

**The 7<sup>th</sup> Annual Meeting of the S.I.V.I.**

**Naples, 2-3 June 2006**

# HIRSCHSPRUNG DISEASE

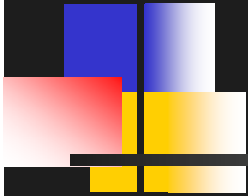
**Luciano Mastroianni**

**S.O.D. Chirurgia Pediatrica**

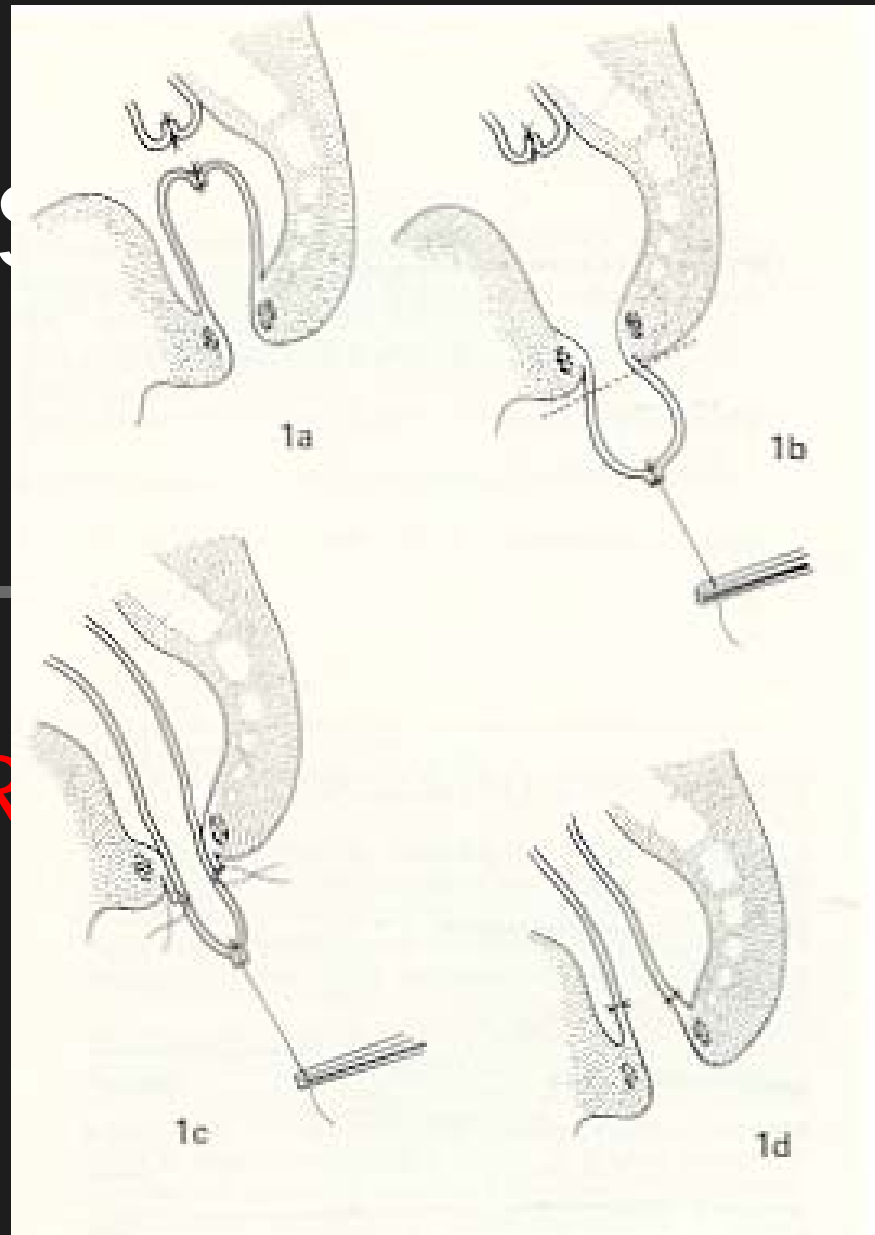
**Presidio Ospedaliero di Alta Specializzazione "G. Salesi"**

**Ancona**





1° INTER



HROUGH

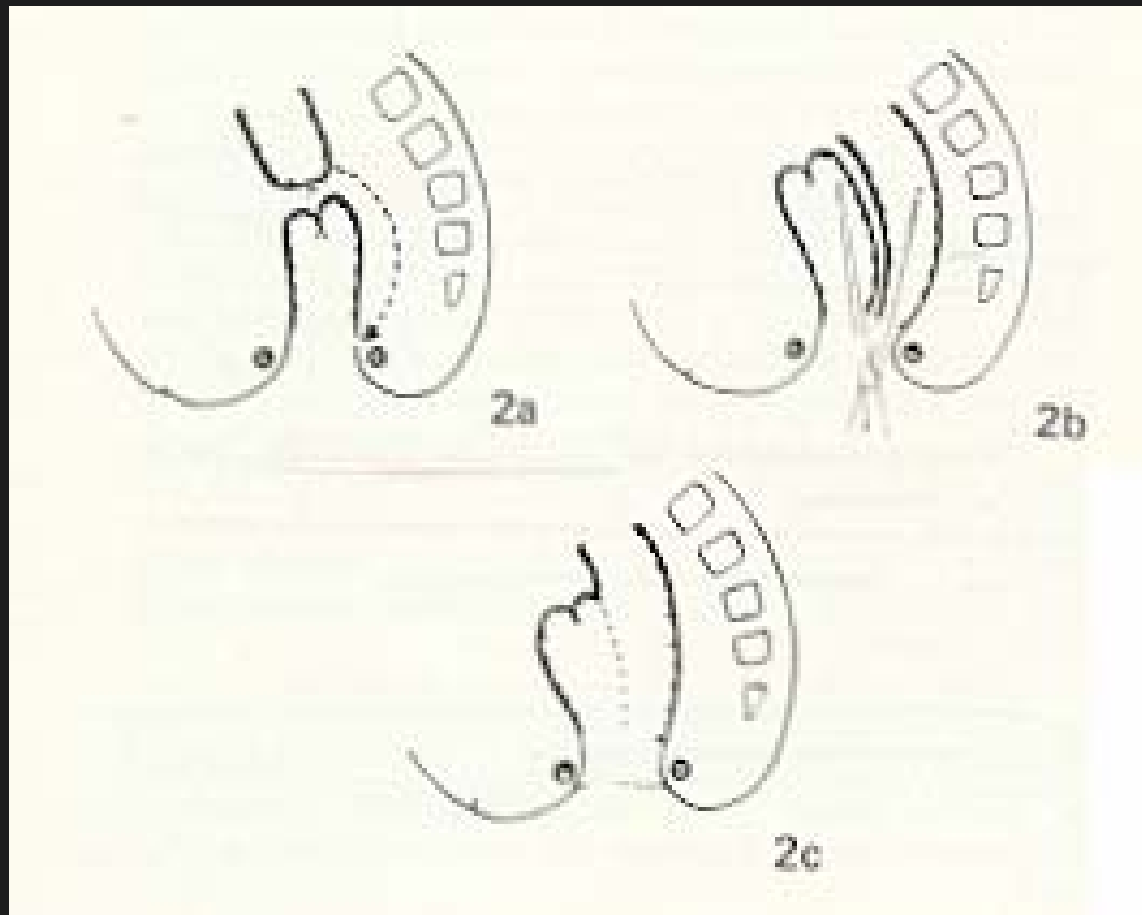
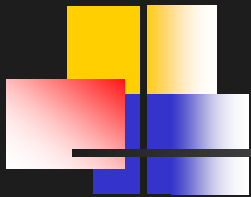


