

Chronische Intestinale Pseudo-Obstructie (CIPO)

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Chronische Intestinale Pseudo-Obstructie (CIPO)

Acute Intestinale Pseudo-Obstructie

- Post-operatieve ileus
- Acute colon-atonie (Ogilvie)

Chronic Intestinal Pseudo-obstruction

First described in 1958

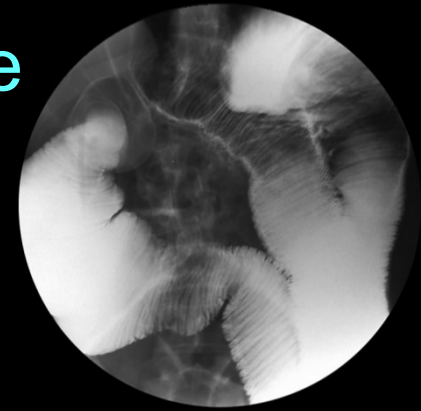
Dudley HAF *et al.* J R Coll Surg Edinb 1958;3:206-217

- Recurrent or chronic symptoms and signs of intestinal obstruction
 - abdominal pain
 - abdominal distension
 - nausea and vomiting
 - clinical signs of ileus
 - radiographic signs of ileus
- No mechanical impediment:
 - radiography
 - laparotomy/laparoscopy



Chronische Intestinale Pseudo-Obstructie

een heterogeen syndroom



Voorkomen

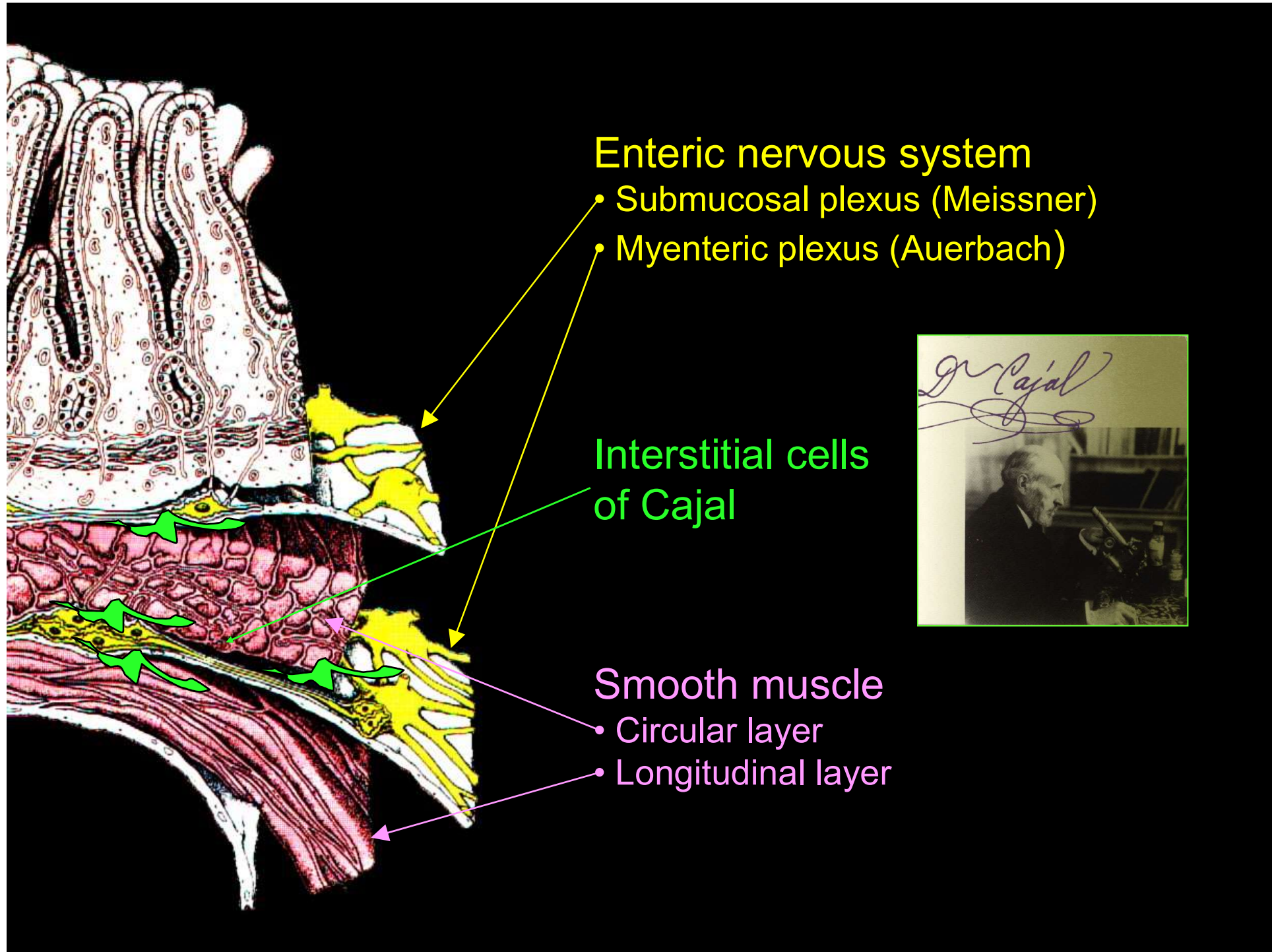
- sporadisch
- erfelijk

Etiologie

- idiopathisch
- secundair aan andere aandoening

Histopathologie

- myopathie
- neuropathie
- 'mesenchymopathie' (ICCs)



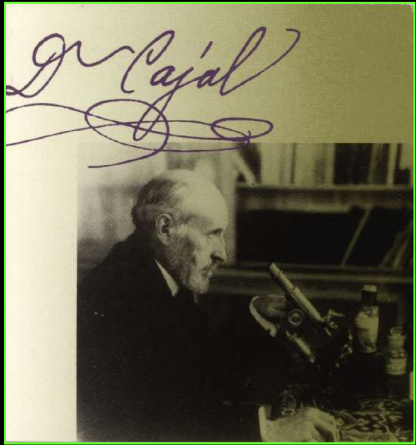
Enteric nervous system

- Submucosal plexus (Meissner)
- Myenteric plexus (Auerbach)

