



The Hirschsprung Hunt

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Hirschsprung disease

First described by Harold Hirschsprung
(1886)

Congenital disorder (1 in 5000 newborns)

Pathophysiology

Cause - Lack of coordinated propulsive movement of the distal portion of the large bowel resulting from loss of intrinsic inhibitory innervation

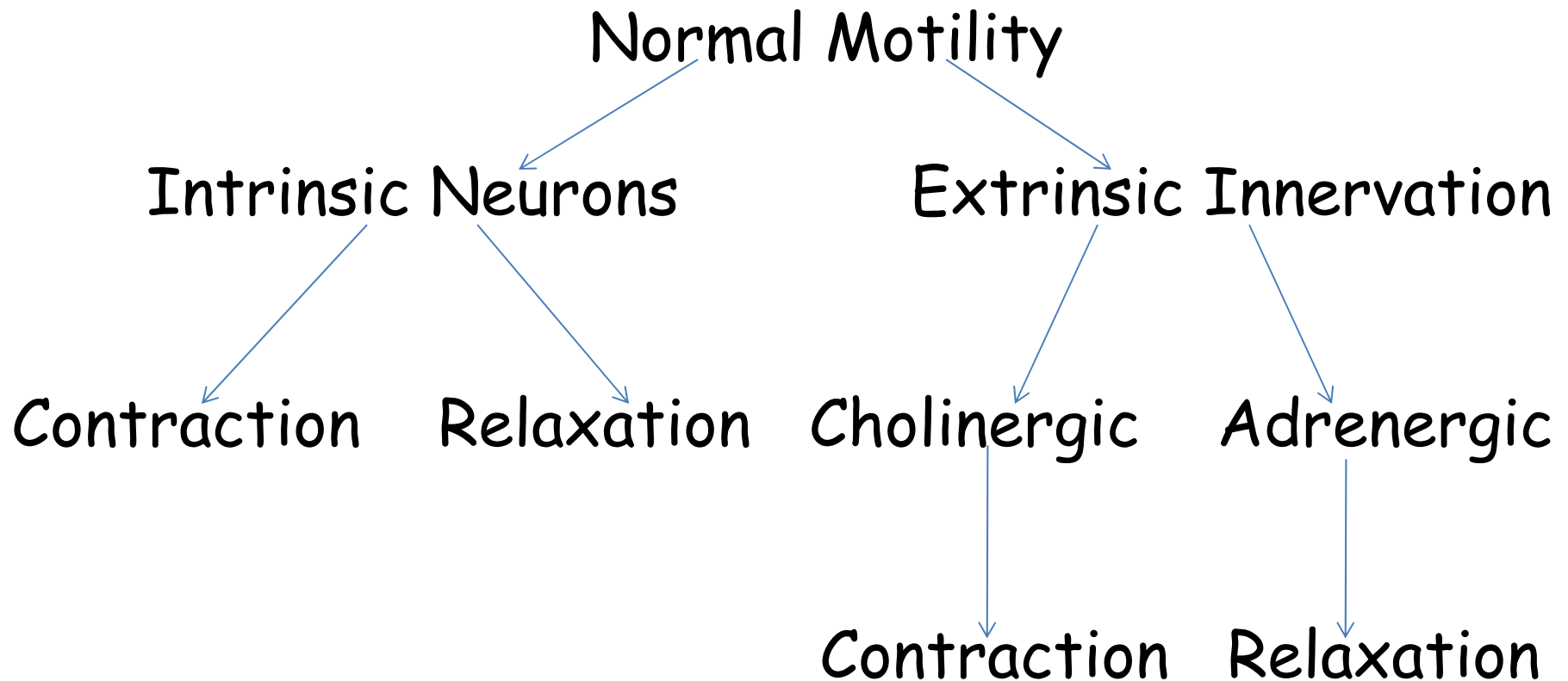
Absence of parasympathetic ganglion cells in the intramural and submucosal plexuses

Failure of migration from neural crest



Hirschsprung disease

Pathophysiology



Symptoms of HD

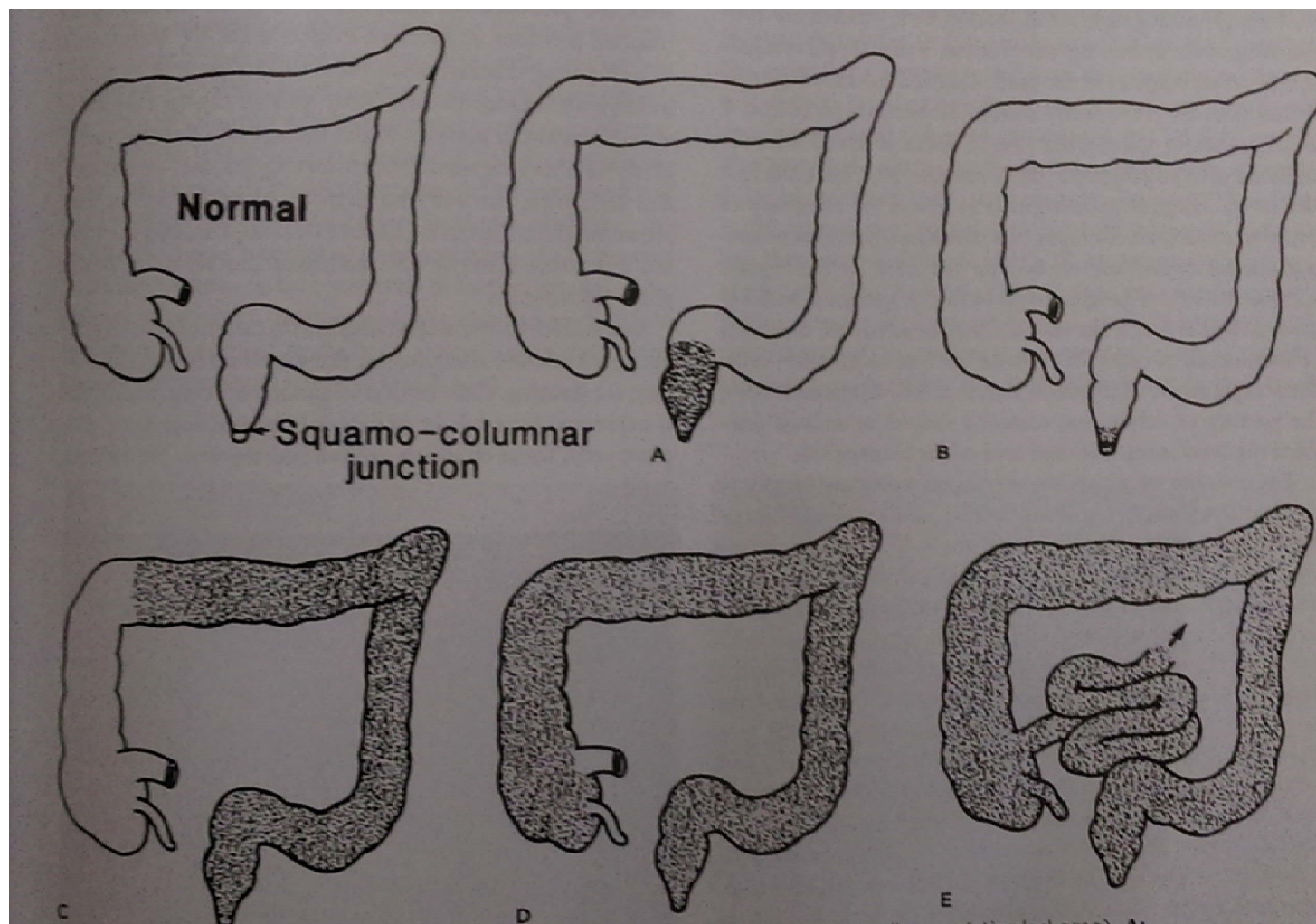
Nearly one-half children - non passage of meconium beyond 48 hrs

Delayed passage of first meconium beyond 36 hrs

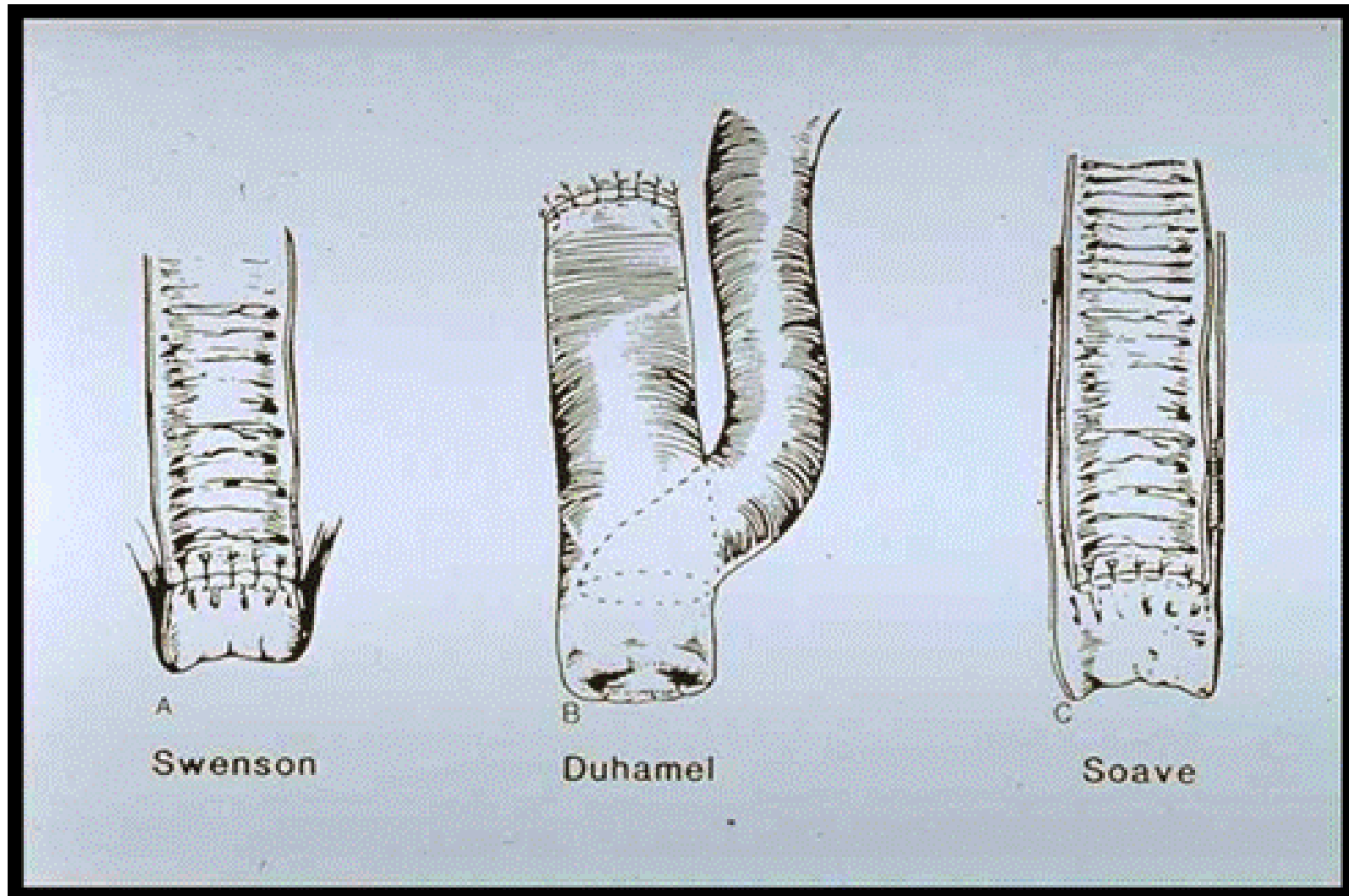
New born period - abdominal distension and repeated vomiting

Older children present with chronic constipation, distension , poor nutrition
May need enema therapy

Symptoms of enterocolitis - abdominal pain , vomiting, fever and foul-smelling or bloody diarrhea



Surgeries for HD



Role of frozen section in diagnosis of HD

- To decide level of colostomy
- To decide the level of pull through - doughnut assay
- For multiple intraoperative biopsies in one stage procedure