# Aganglioneosis Management

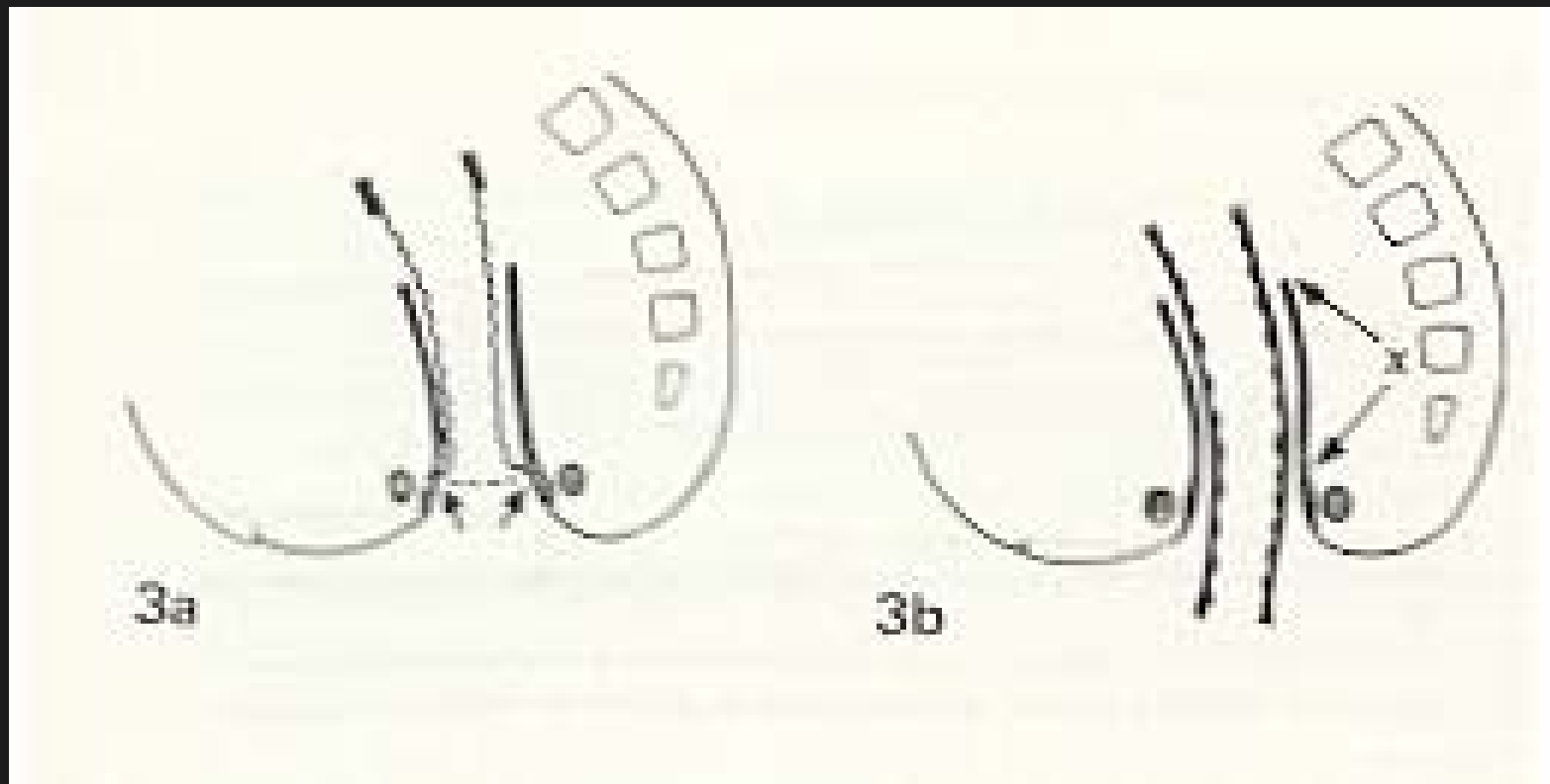
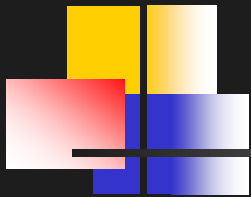
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In the last 60 years the Hirschsprung's disease treatment has evolved and different surgical techniques have been proposed

# Duhamel/Martin 1956 /67



# Soave 1964

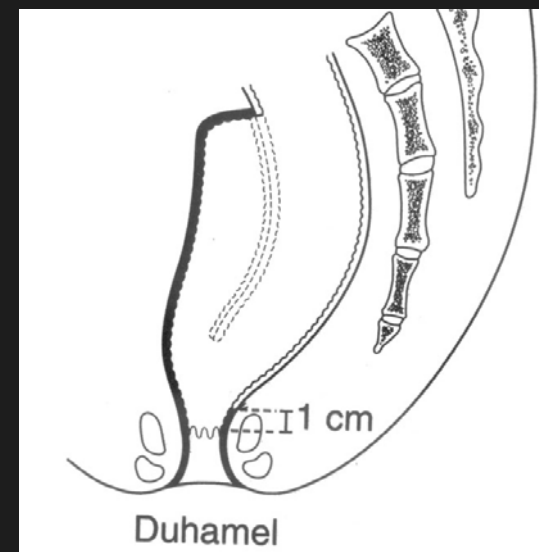
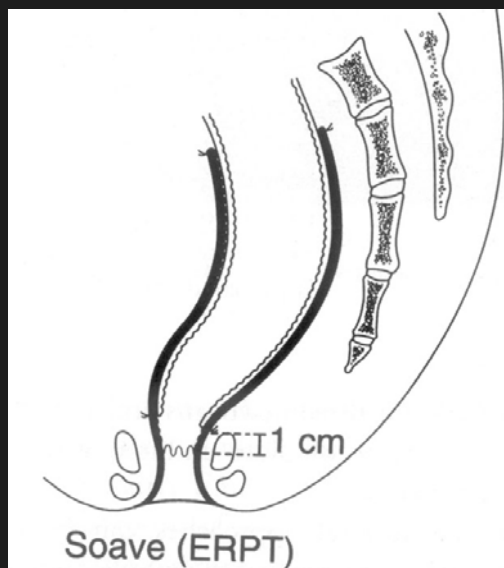
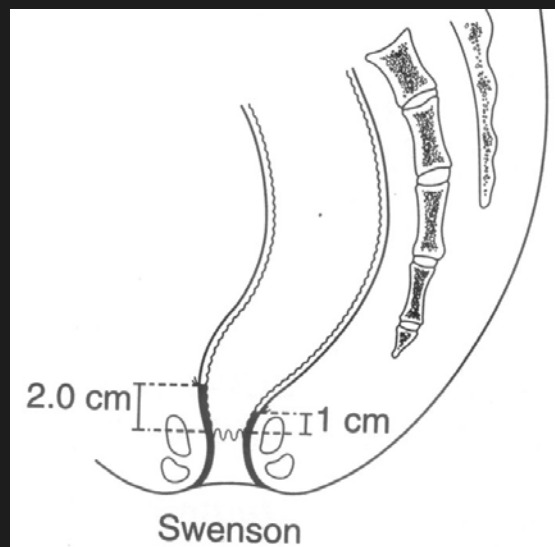
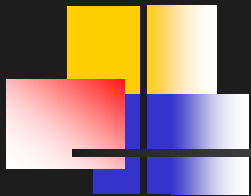