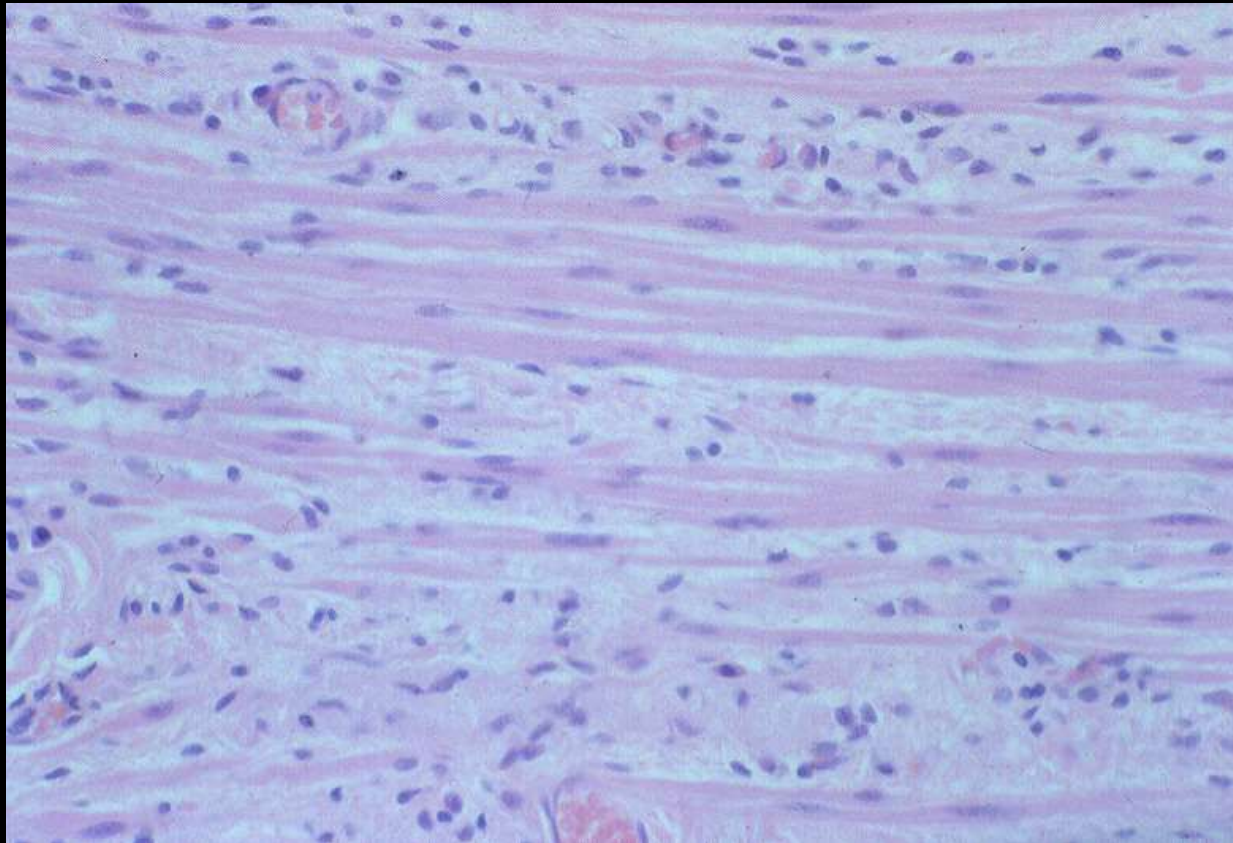
Interstitial cells of Cajal

Smooth muscle

- Circular layer
- Longitudinal layer



Degeneration of intestinal smooth muscle in a patient with idiopathic intestinal pseudo-obstruction syndrome



The London classification of gastrointestinal neuromuscular pathology

Knowles CH *et al.* Gut 2010;59:882-887

1. Neuropathies

- 1.1 Absent neurons
 - 1.1.1 Aganglionosis*
- 1.2 Decreased numbers of neurons
 - 1.2.1 Hypoganglionosis
- 1.3 Increased numbers of neurons
 - 1.3.1 Ganglioneuromatosis†
 - 1.3.2 IND, type B‡
- 1.4 Degenerative neuropathy§
- 1.5 Inflammatory neuropathies
 - 1.5.1 Lymphocytic ganglionitis¶
 - 1.5.2 Eosinophilic ganglionitis
- 1.6 Abnormal content in neurons
 - 1.6.1 Intraneuronal nuclear inclusions
 - 1.6.2 Megamitochondria
- 1.7 Abnormal neurochemical coding**
- 1.8 Relative immaturity of neurons
- 1.9 Abnormal enteric glia
 - 1.9.1 Increased numbers of enteric glia

2. Myopathies

- 2.1 Muscularis propria malformations††
- 2.2 Muscle cell degeneration
 - 2.2.1 Degenerative leiomyopathy/‡‡
 - 2.2.2 Inflammatory leiomyopathy
 - 2.2.2.1 Lymphocytic leiomyositis
 - 2.2.2.2 Eosinophilic leiomyositis
- 2.3 Muscle hyperplasia/hypertrophy
 - 2.3.1 Muscularis mucosae hyperplasia
- 2.4 Abnormal content in myocytes
 - 2.4.1 Filament protein abnormalities
 - 2.4.1.1 Alpha-actin myopathy§§
 - 2.4.1.2 Desmin myopathy
 - 2.4.2 Inclusion bodies
 - 2.4.2.1 Polyglucosan bodies
 - 2.4.2.2 Amphophilic
 - 2.4.2.3 Megamitochondria¶¶
- 2.5 Abnormal supportive tissue
 - 2.5.1 Atrophic desmosis***

3. ICC abnormalities (enteric mesenchymopathy)

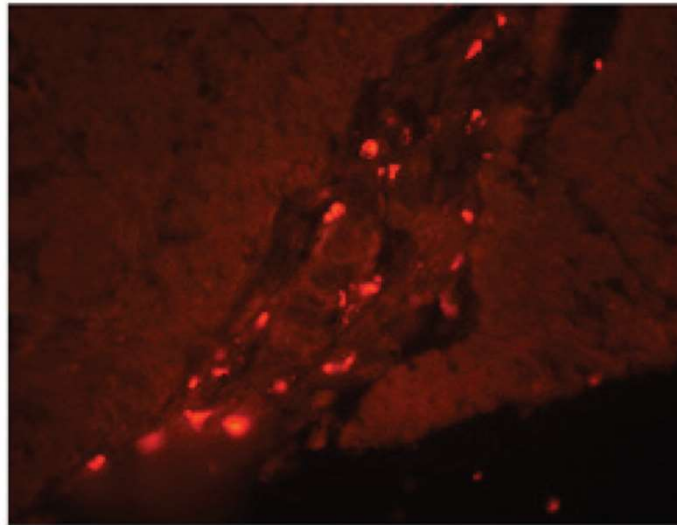
- 3.1 Abnormal ICC networks†††

JC virus infects the enteric glia of patients with chronic idiopathic intestinal pseudo-obstruction

M Selgrad,¹ R De Giorgio,² L Fini,¹ R F Cogliandro,² S Williams,³ V Stanghellini,²
G Barbara,² M Tonini,⁴ R Corinaldesi,² R M Genta,⁵ R Domiati-Saad,³ R Meyer,³ A Goel,¹
C R Boland,¹ L Ricciardiello¹

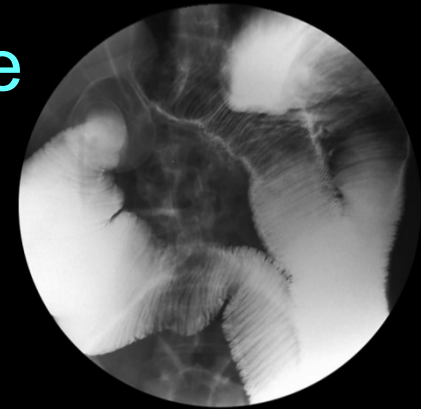
Gut 2009;**58**:25–32.