Implications of false positive or false negative

- Failure to recognize ganglion cells that are present - unnecessary extensive resection
- Misclassification of other cell types as ganglion cells - second surgery

Types of biopsies received

- Full thickness
- Seromuscular
- Suction/Mucosal/Submucosal

*Advantages of seromuscular biopsy over mucosal/
submucosal biopsy -*

Myenteric plexus is included

Ideal biopsy:

- Seromuscular biopsy
- Site - 2 cm above the dentate line
- Length - Min 5 mm
- Shrimp-like or a C-shaped profile (hemispherical)



Orientation:

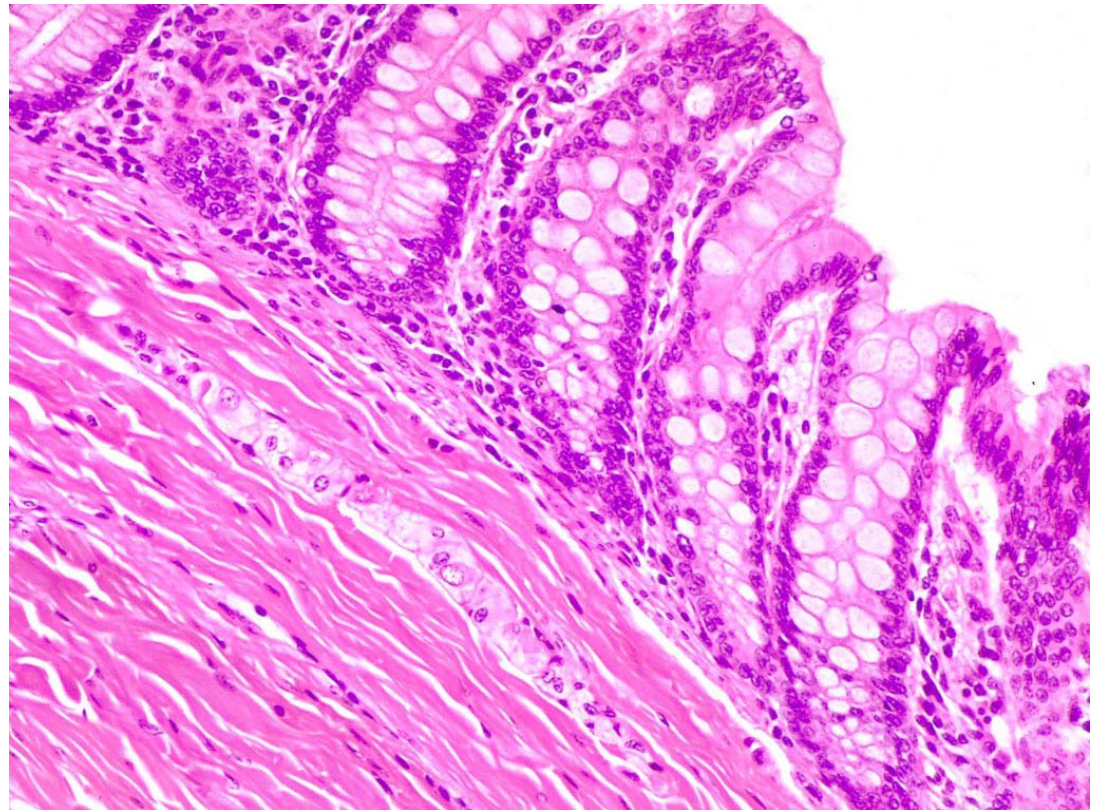
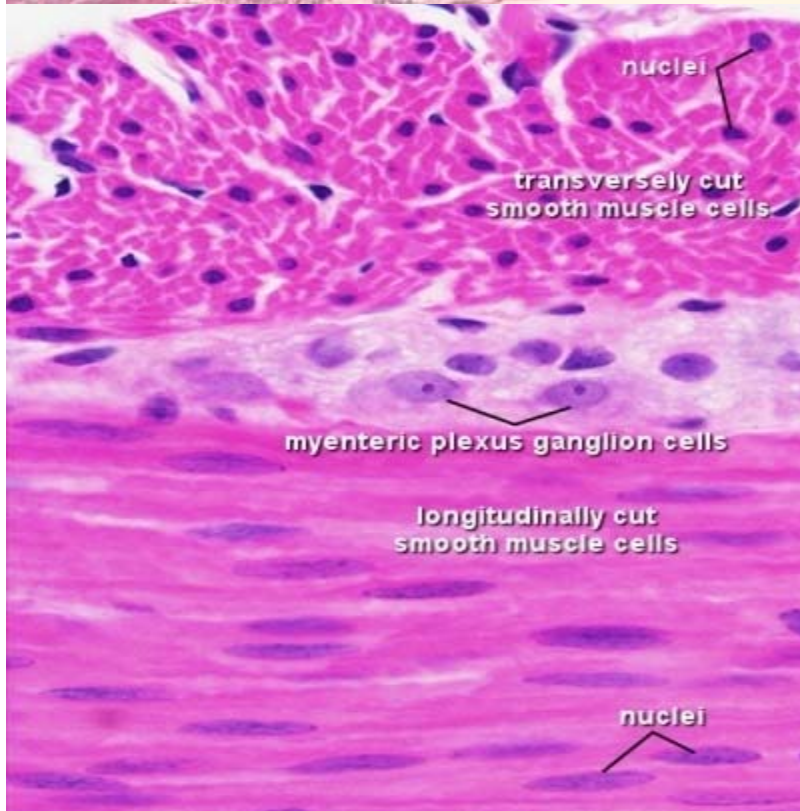
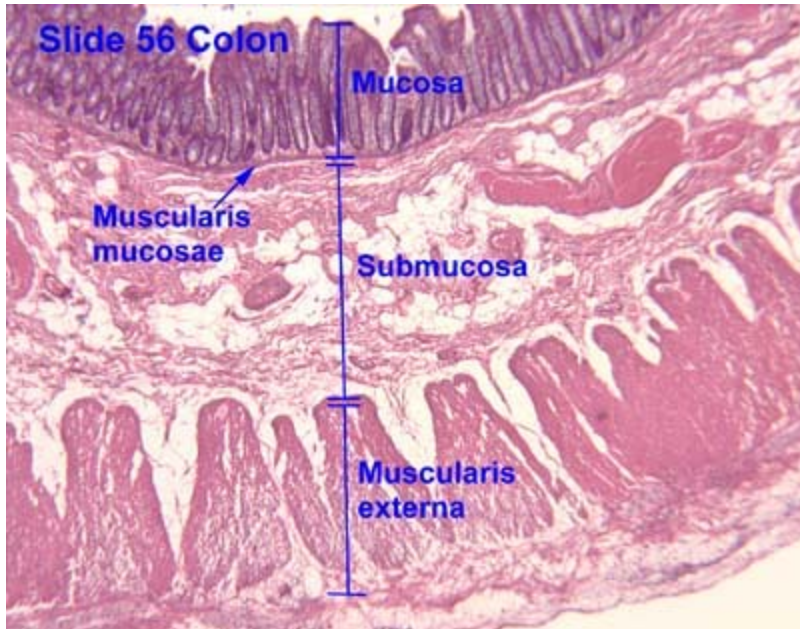
- Properly oriented
- Embedded so that full thickness and entire circumference of specimen is seen



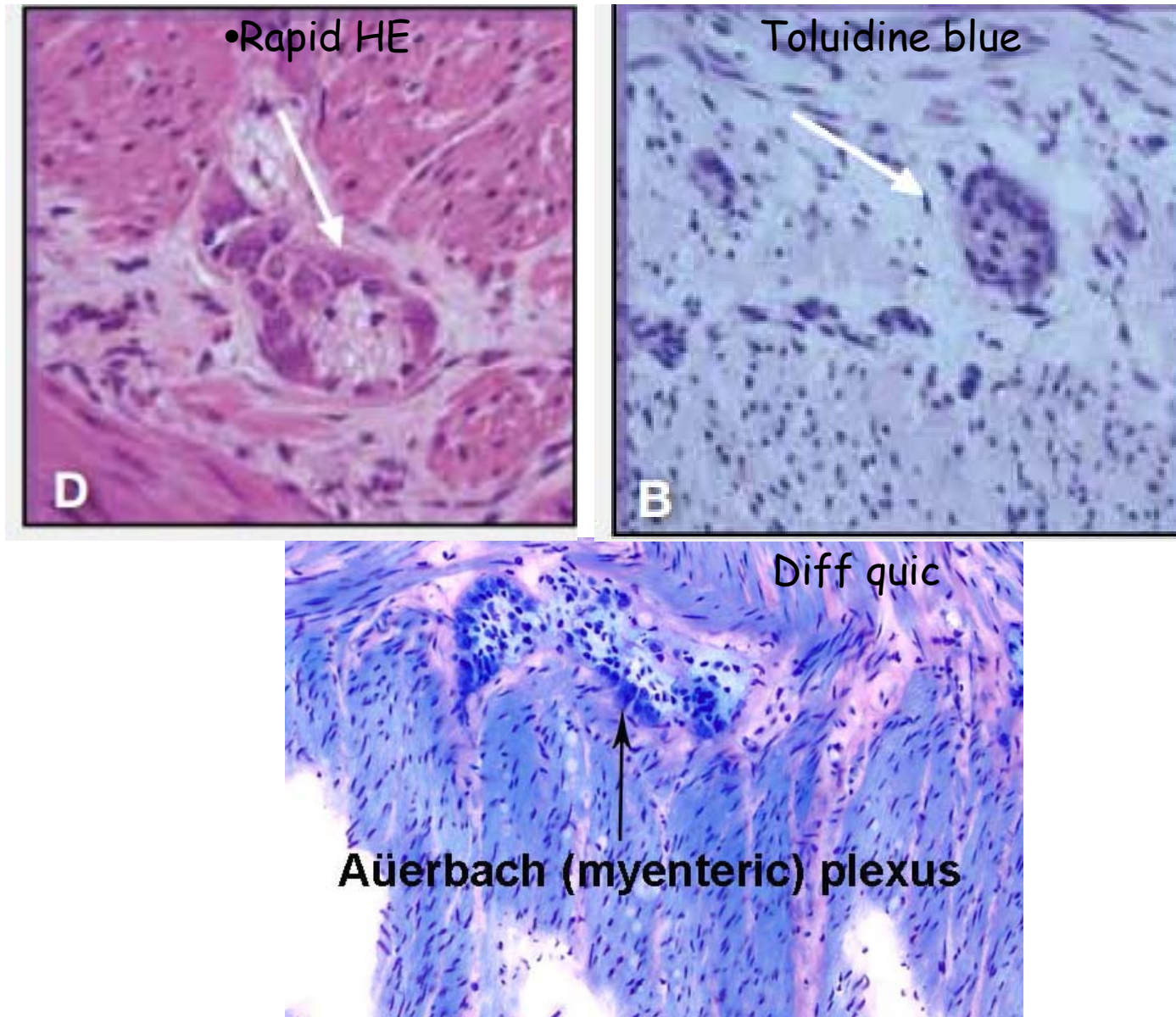
Diagnosis at Frozen section

- Presence of ganglion cells
- Presence of hypertrophied nerve bundles

Identification of ganglion cells



Stains used at Frozen section



Histologic – Technical bulletin for histotechnology, 2007

Methods to improvise the frozen sections

- Thicker sections
- Staining for a longer duration
- Taking additional 2-3 cuts

Problems at Frozen Section

- Newborns - immature ganglion cells
- *Mimics of ganglion cells* -
 - Plump endothelial cells lining the blood vessels
 - Heavy inflammatory infiltrate - large plasma cells, activated lymphocytes and activated endothelial cells
 - Adjacent smooth muscle cells, Schwann cells
- "Leading edge" can have normal innervation and normal number of ganglion cells in a portion of the wall