Stockman PT, Philippart AI 1998.

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*“the current goal of all procedures for hirschsprung’s disease is to place normally functioning ganglionated intestine within 1 cm. of the anal verge”.*





# Treatment options

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## Staged Treatment

- Nursing/Colostomy 2-13% complications
- Pull-Through after 6-12 months

## Primary Treatment

- Pull-through at diagnosis
- Even in the newborn



# Treatment criteria selection

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Parameters

Primary Treatment

Staged treatment

|                          |                        |                     |
|--------------------------|------------------------|---------------------|
| <b>Disease</b>           | <b>Confirmed</b>       | <b>No confirmed</b> |
| <b>Aganglionicosis</b>   | <b>Recto-sigmoid</b>   | <b>Long segment</b> |
| <b>Nursing</b>           | <b>Successful</b>      | <b>Unsuccessful</b> |
| <b>Enterocolitis</b>     | <b>Absent/resolved</b> | <b>Persistent</b>   |
| <b>Bowel dilatation</b>  | <b>Minimal</b>         | <b>Marked</b>       |
| <b>Nutritional State</b> | <b>Normal</b>          | <b>Compromised</b>  |
| <b>Gestational Age</b>   | <b>A term</b>          | <b>Pre-term</b>     |



# Primary Treatment

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- SOAVE So H.B et al. 1980
- SWENSON Carcassonne et al. 1982
- DUHAMEL Samuel M.,Freeman N.V. 1994



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**Laparoscopic Duhamel pull-through procedure for Hirschsprung's disease in childhood**

Smith B.M., Steiner R.B., Lobe T.E.

*J Laparosc Surg* 4:273-276, 1994

**Primary laparoscopic pull-through for Hirschsprung's disease in infants and children**

Georgeson K.E., Fuenfer M.M., Hardin W.D.

*J Pediatr Surg*, 30:1017-1022, 1995

**Laparoscopic Swenson pull-through: a comparison with the open procedure**

Curran T.J., Raffensperger J.G.

*J Pediatr Surg* 31:1155-1156, 1996



In the first half of 1990

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Endorectal Pull-through

Age 3 – 6 months



# Now

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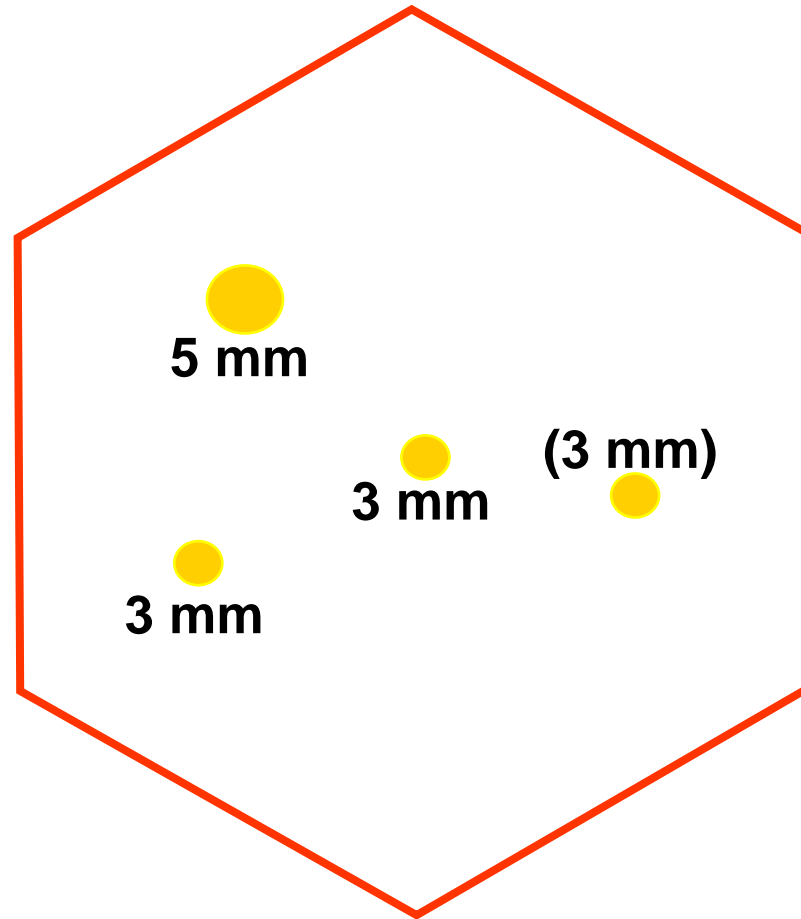
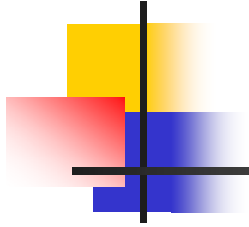
The ideal treatment of classic  
Hirschsprung disease is the Laparoscopic  
assisted endorectal Pull-through.

To be performed in the newborn or  
infant.

Without colostomy.