JCV



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- sporadisch
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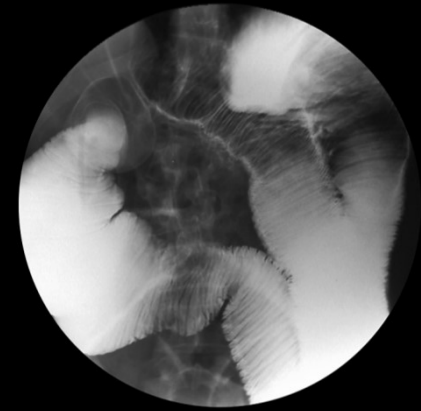
Histopathologie

- myopathie
- neuropathie
- 'mesenchymopathie' (ICCs)

- Sclerodermie
- SLE
- MEN 2B syndroom
- Dystrofia myotonica
- Paraneoplastisch

Chronic Intestinal Pseudo-obstruction

Affected organs

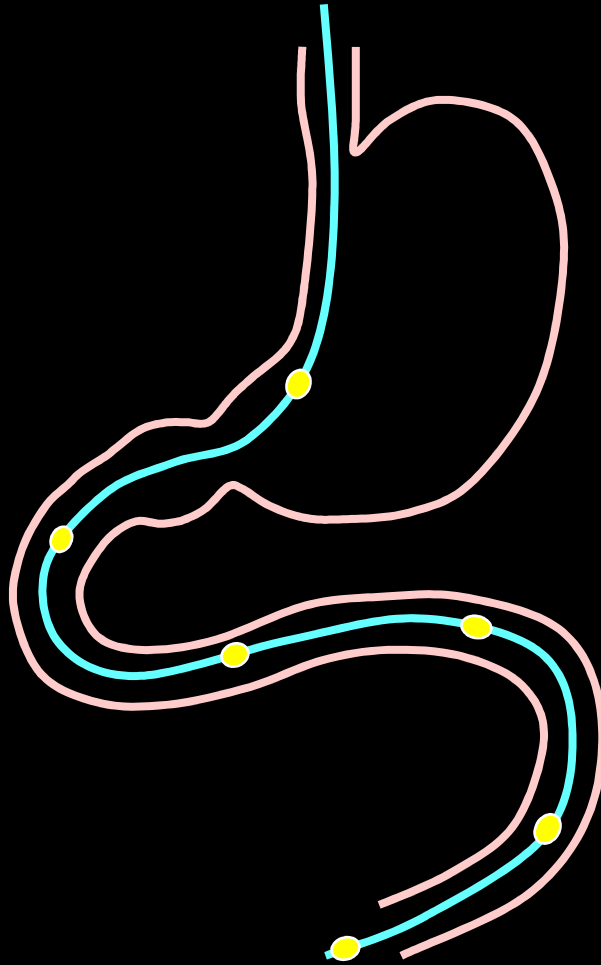


Small intestine

plus:

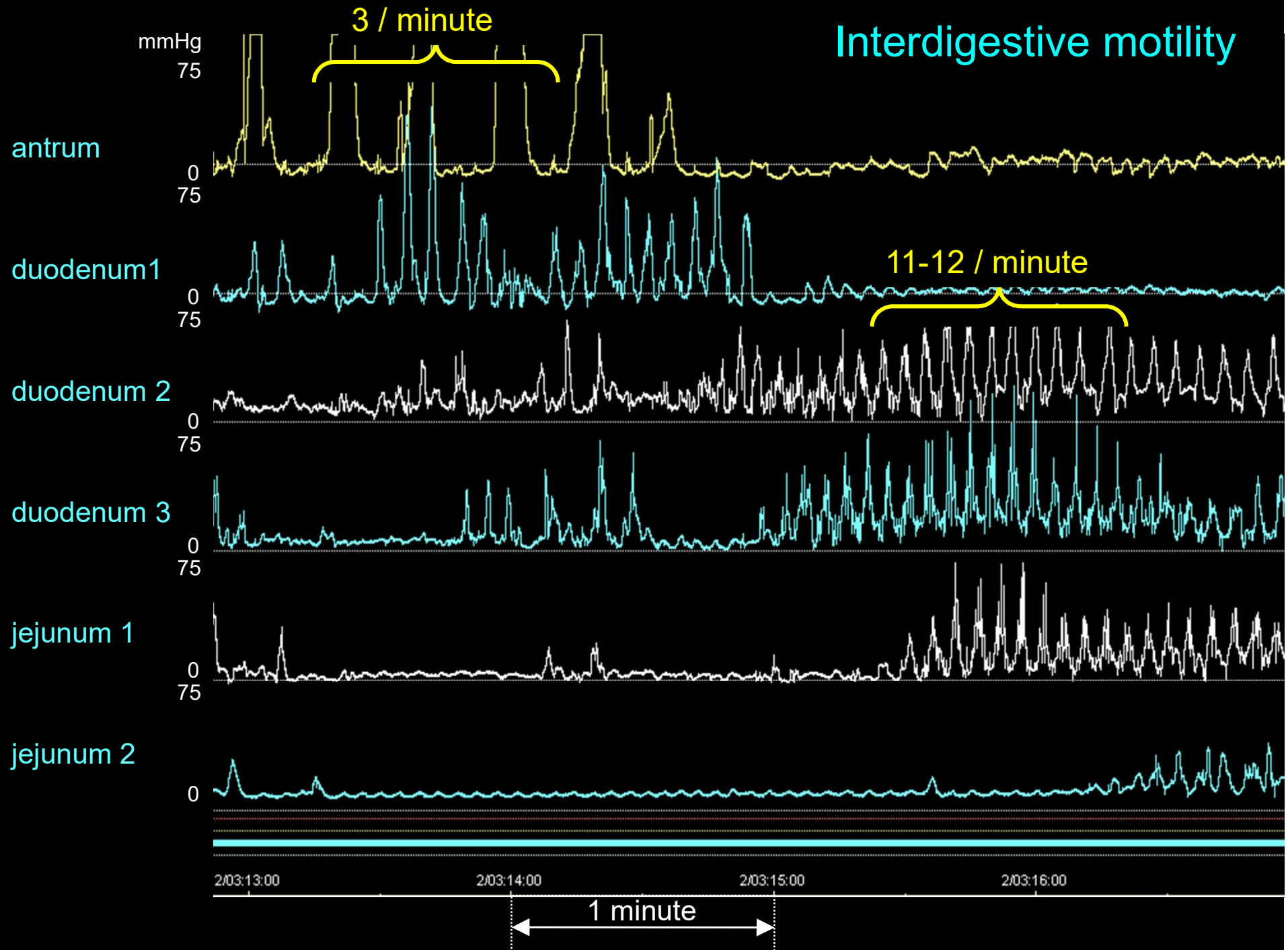
- Stomach
- Oesophagus
- Colon
- Gallbladder
- Smooth muscle elsewhere
 - Urinary bladder
 - Ureters
 - Uterus
 - M. constrictor pupillae