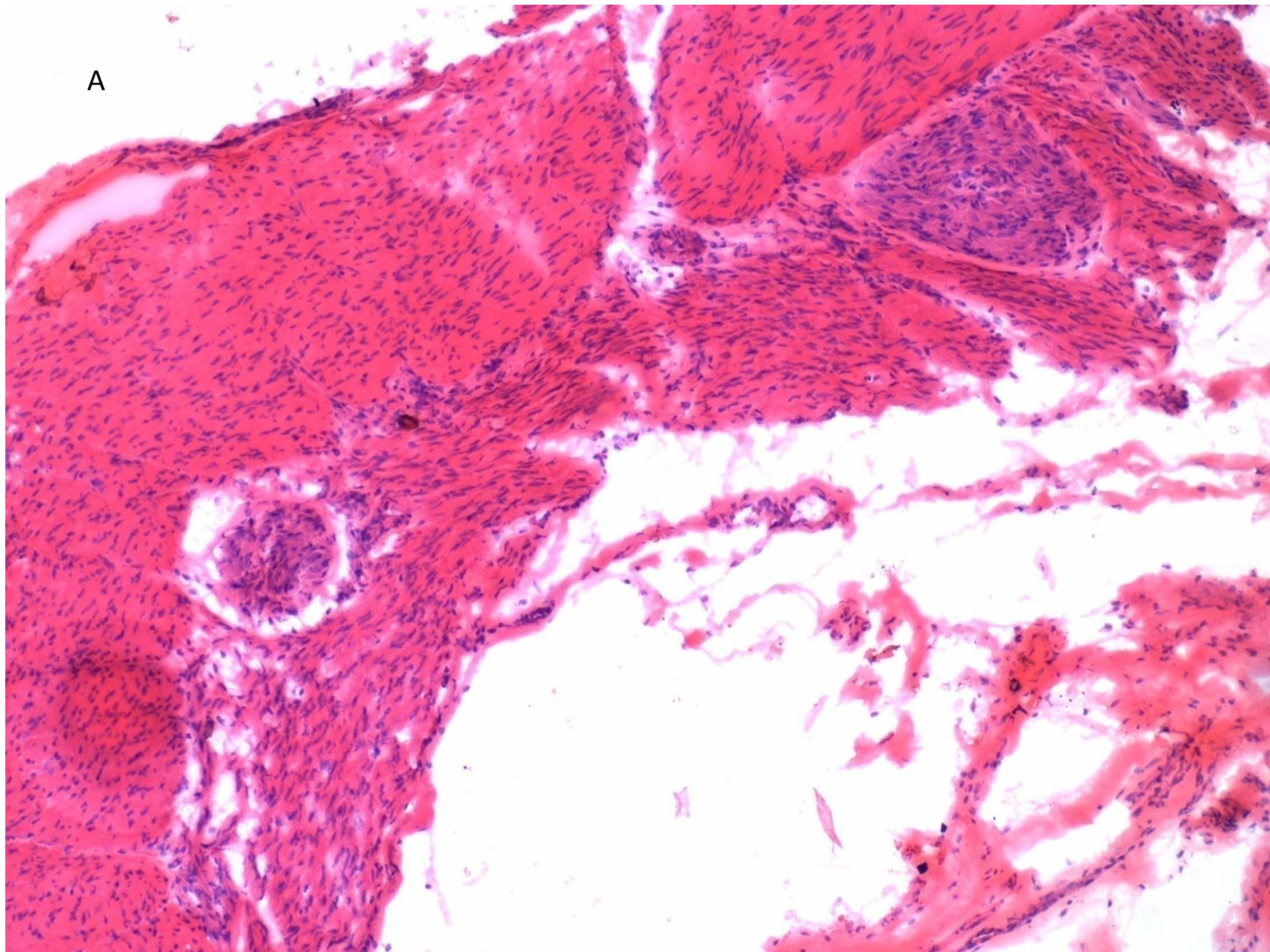
Point to remember

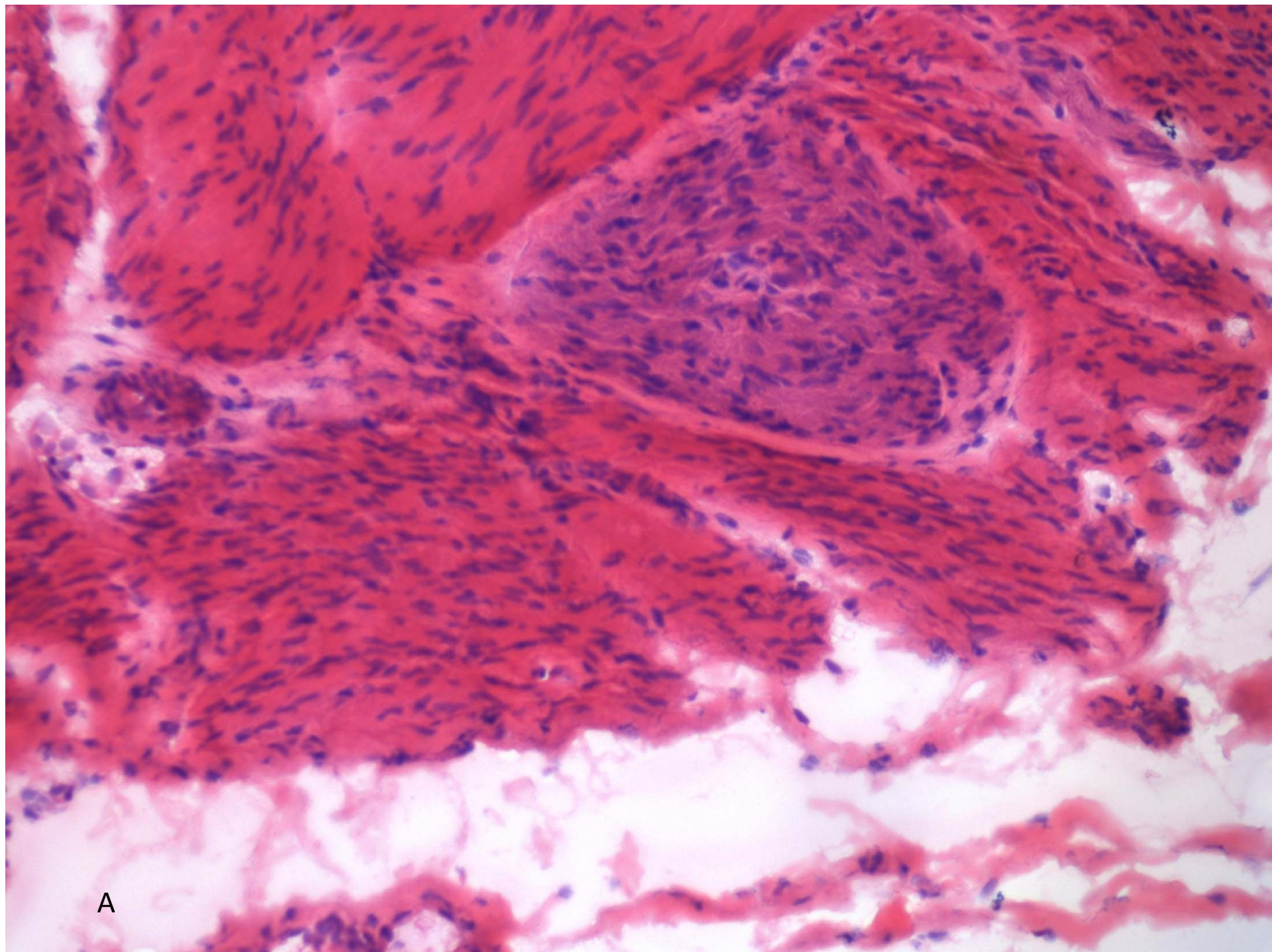
- Isolated ganglion cells are insufficient
- Look for the neural units
- Hypoganglionosis and hypertrophied nerves immediately proximal to the aganglionic segment. This should be interpreted as transition zone. This is not a good level for anastomosis
- Ensure that segment of bowel used for anastomosis has normal ganglion cells and normal-caliber nerves

Case 1

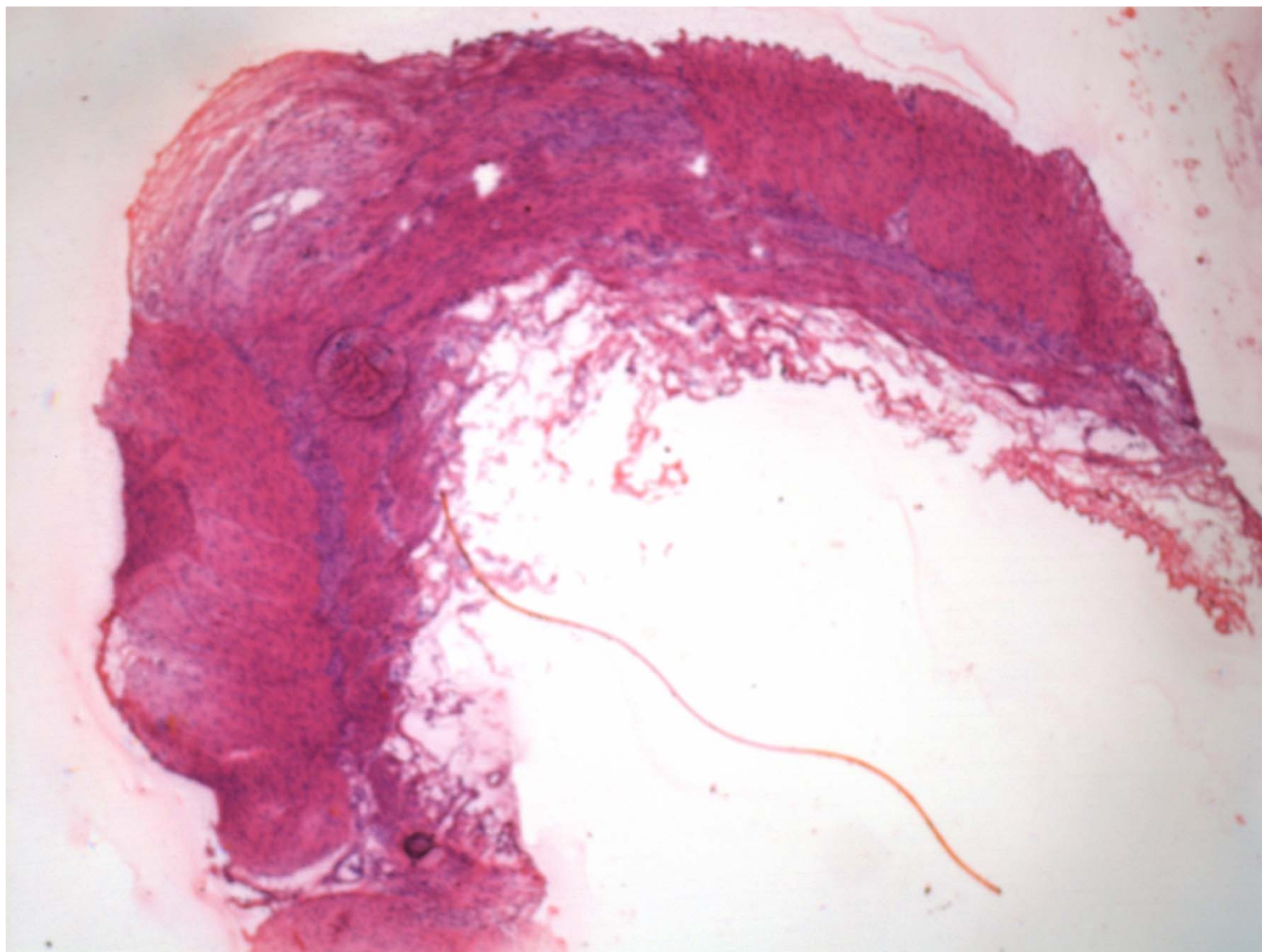
- An eleven months, Male
- Diagnosed on FTRB
- Transverse colostomy performed 4 months ago