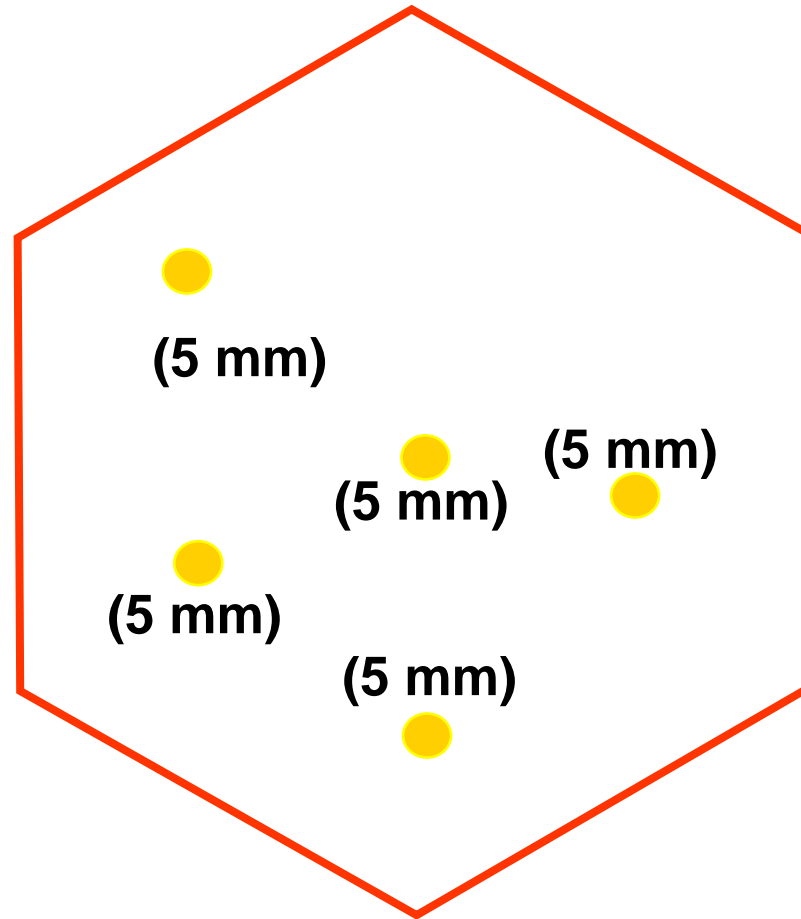
# Technique

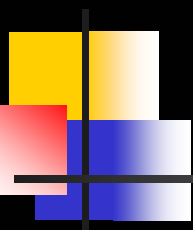
## *4 trocars*



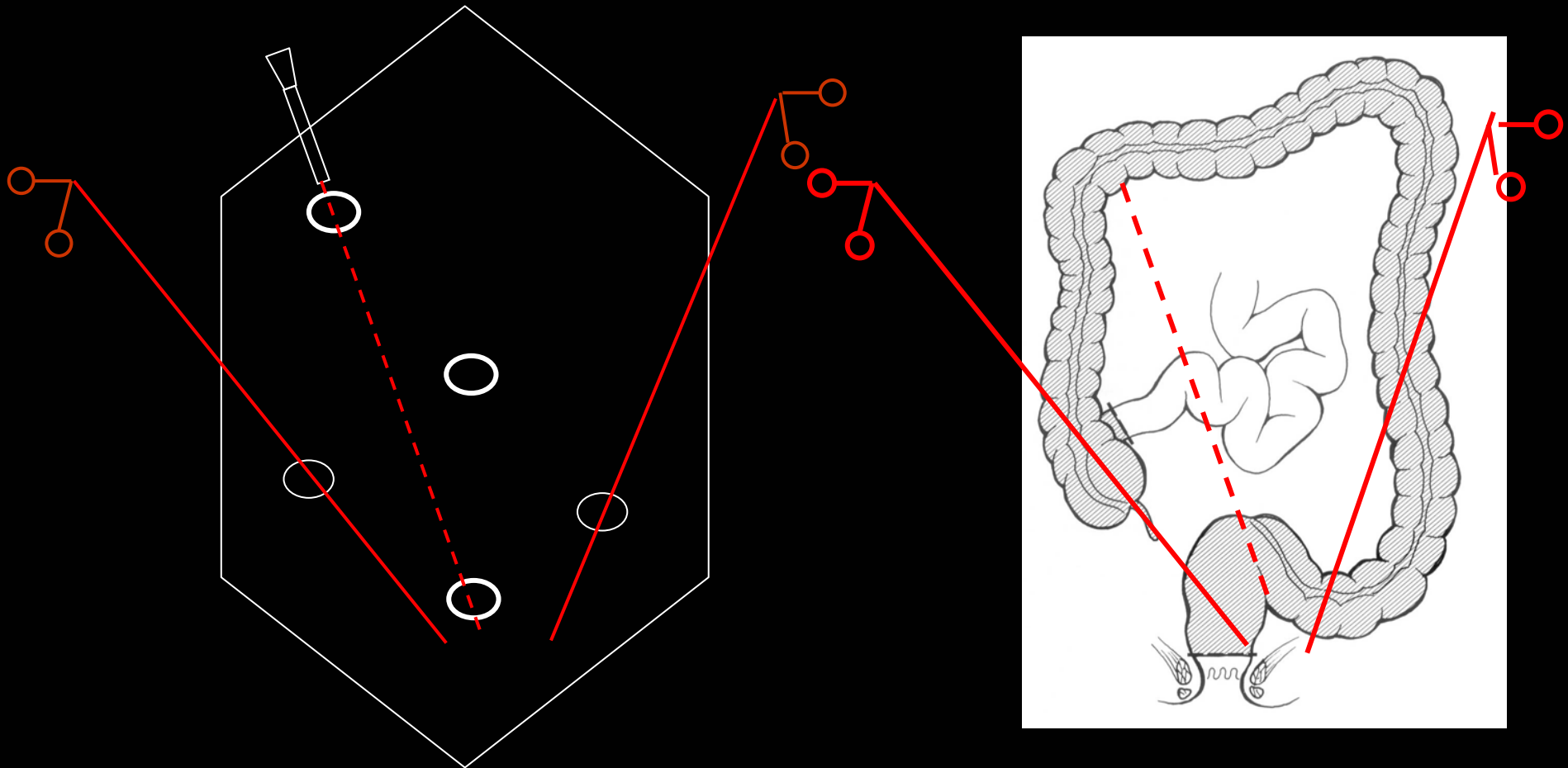
# Technique

## *5 trocars (long segment)*

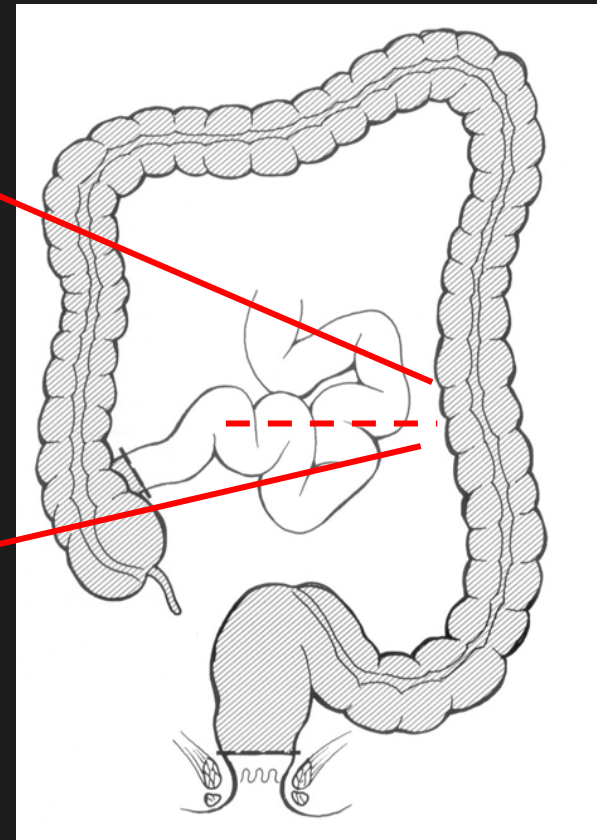
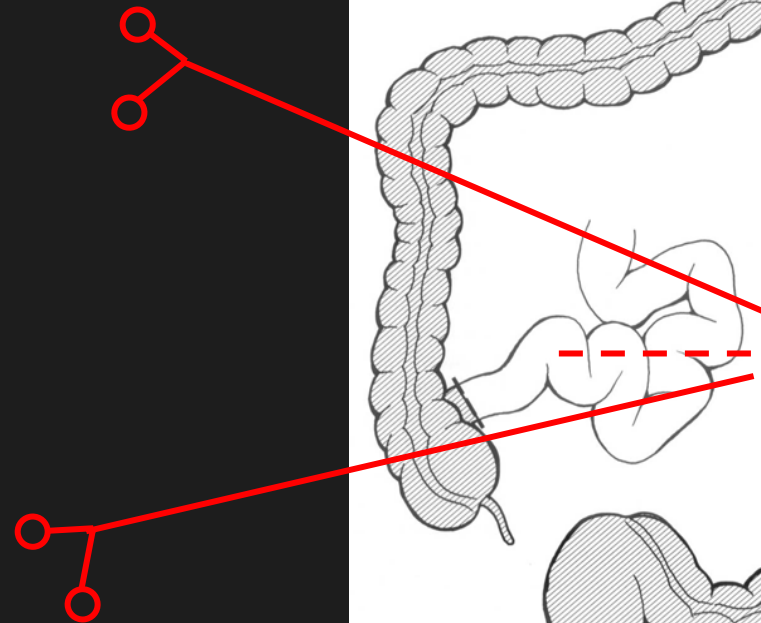
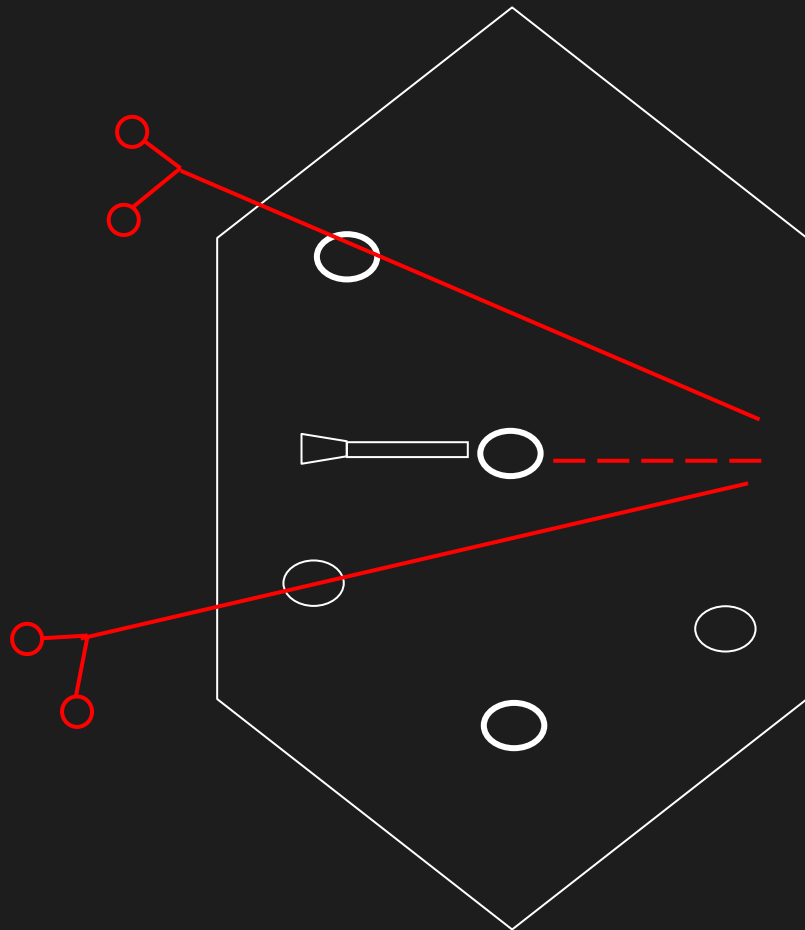
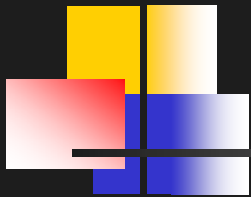