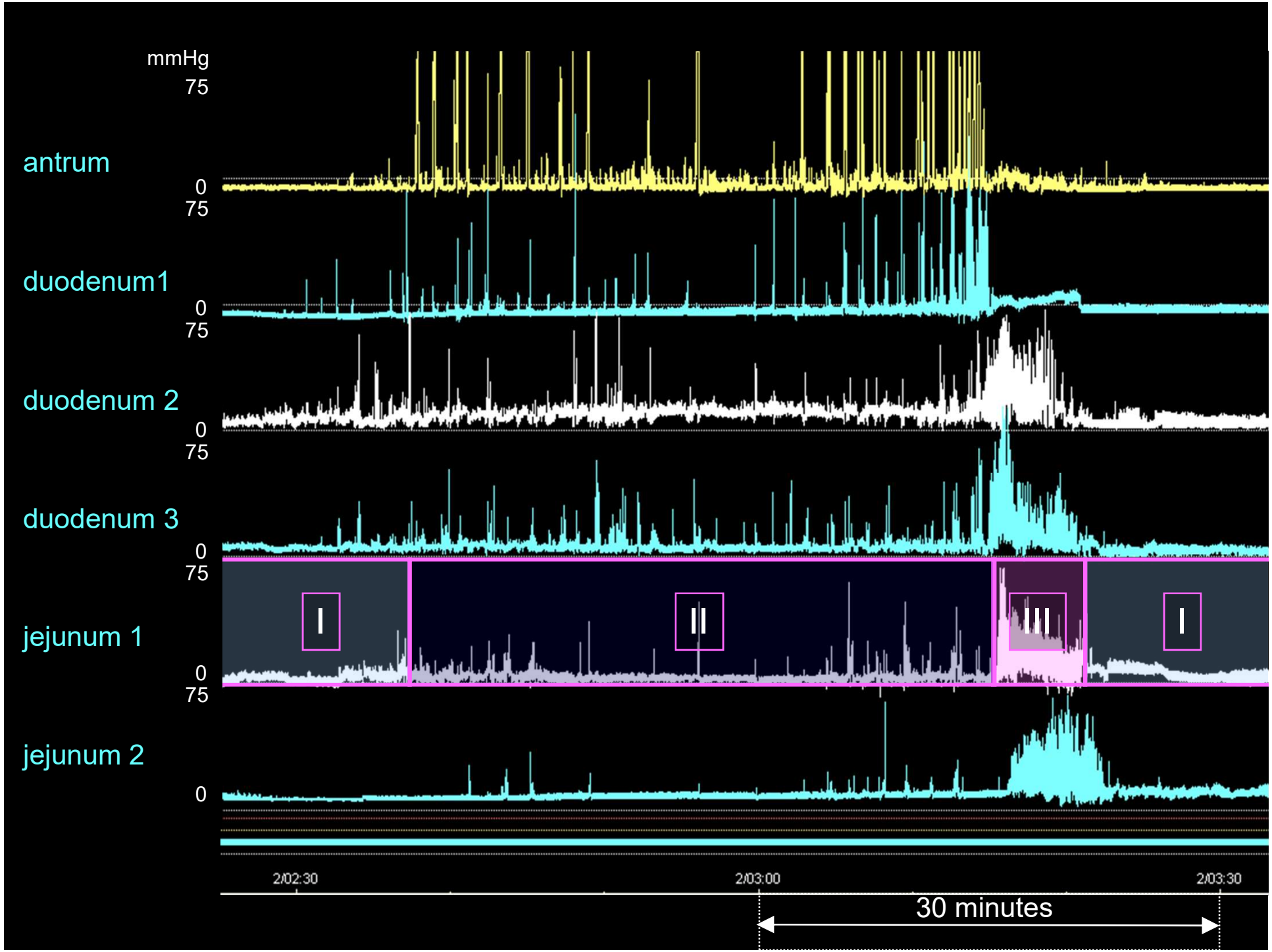
Antroduodenojejunal manometry

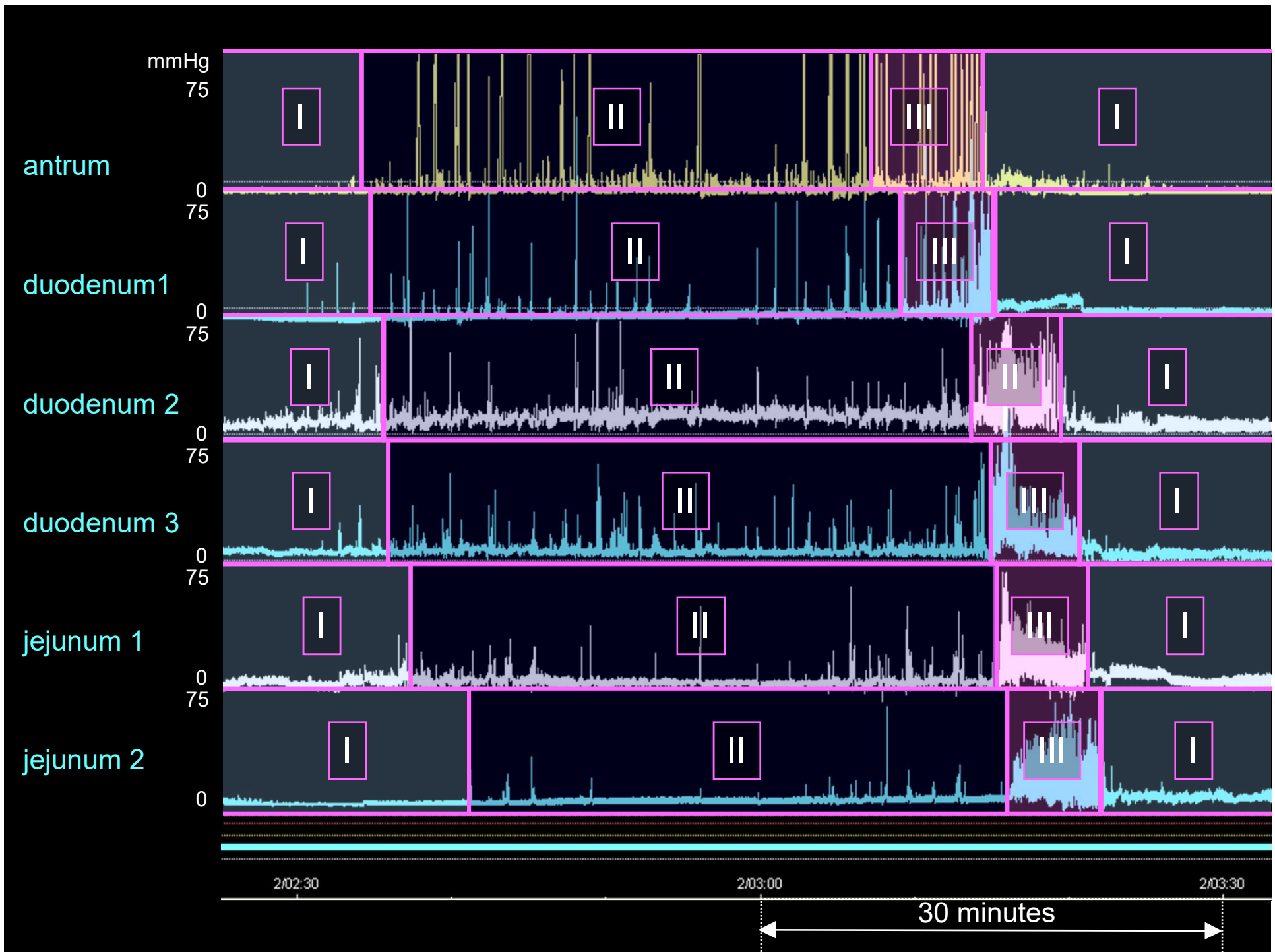


- Sufficiently long fasting period (night)