A

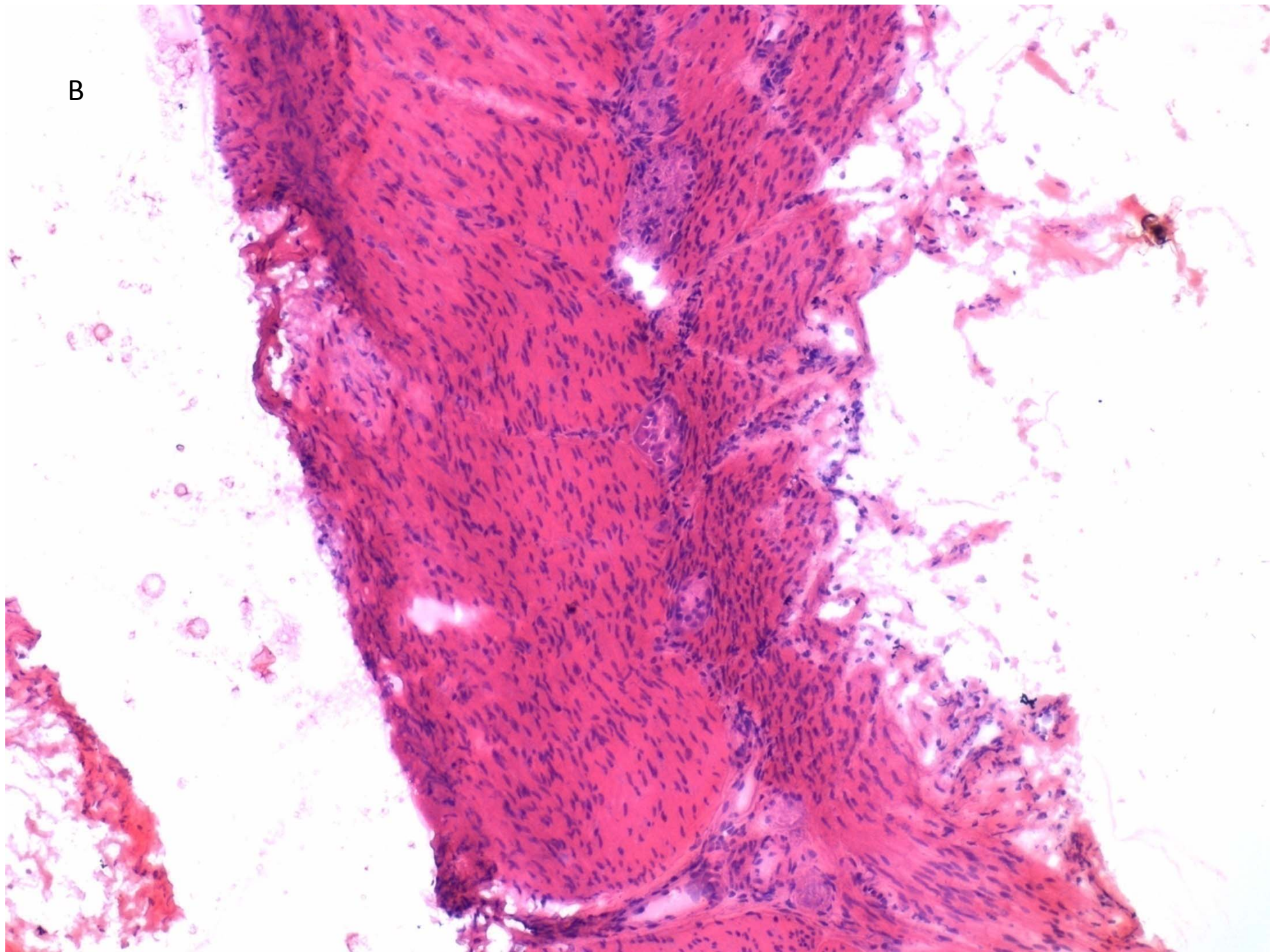


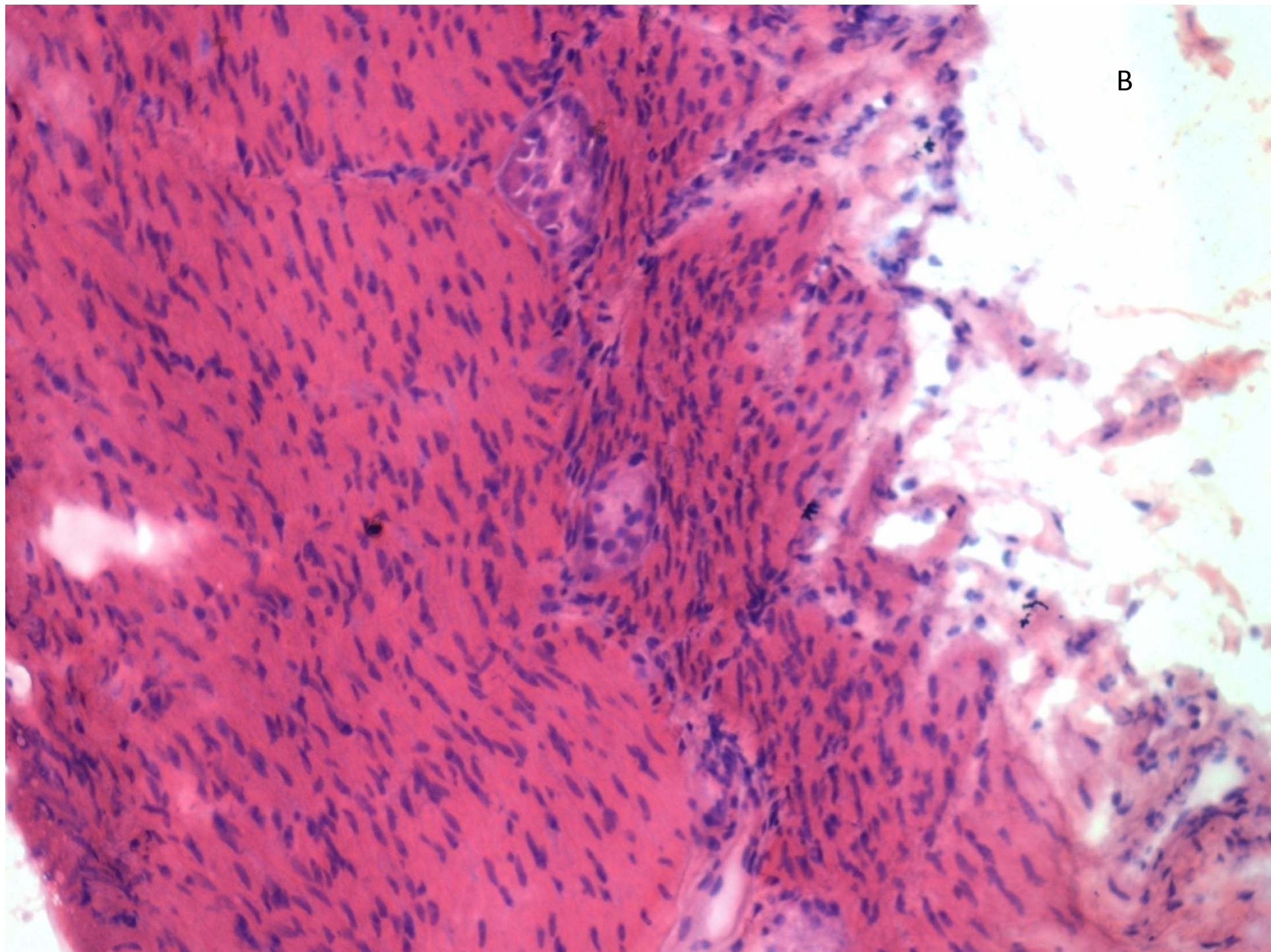


B

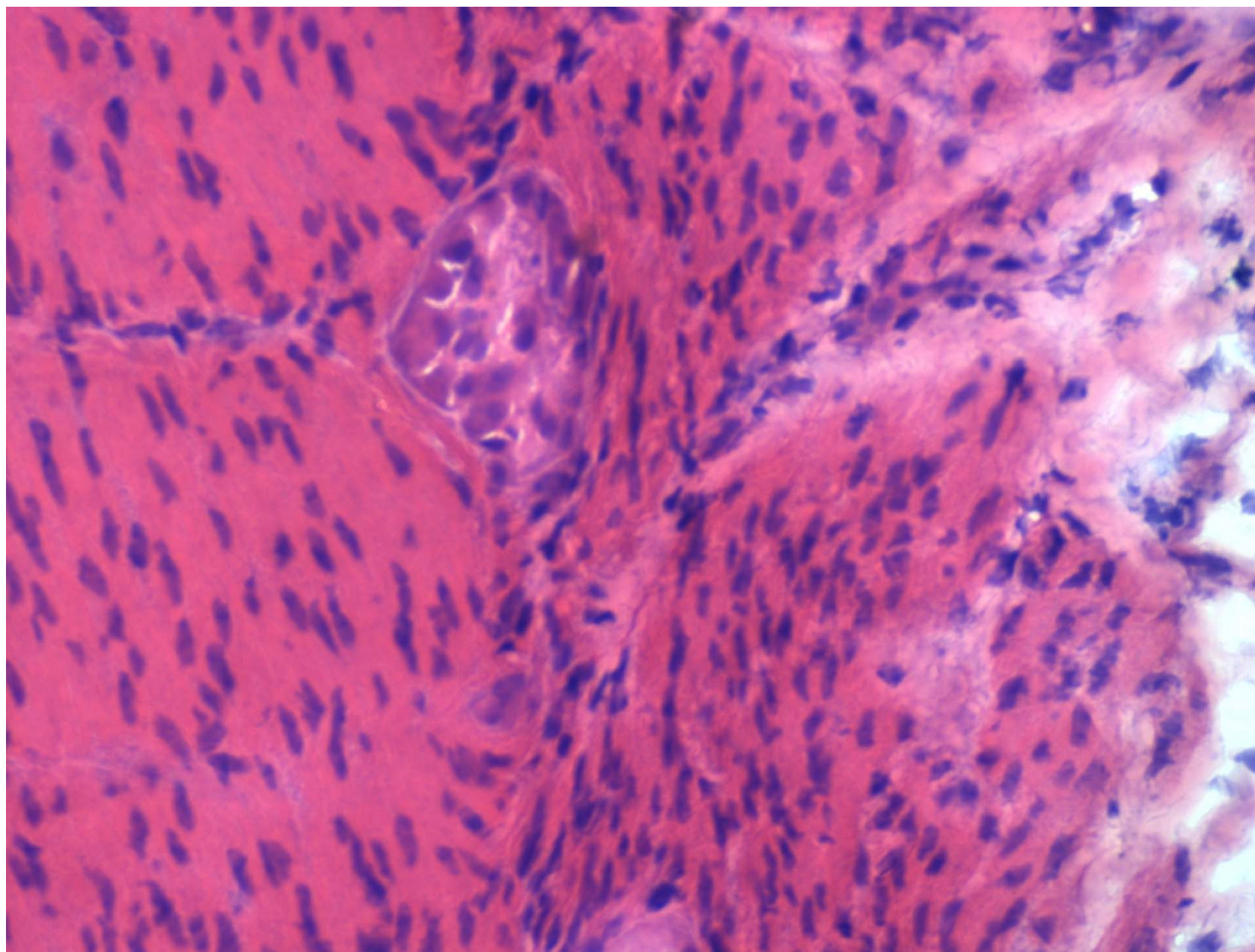


B



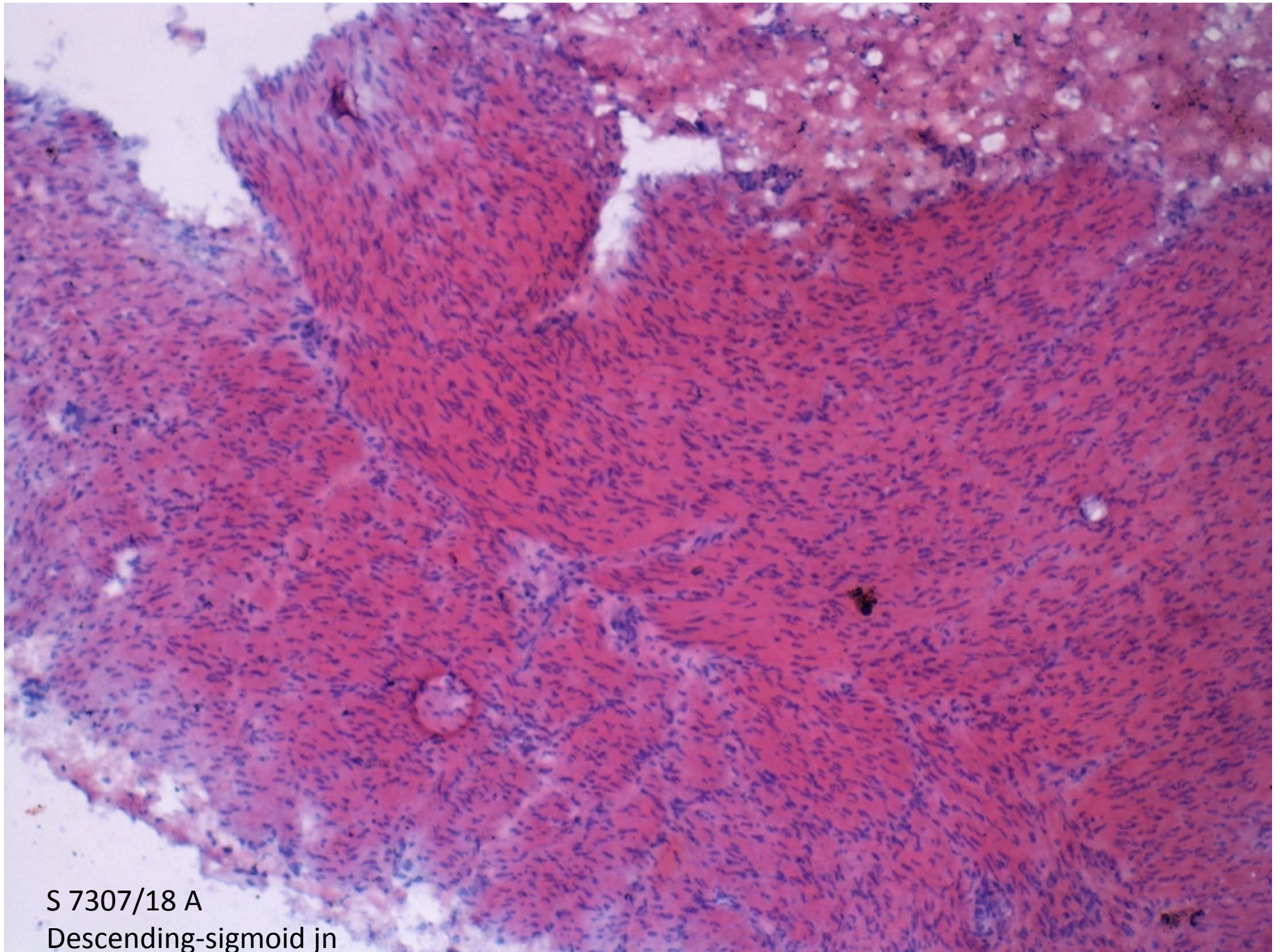


B