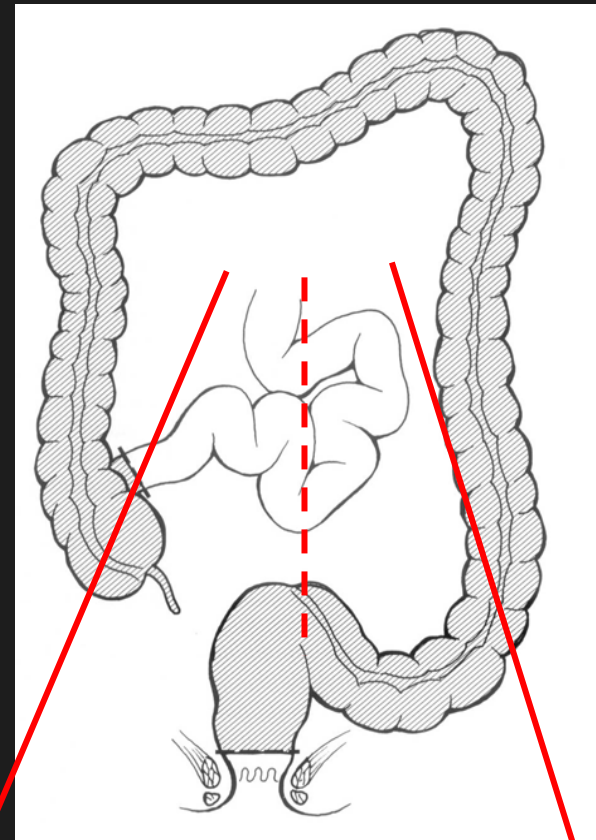
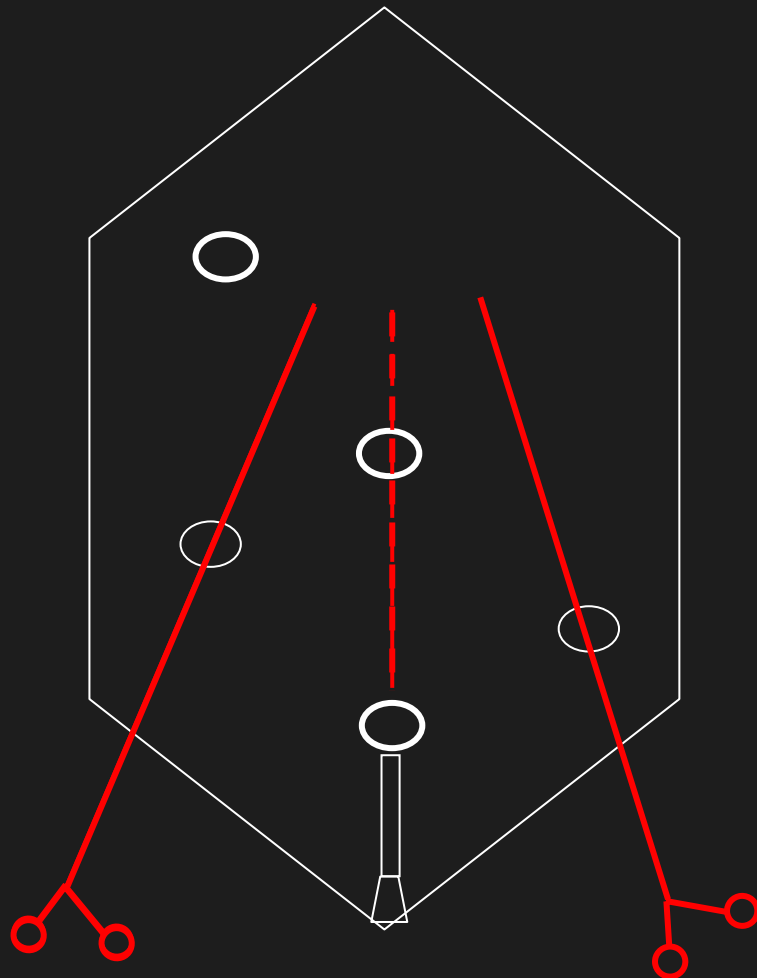
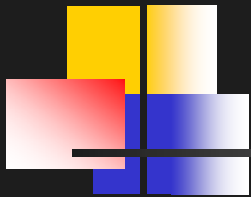
# Recto-sigmoid Mobilization



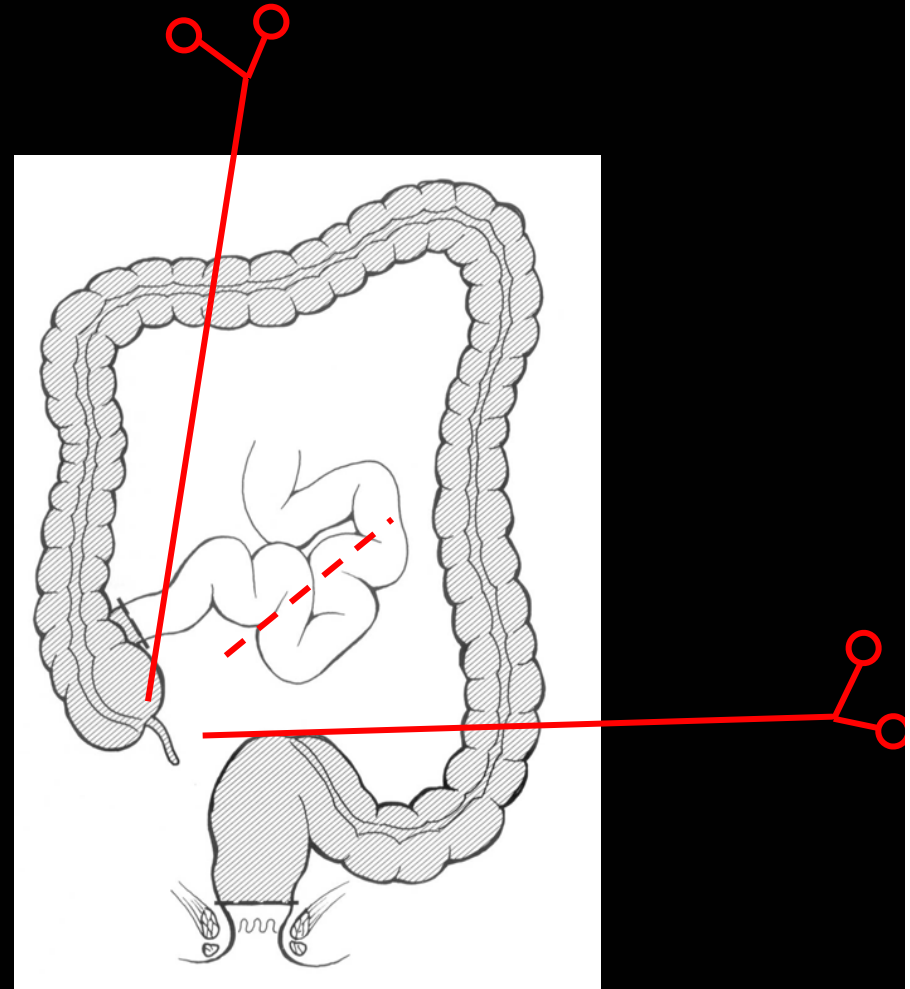
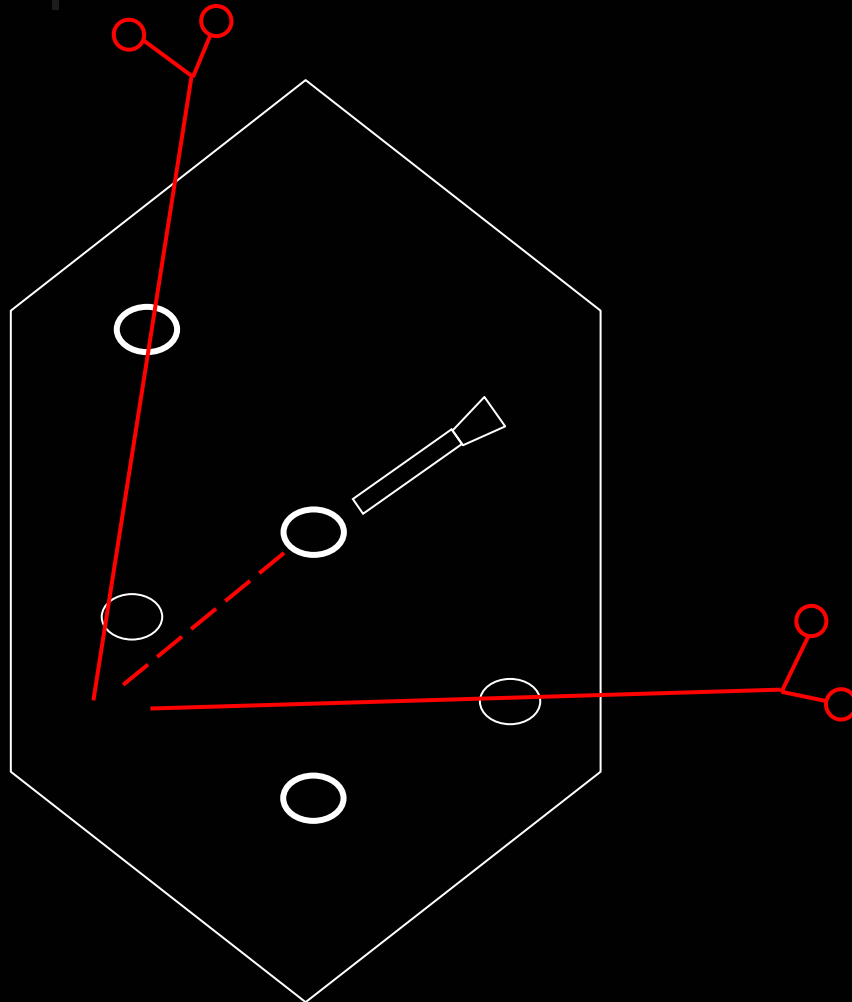
# Left colon Mobilization