- At least one normal meal

Interdigestive motility

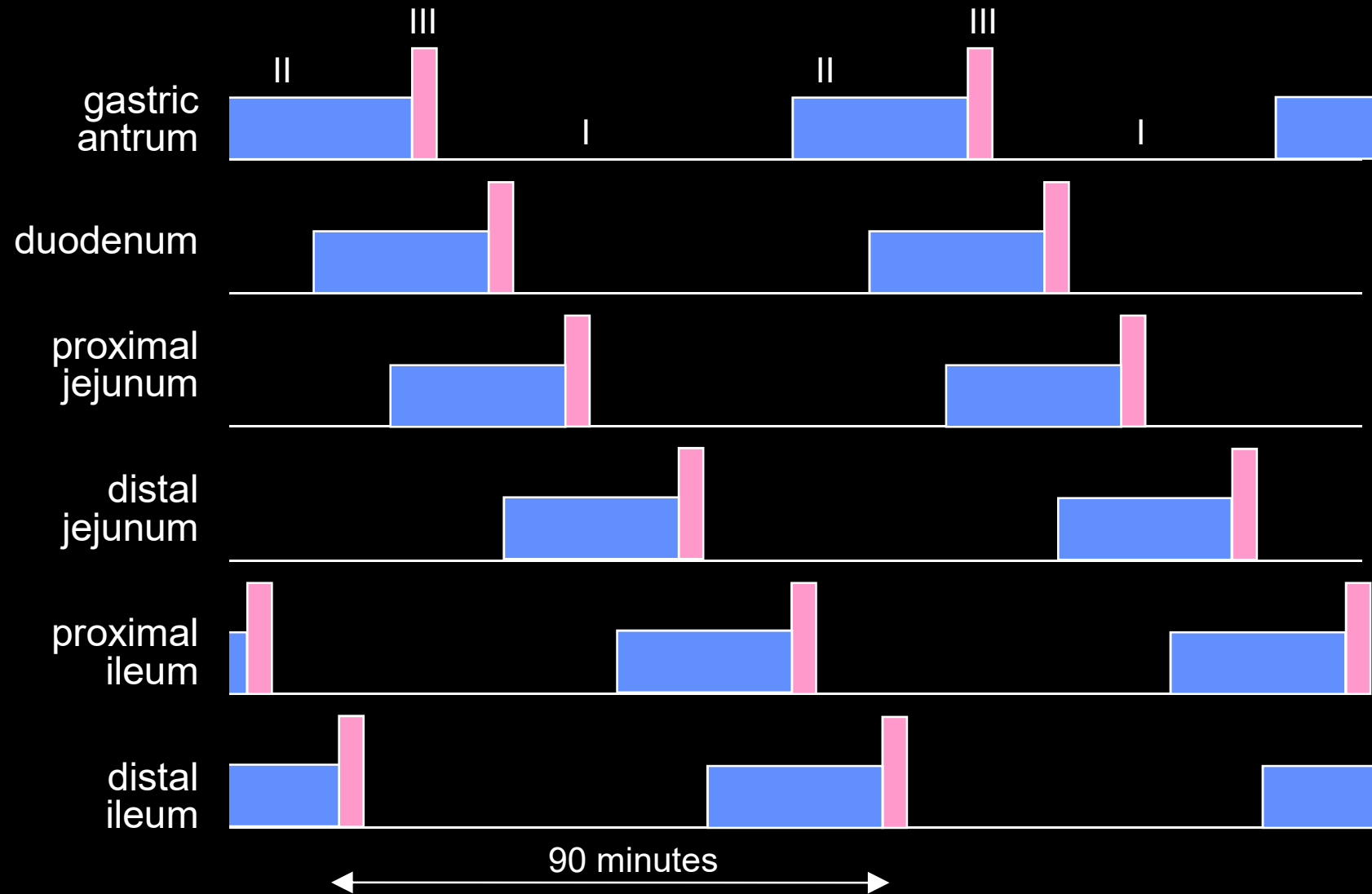




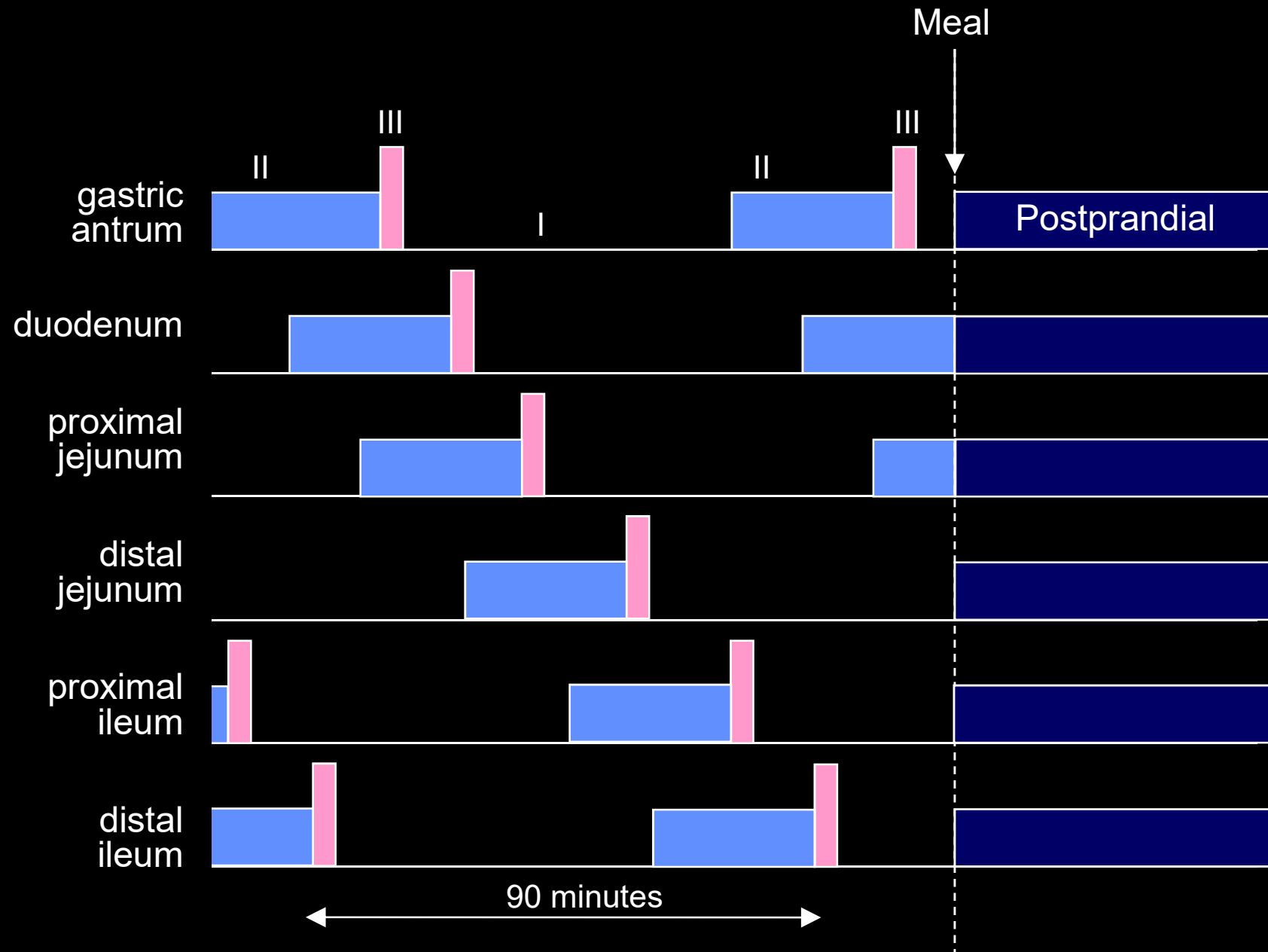


