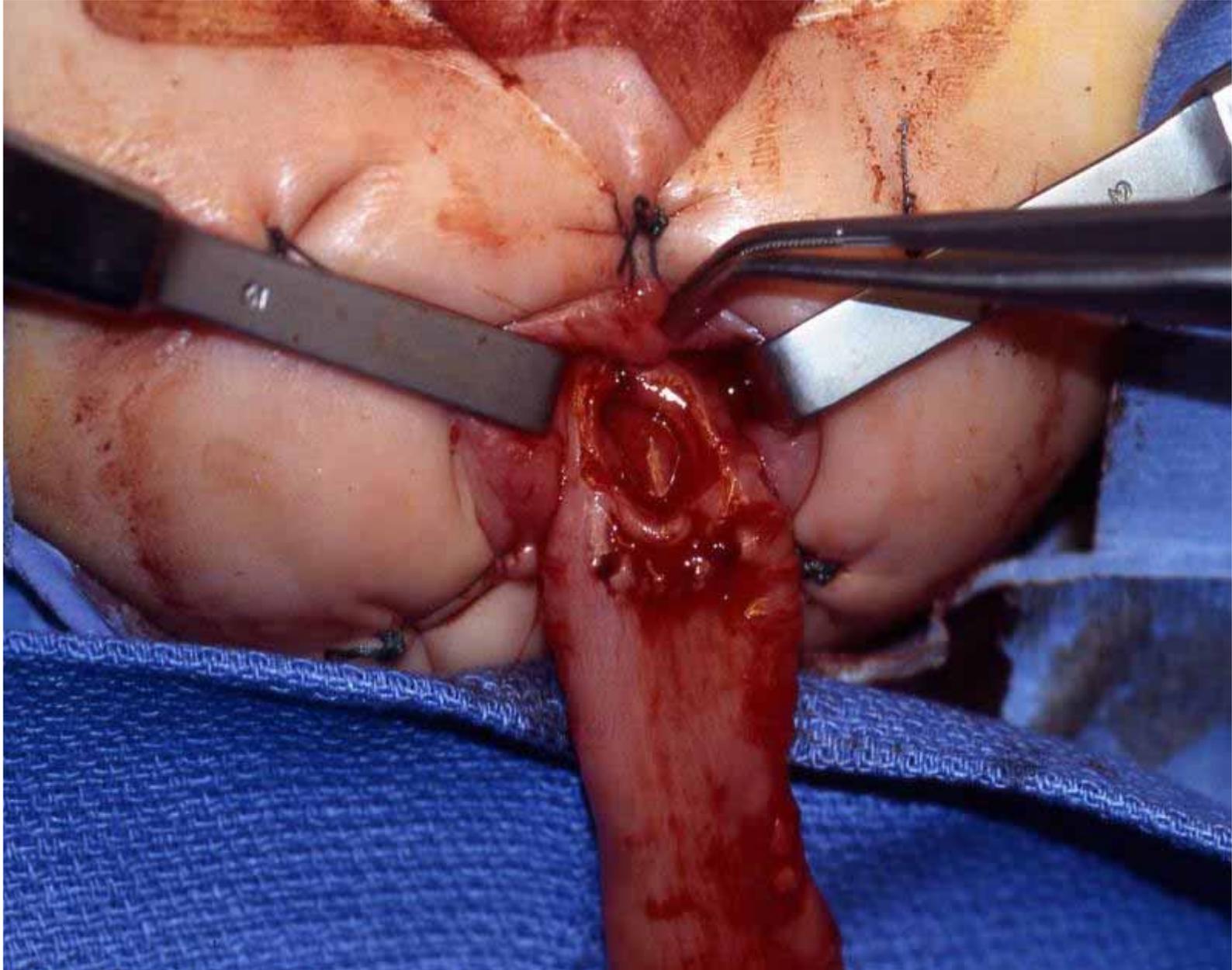


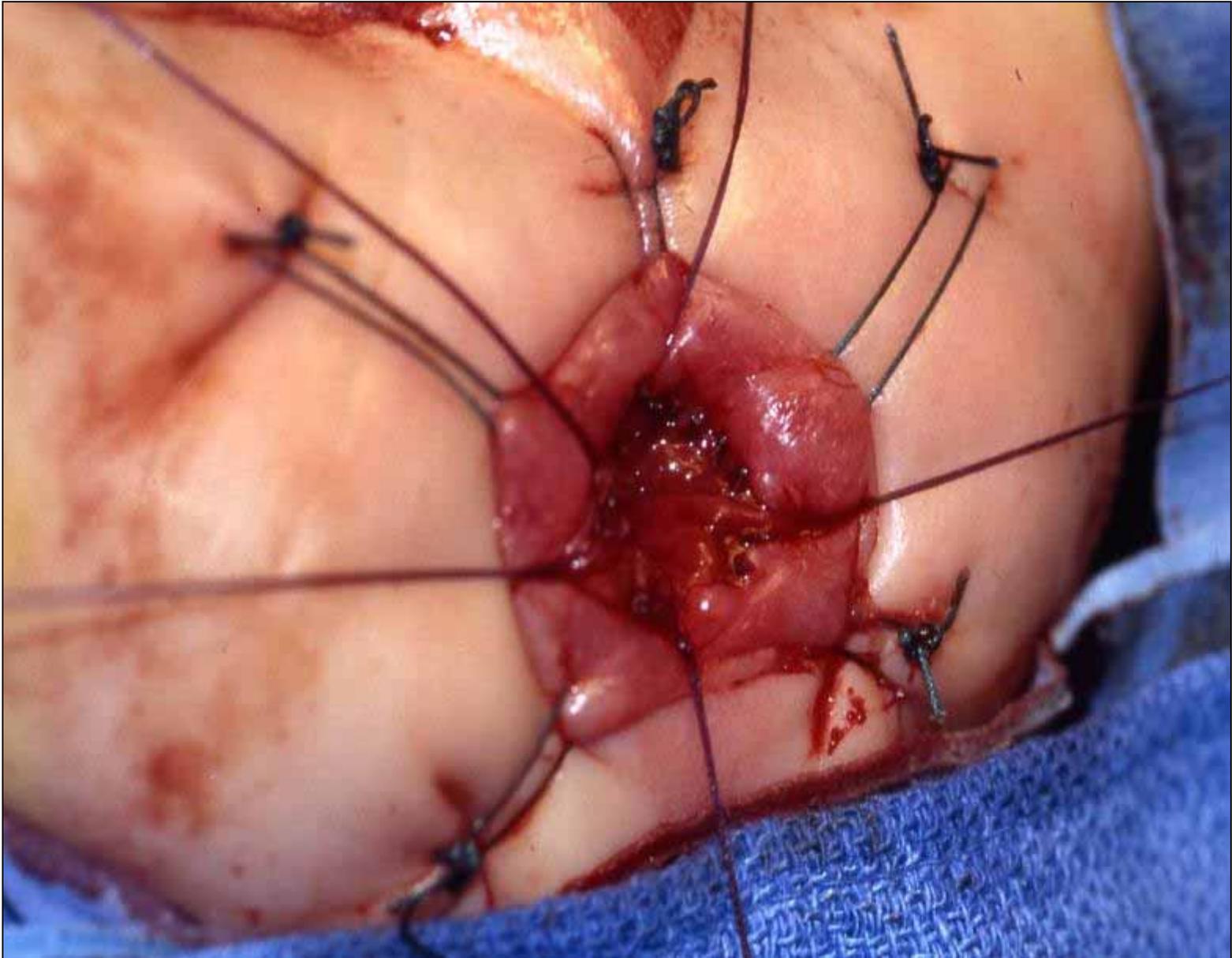
SOAVE













POST-OP

1) Immediate : extubated on table, passed stools on Day2, on full feeds on Day4, discharged Day5 post-op

2) Intermediate : had a prophylactic anal dilatation at Day15 just before flying back to Comoros,

recent news are good : 5 stools/day, no need for laxatives / baby is thriving well 2 months after surgery

3) Long-term : to monitor continence