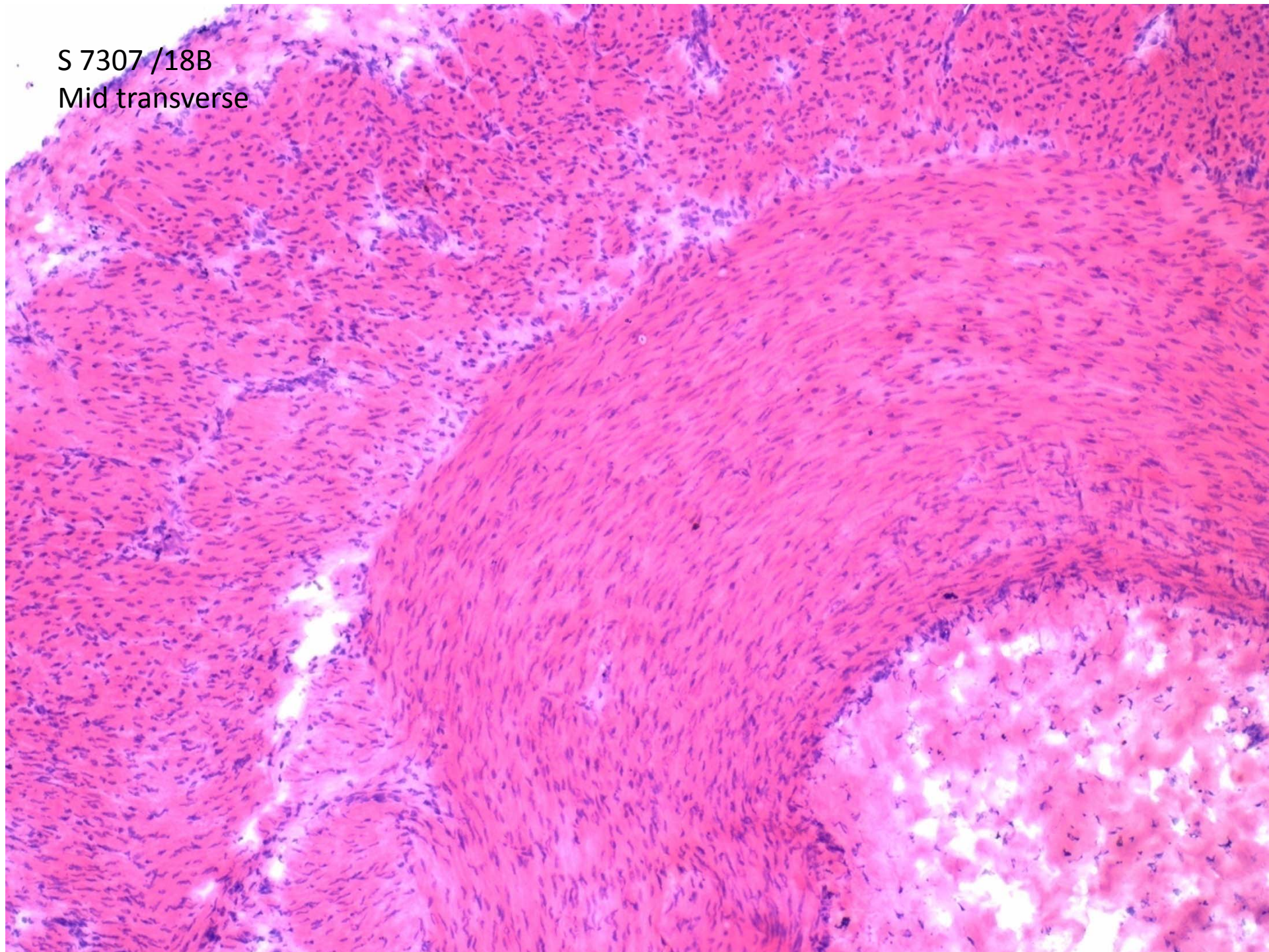
Case 2

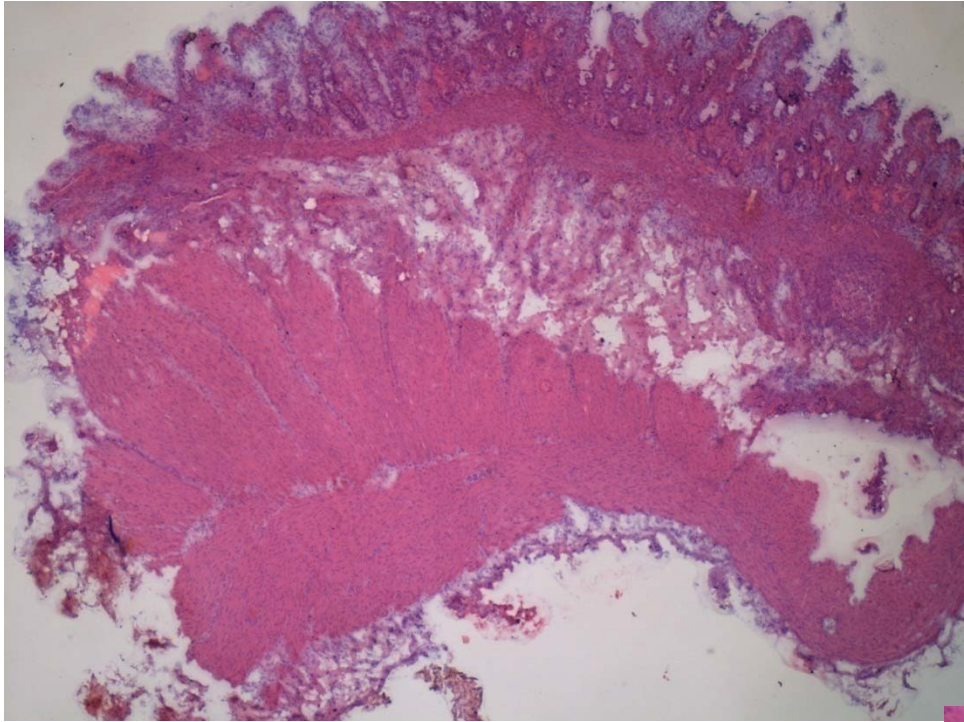
- A two years, male
- Diagnosed case of Hirschsprung dis in another hospital
- Ileostomy performed