Interdigestive migrating motor complex

Housekeeper of the GI tract

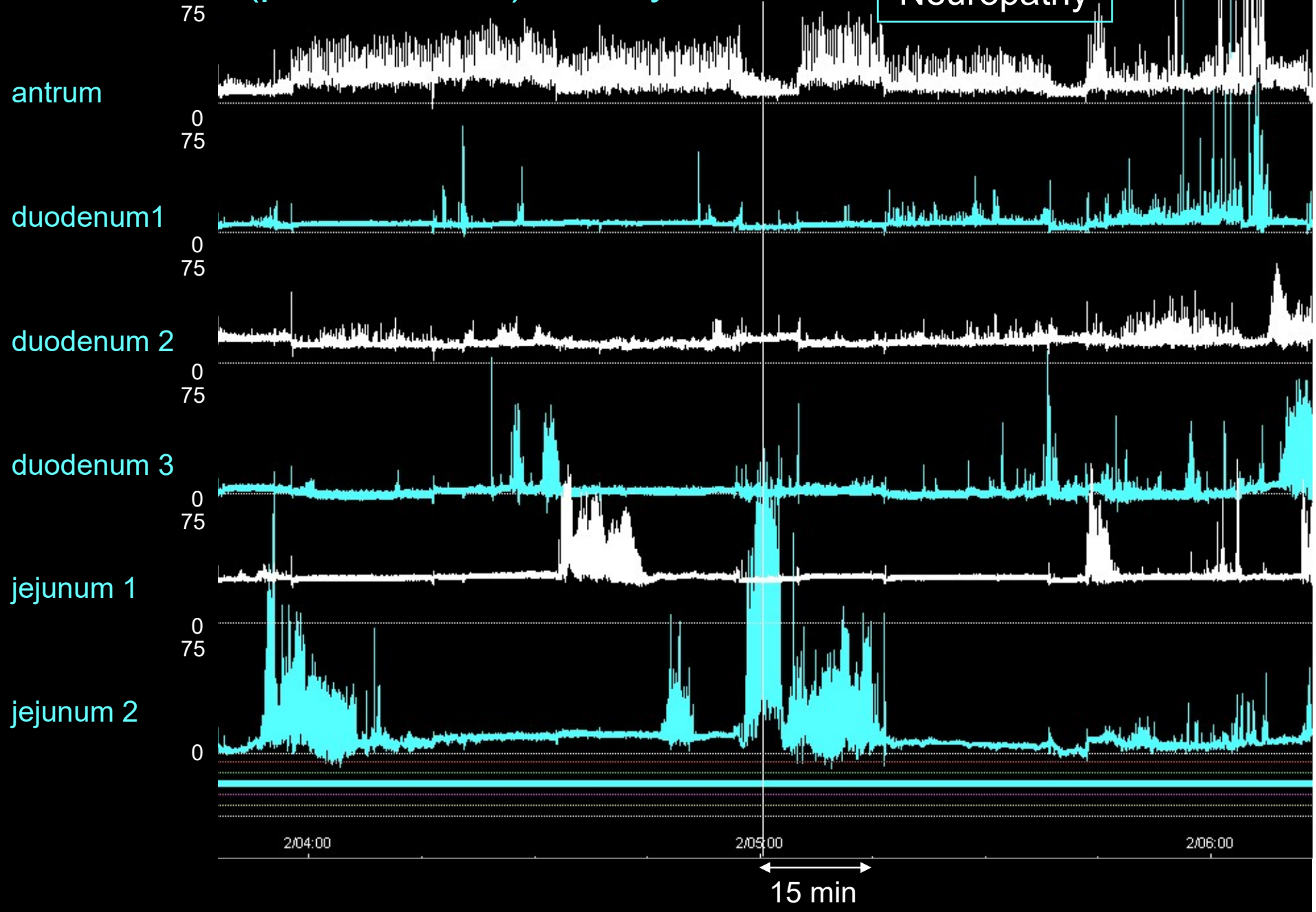