# Transvers colon Mobilization



# Right colon Mobilization





The choice of operative approach

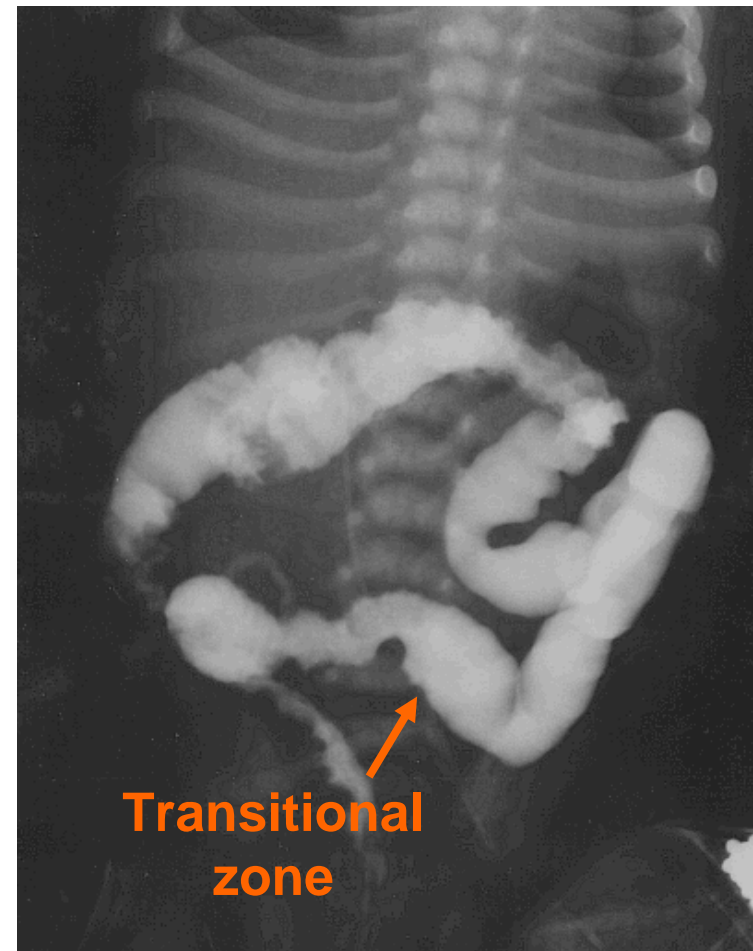
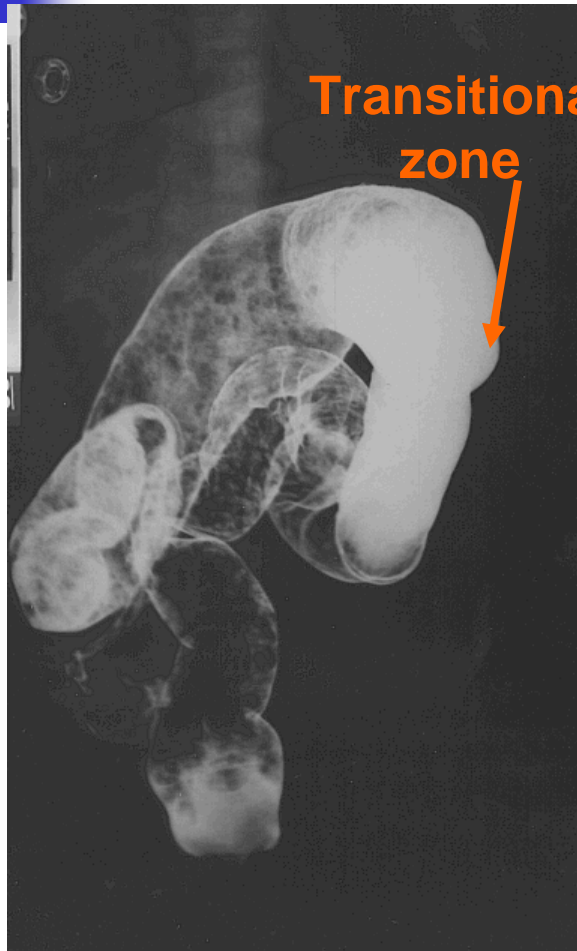
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is dictated

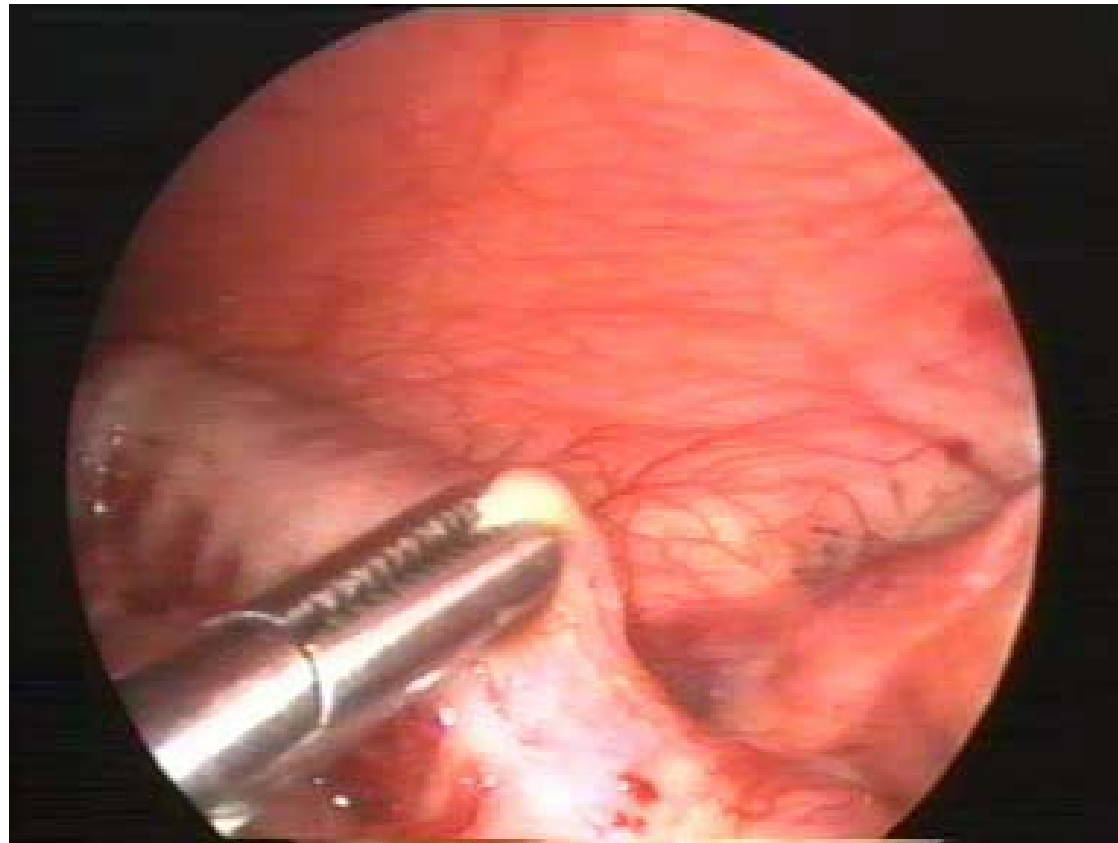
by identification of distal  
ganglionic intestinal segment.