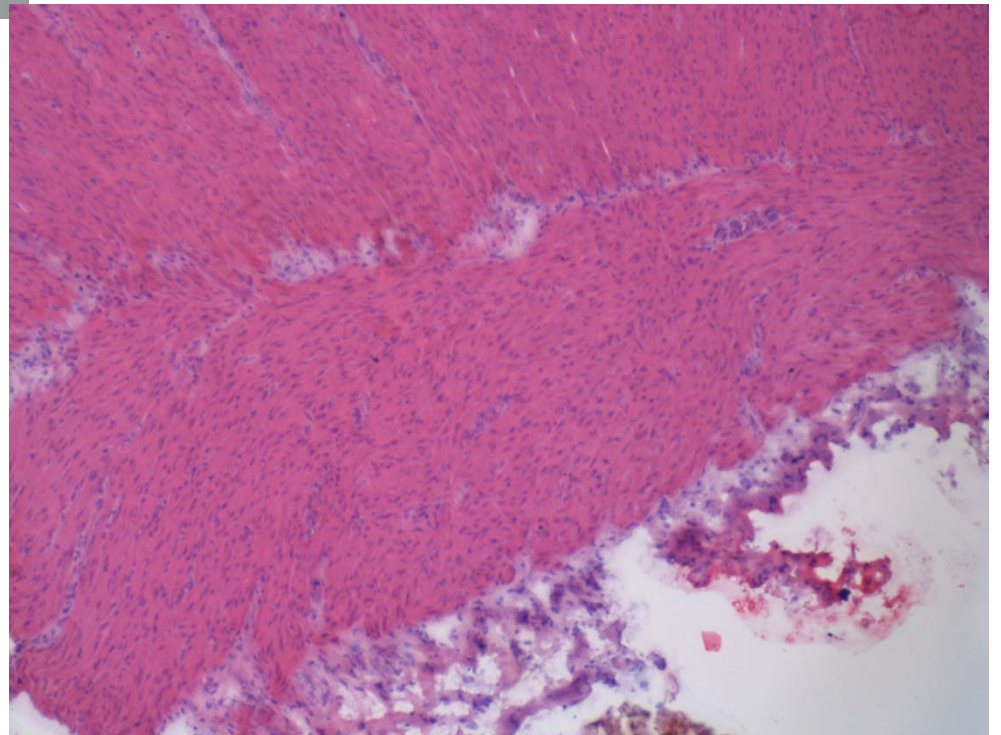
S 7307/18 A
Descending-sigmoid jn

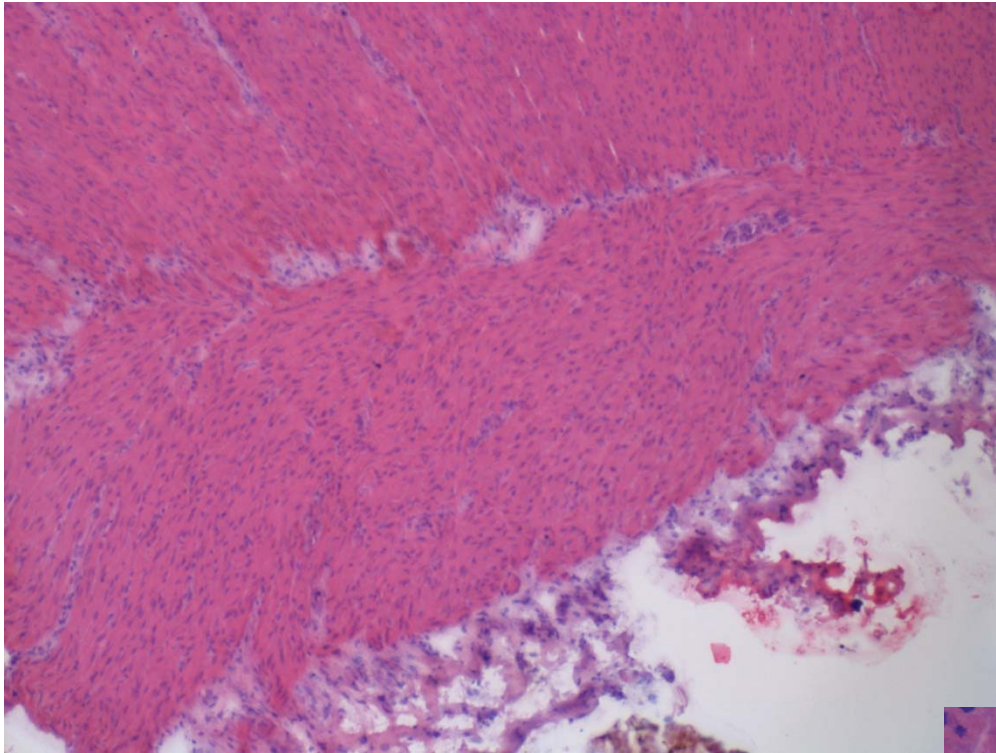
S 7307 /18B
Mid transverse



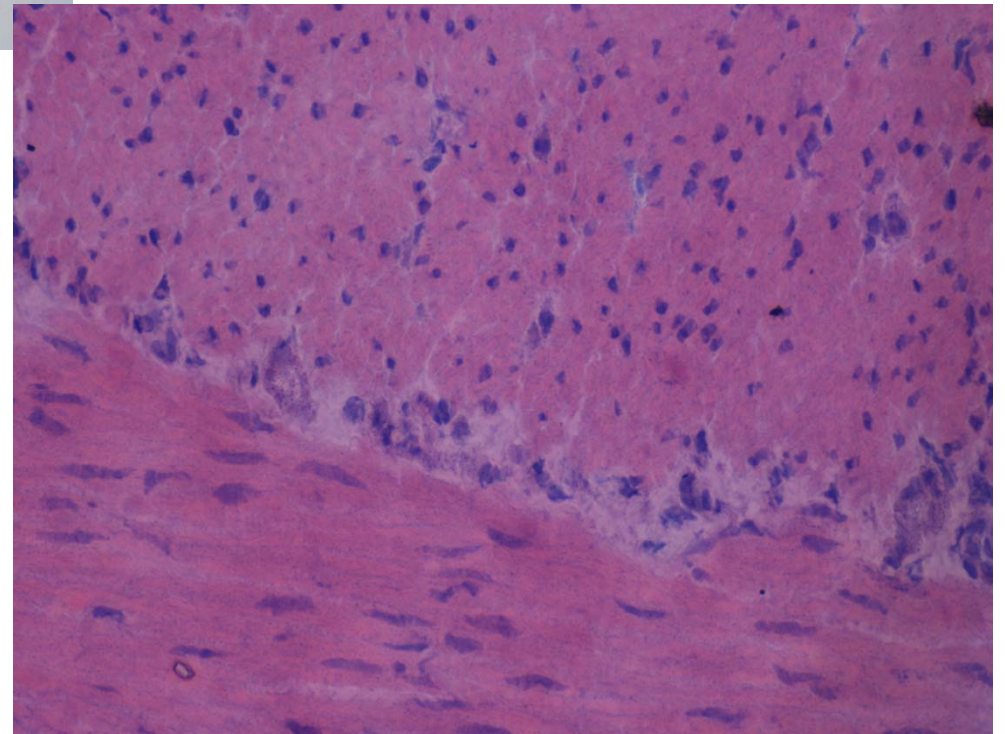


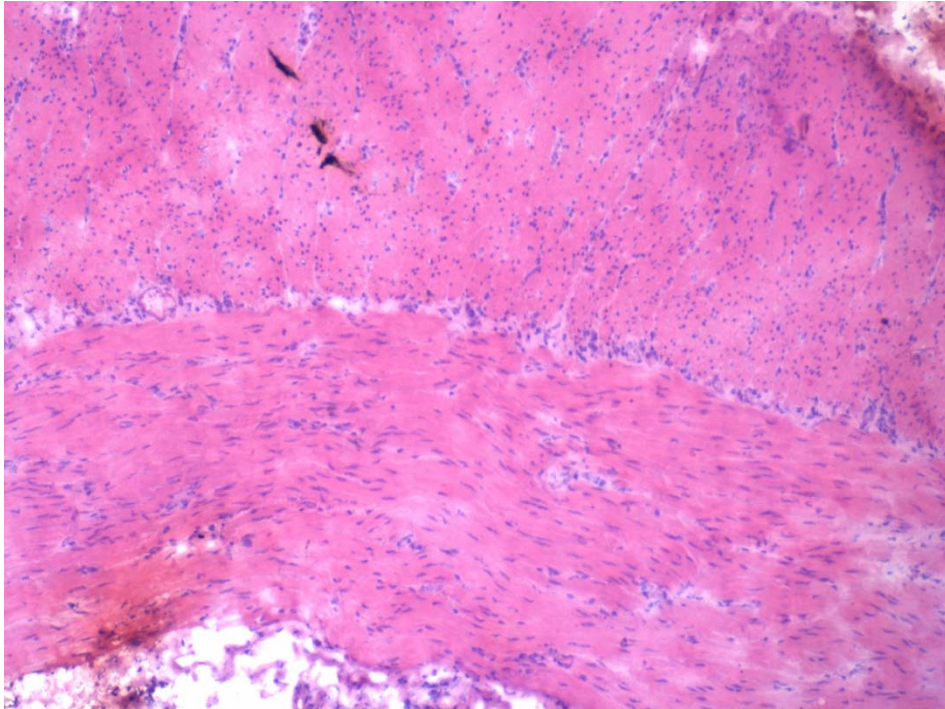
S 7307/18 C
Cecum-ascending jn



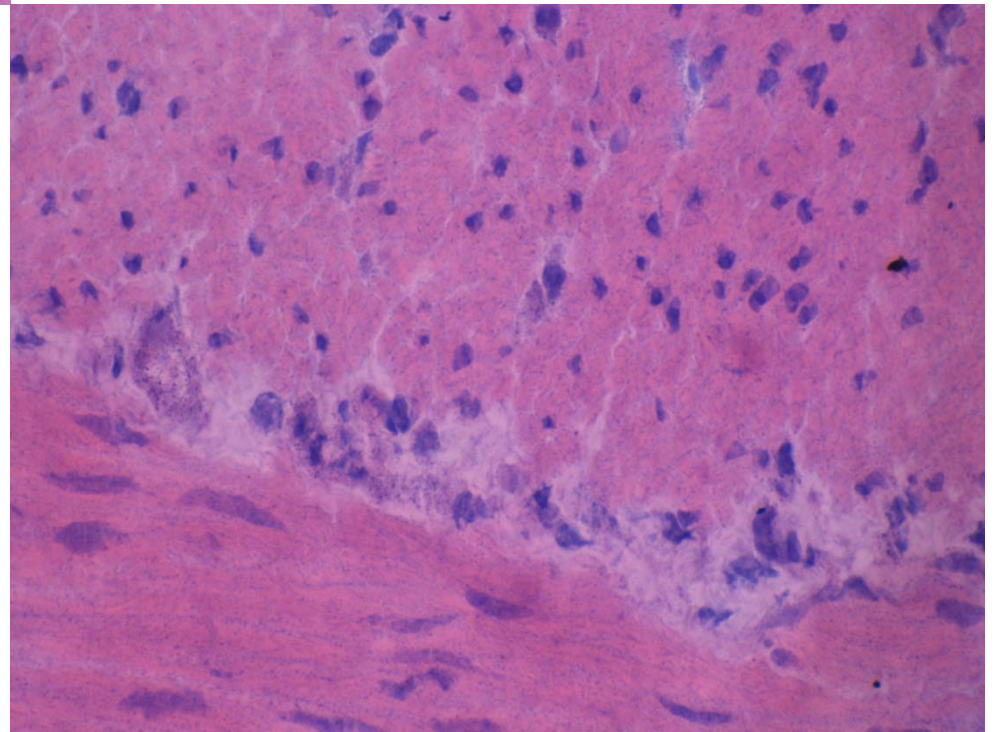


S 7307/18 D
10 cm prox to ileostomy

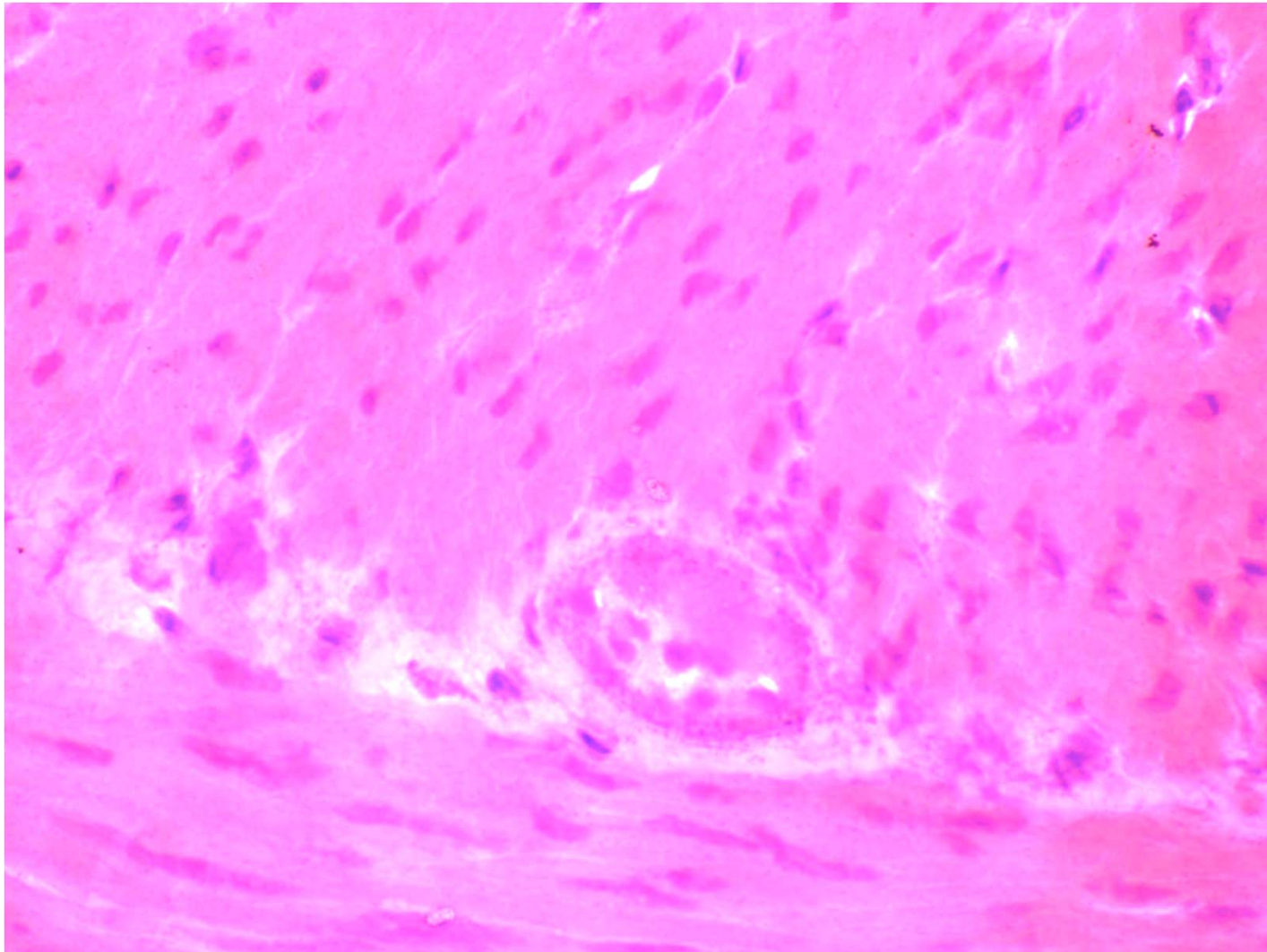