Food interrupts the interdigestive migrating motor complex



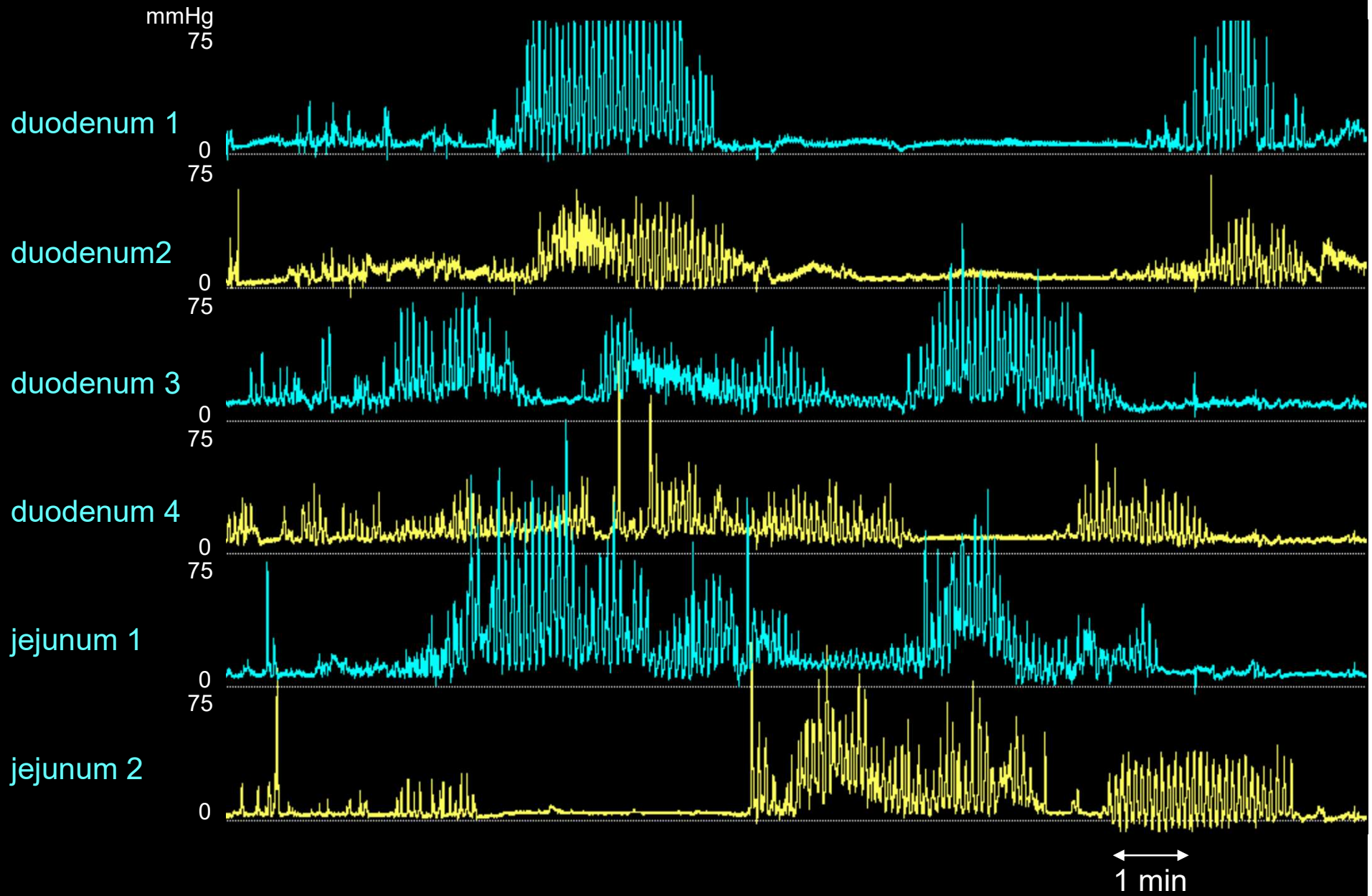
Bursts (phase II-like) activity

'Neuropathy'