# Hirschsprung's Disease

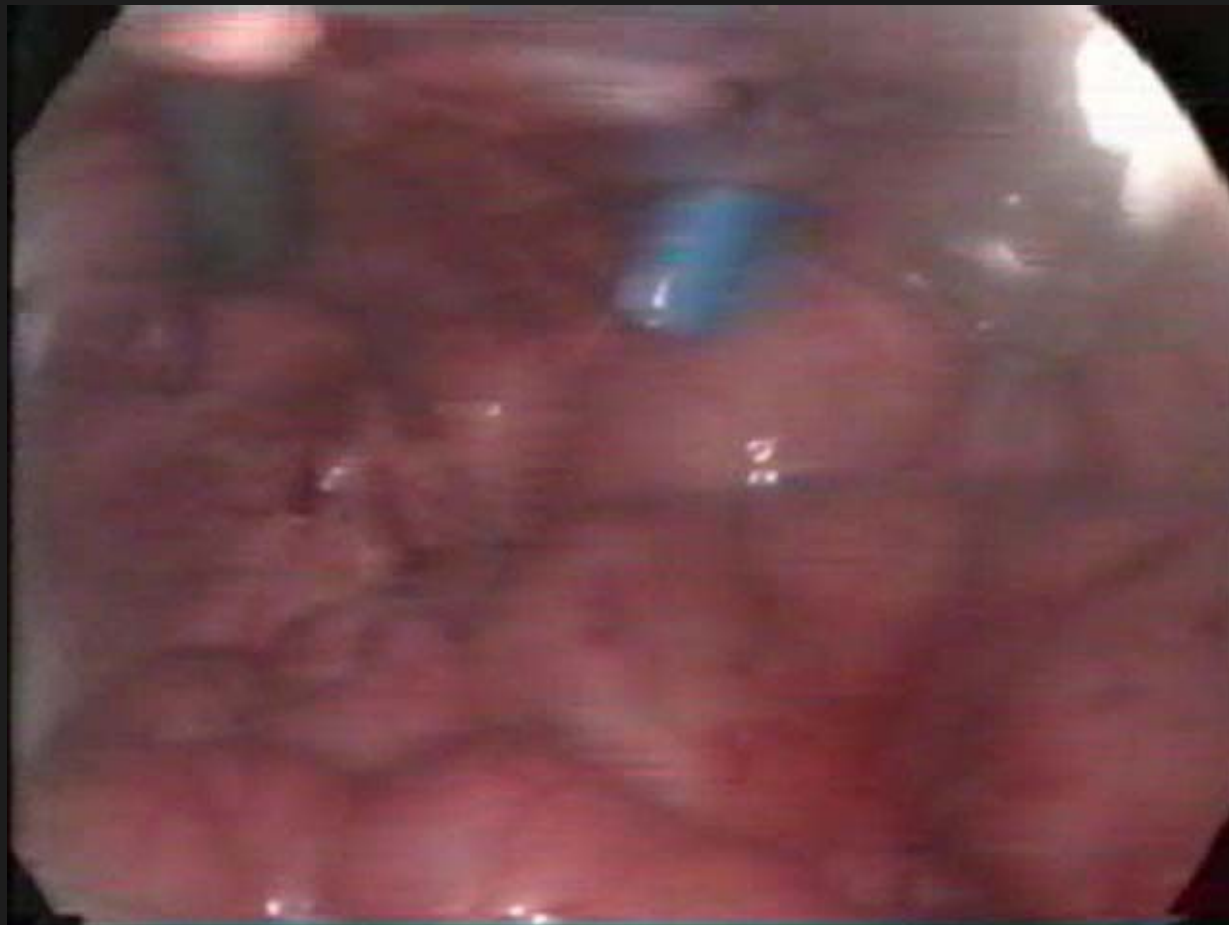
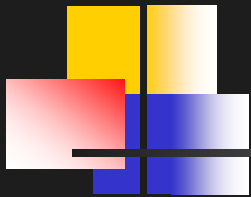
*contrast enema radiography*



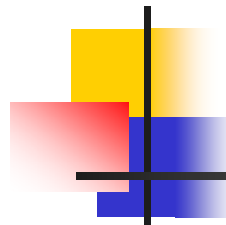
# Seromuscular Biopsy



# Total Colectomy



# Procedures for complications after transanal pull-through



| <u>Diagnosis</u>                       | <u>Procedure</u>                    |
|--|-------------------------------------|
| • peritonitis                          | laparotomy: washing and enterostomy |
| • pneumoperitoneum                     | laparoscopy: washing and suture     |
| • urethral injury                      | colostomy: urethral repair          |
| • anastomosis in aganglionic zone      | re-do                               |
| • anal stenosis and mild enterocolitis | Colostomy                           |



# Conclusion

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Laparoscopy has changed the management of rectosigmoid aganglionosis allowing the surgeon to use safely an established concept (pull-through) while eliminating a major source of morbidity (staged - correction and laparotomy).



# Critical factors

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- Experienced Surgeon and Pathologist.
- Long-term outcomes regarding continence and costipation are not established.

**But**

The preliminary data suggest similar results compared to the open techniques.

A landscape photograph featuring a bright sun in the upper center of the sky, creating a lens flare effect. A small airplane is visible in the distance, flying across the sky. The foreground consists of a dark, textured field. The text "Thank you for your attention." is overlaid in a yellow, cursive font, slanted upwards from left to right.

*Thank you for your attention.*