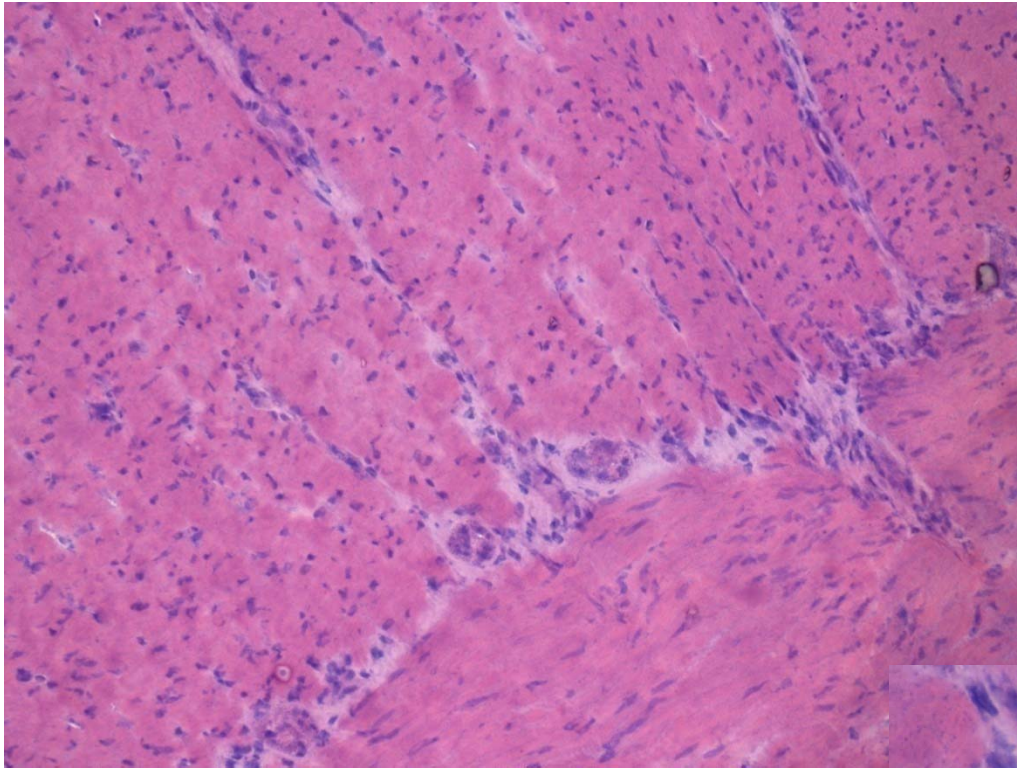


S 7307/18 E
More proximal to ileostomy

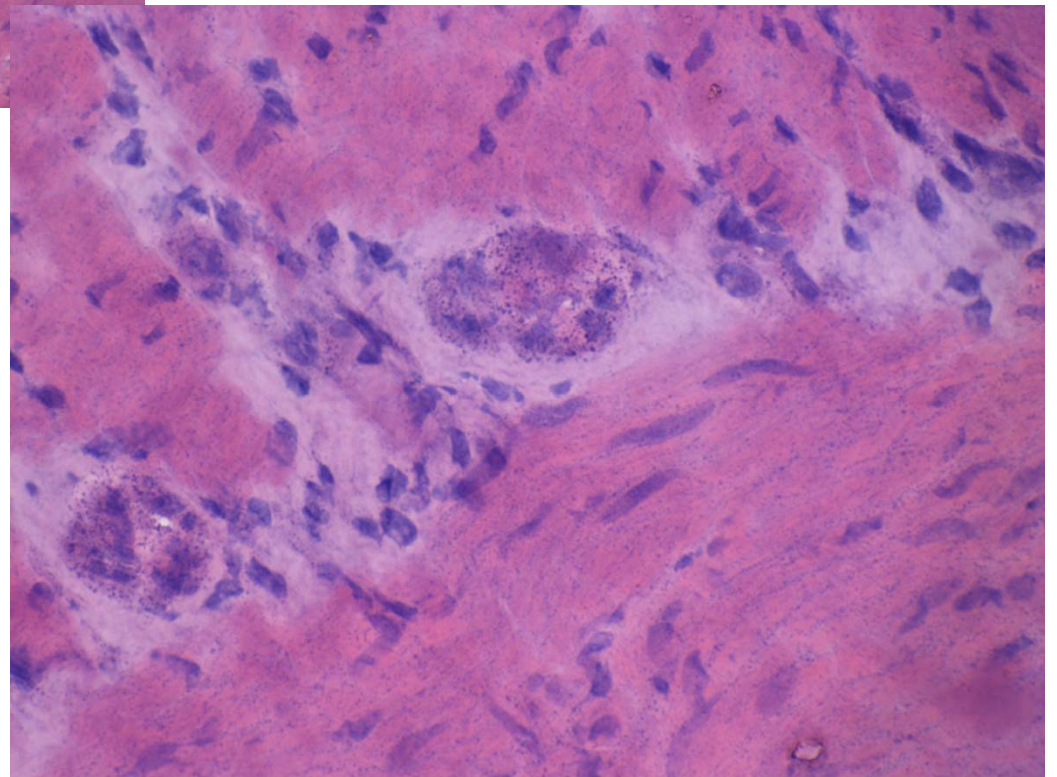


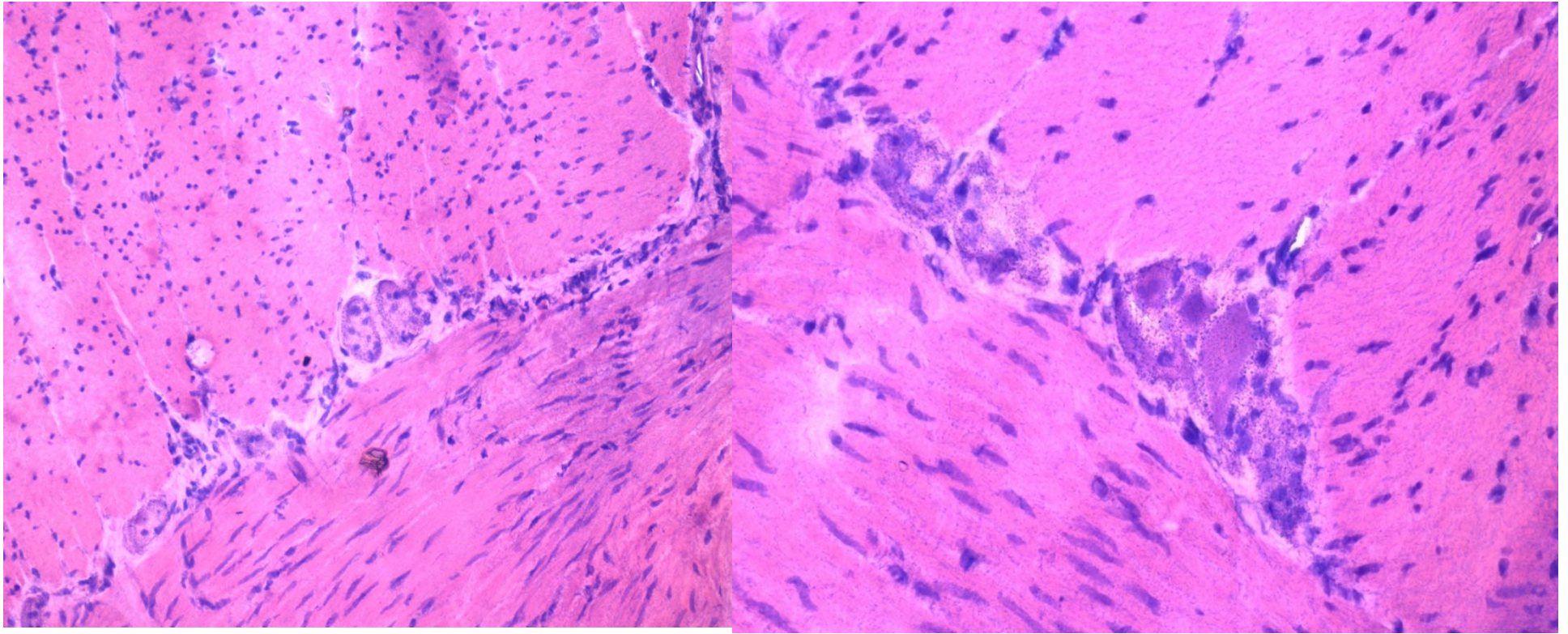
DEEP CUT S 7307/18 E



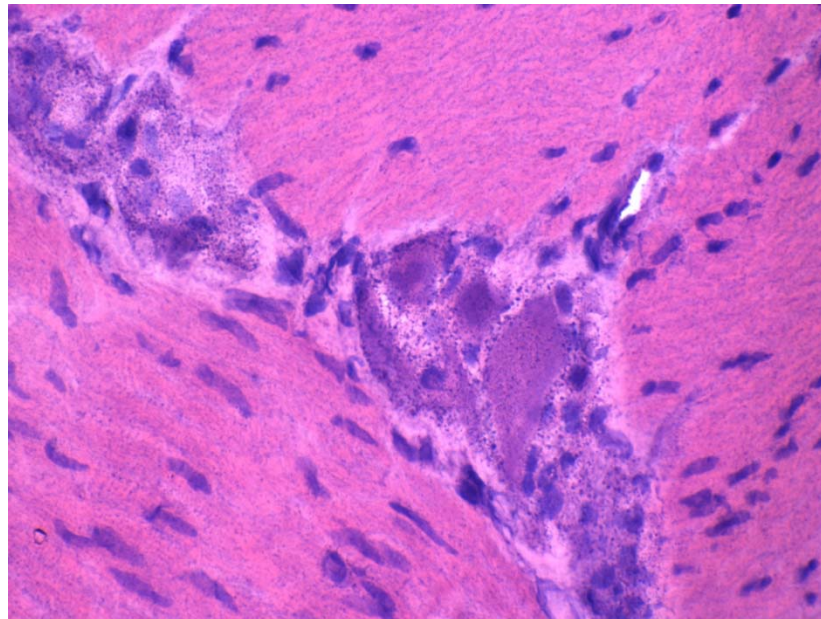


S 7307/18 F
More proximal to ileostomy





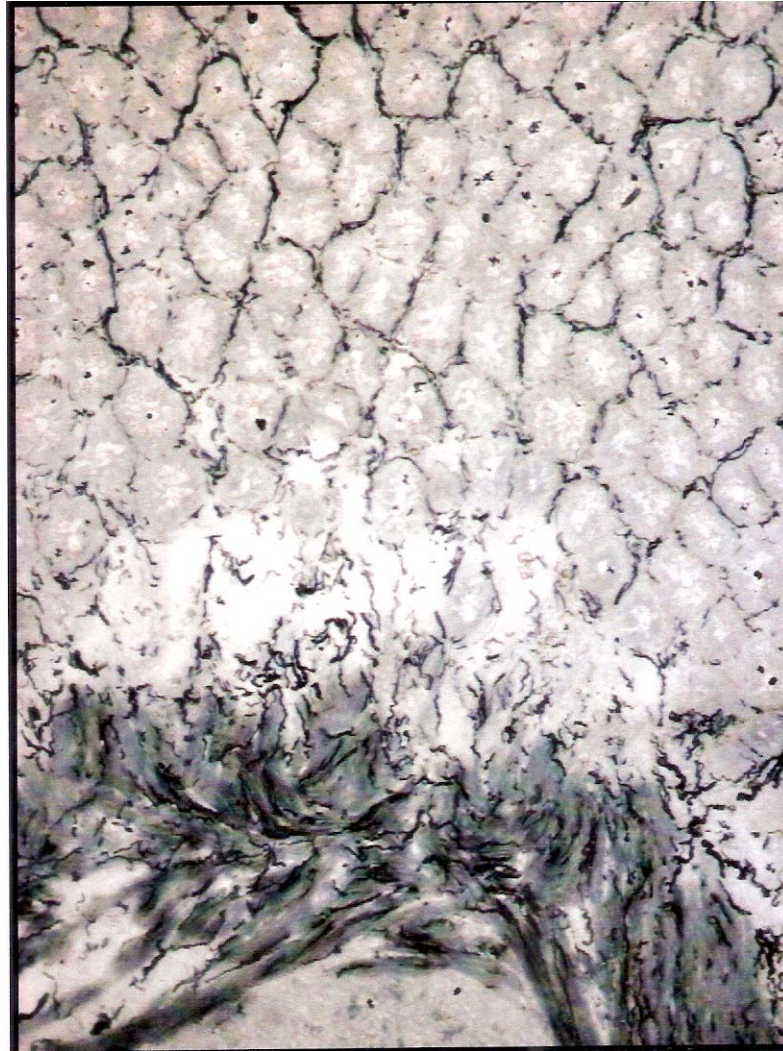
S 7307/18 G
5 cm proximal to previous
ileal biopsy



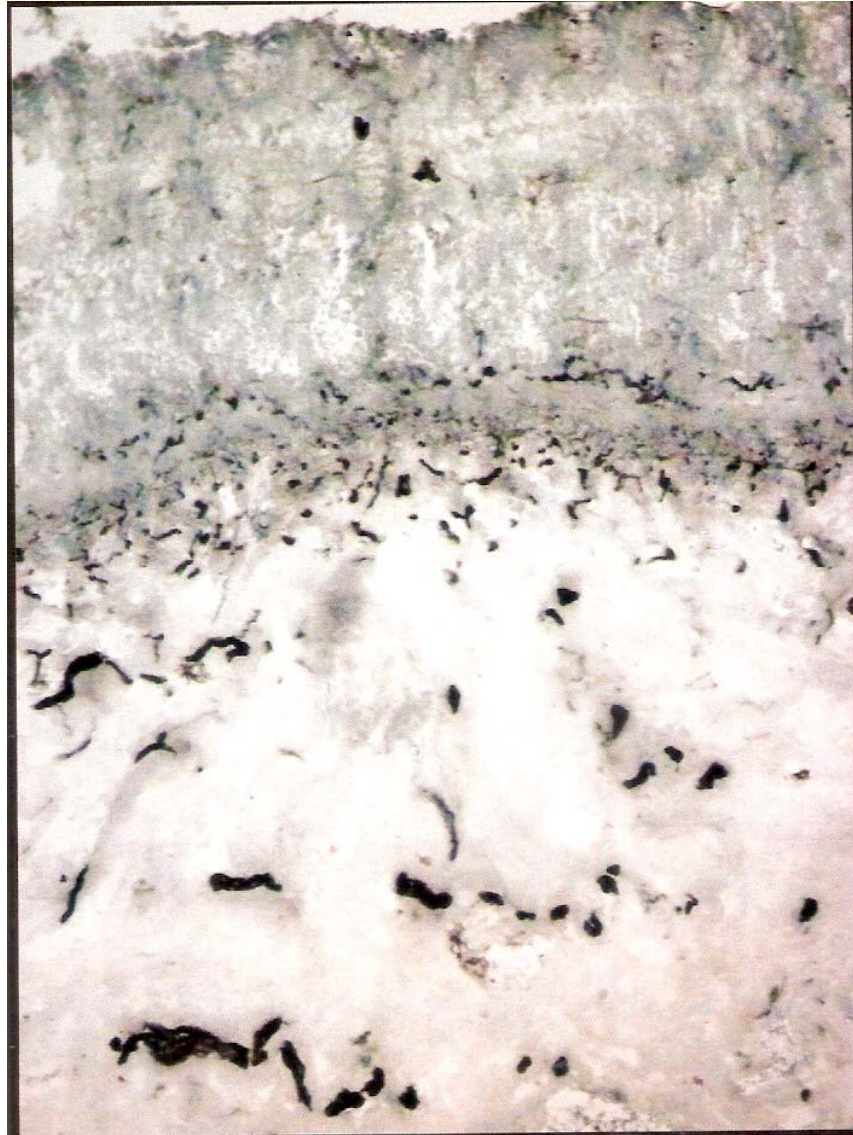
Role of acetyl choline esterase

- Useful as the diagnosis of HD is based on a negative finding
- As an adjunct to H&E
- The increased density and coarseness of cholinergic fibres - demonstrated by Ach E
(Normal nerves do not stain for AChE, but increased AChE expression is associated with the hypertrophied extrinsic nerve fibres of the aganglionic segment)

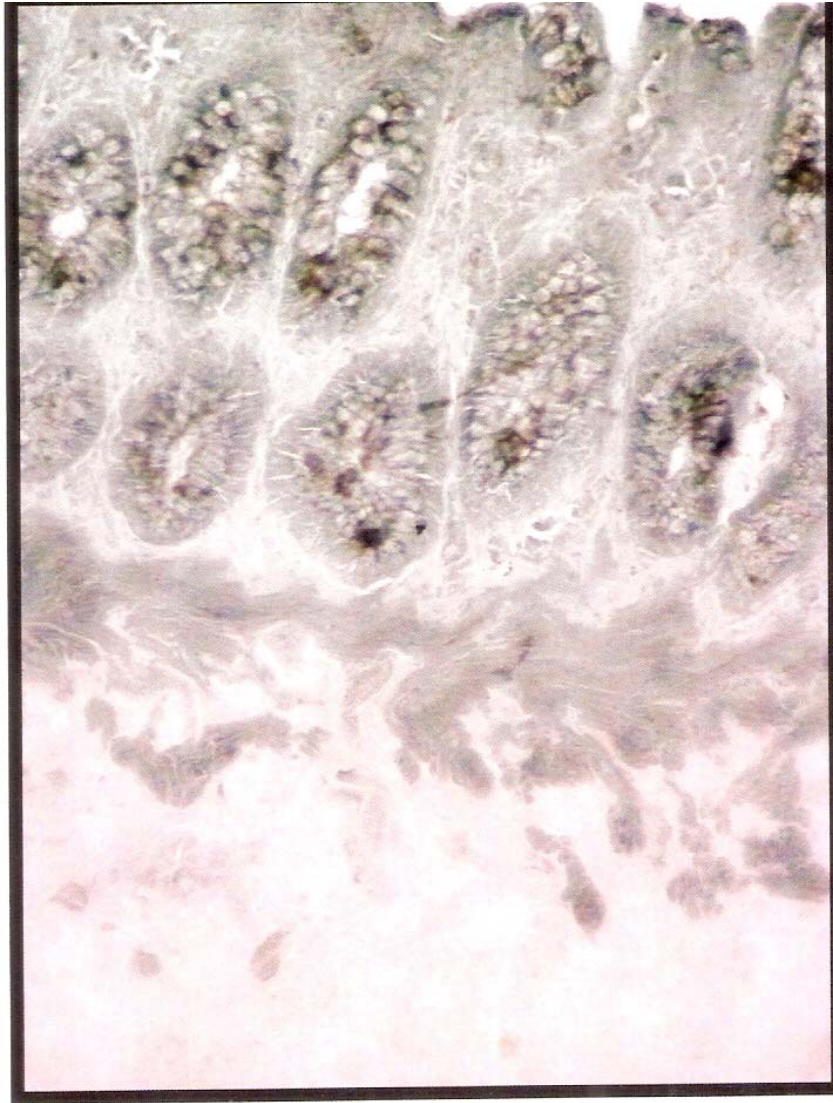
Pattern A



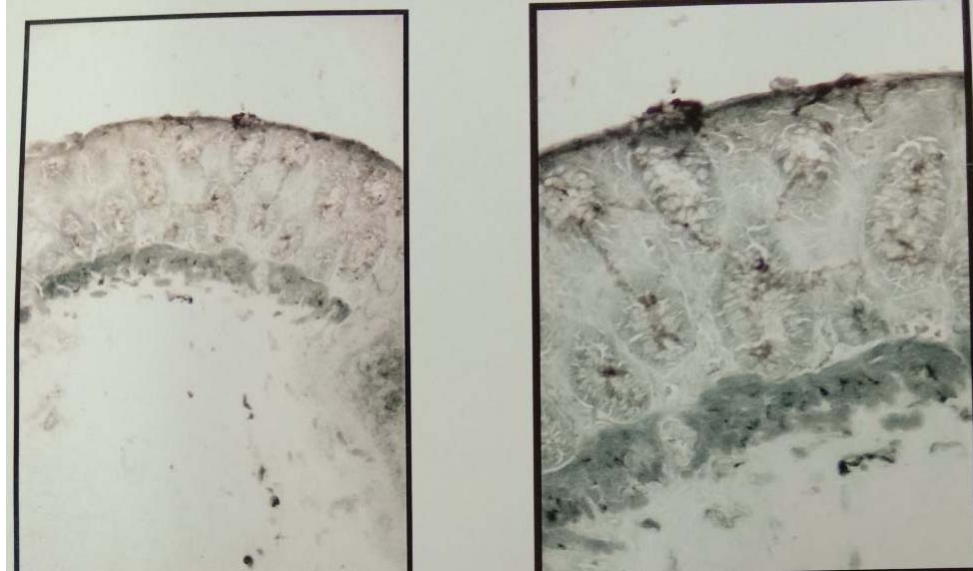
Pattern B



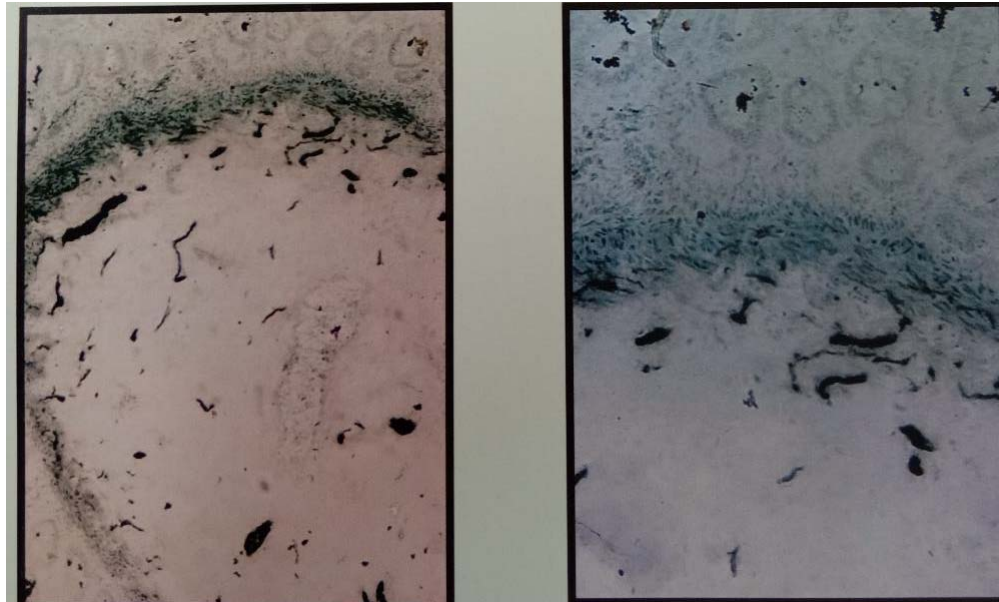
Negative staining



Equivocal pattern without hypertrophied nerve bundles



Equivocal pattern with hypertrophied nerve bundles



Modifications of Ach E staining

Original Method- Karnovsky and Roots

Rapid AchE - 40 min



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[Original Article](#)

Volume 16-5, September/October 2003

A modified technique for the diagnosis of Hirschsprung disease from rectal biopsies: [\[PDF\]](#)

M. K. BABU, USHA KINI, KANISHKA DAS, ANAND ALLADI, ASHLEY J. D'CRUZ

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Problems in AchE

False negative staining-

- Neonates - abnormal twigs may not be fully developed
- Total colonic aganglionosis
- Ultra short HD
- Technical factors
- Poor orientation of small biopsies

False positive staining-

- Hemorrhagic specimens
- Colitis
- Intestinal neuronal dysplasia

Immunohistochemical stains that have been used on frozen section

- Synaptophysin
- Tubulin
- Neurofilament
- Peripherin
- NCAM/CD 56