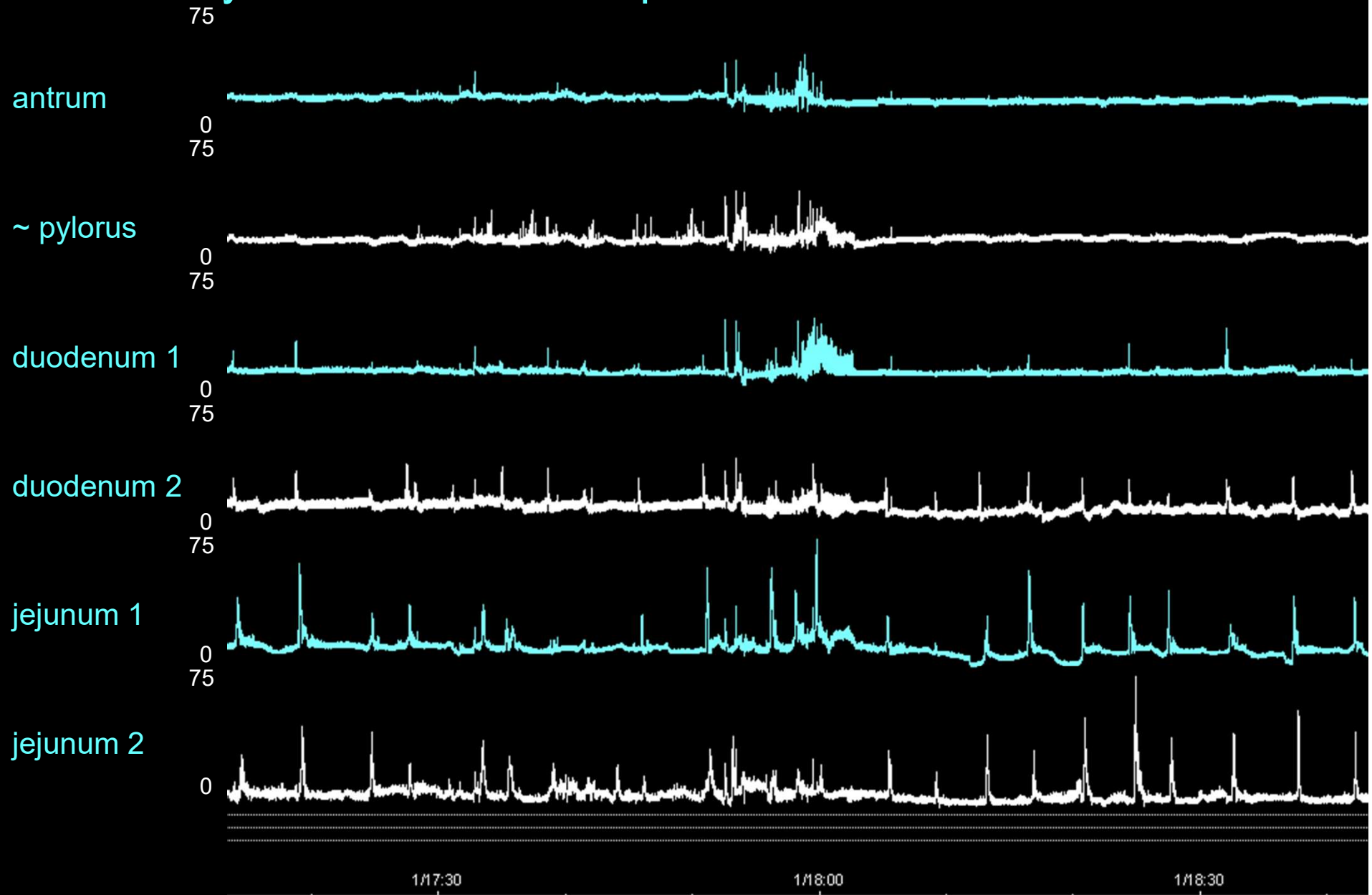


Abnormal phase III propagation

'Neuropathy'

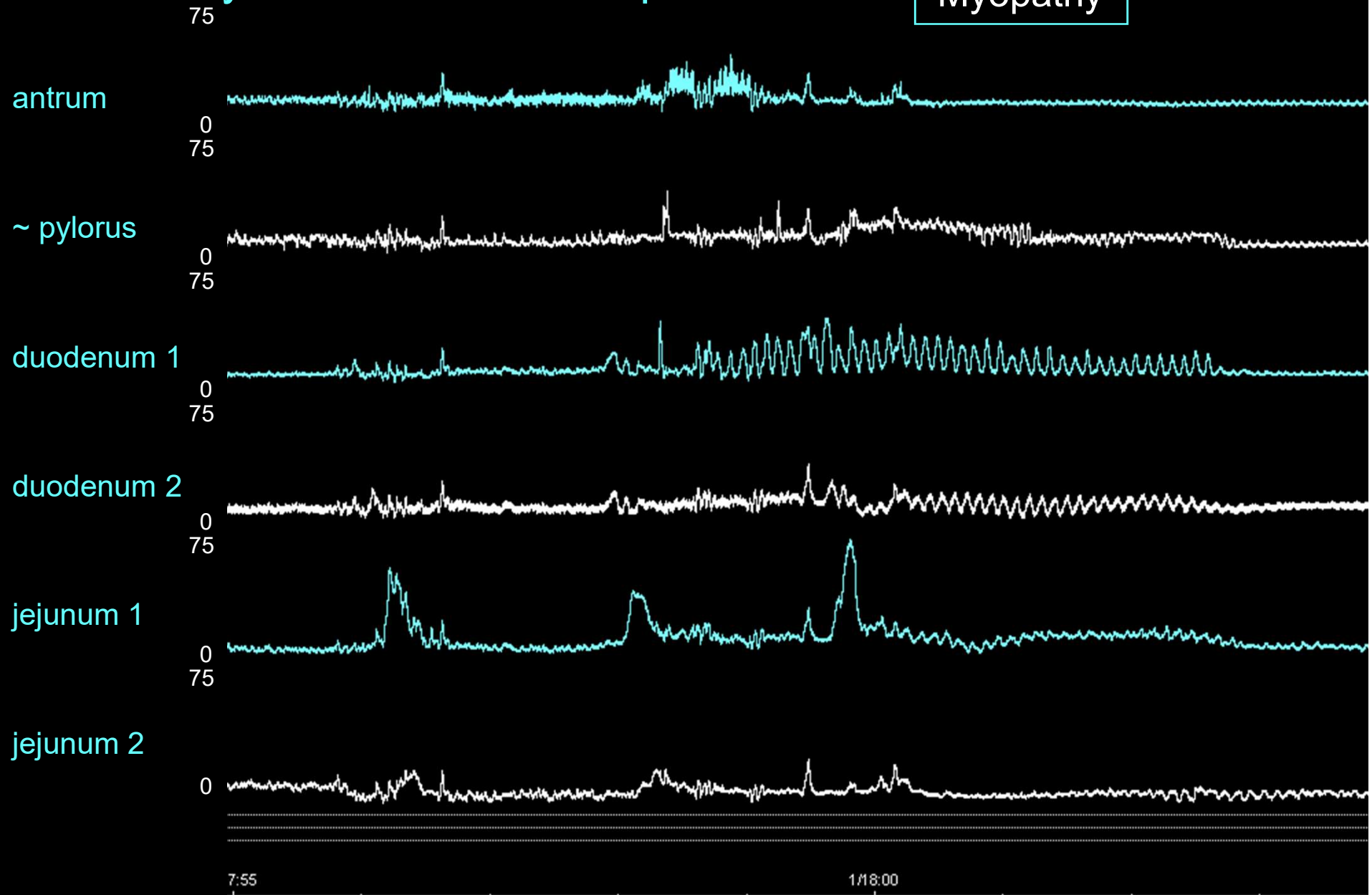


Abnormally low contraction amplitudes

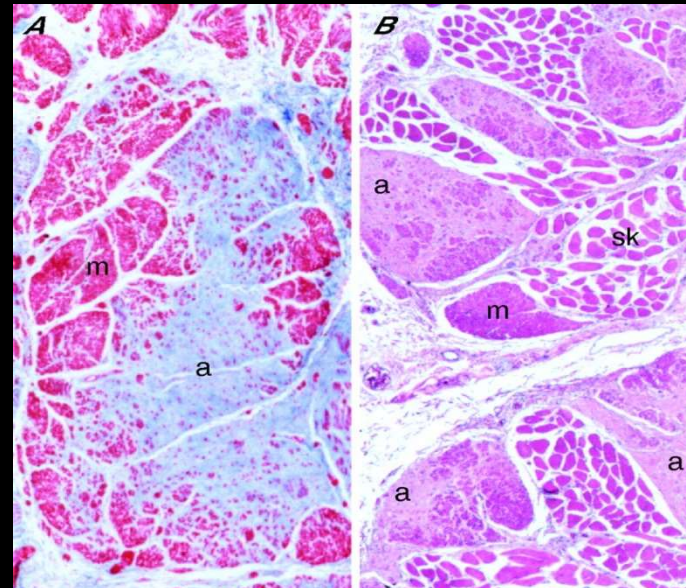


Abnormally low contraction amplitudes

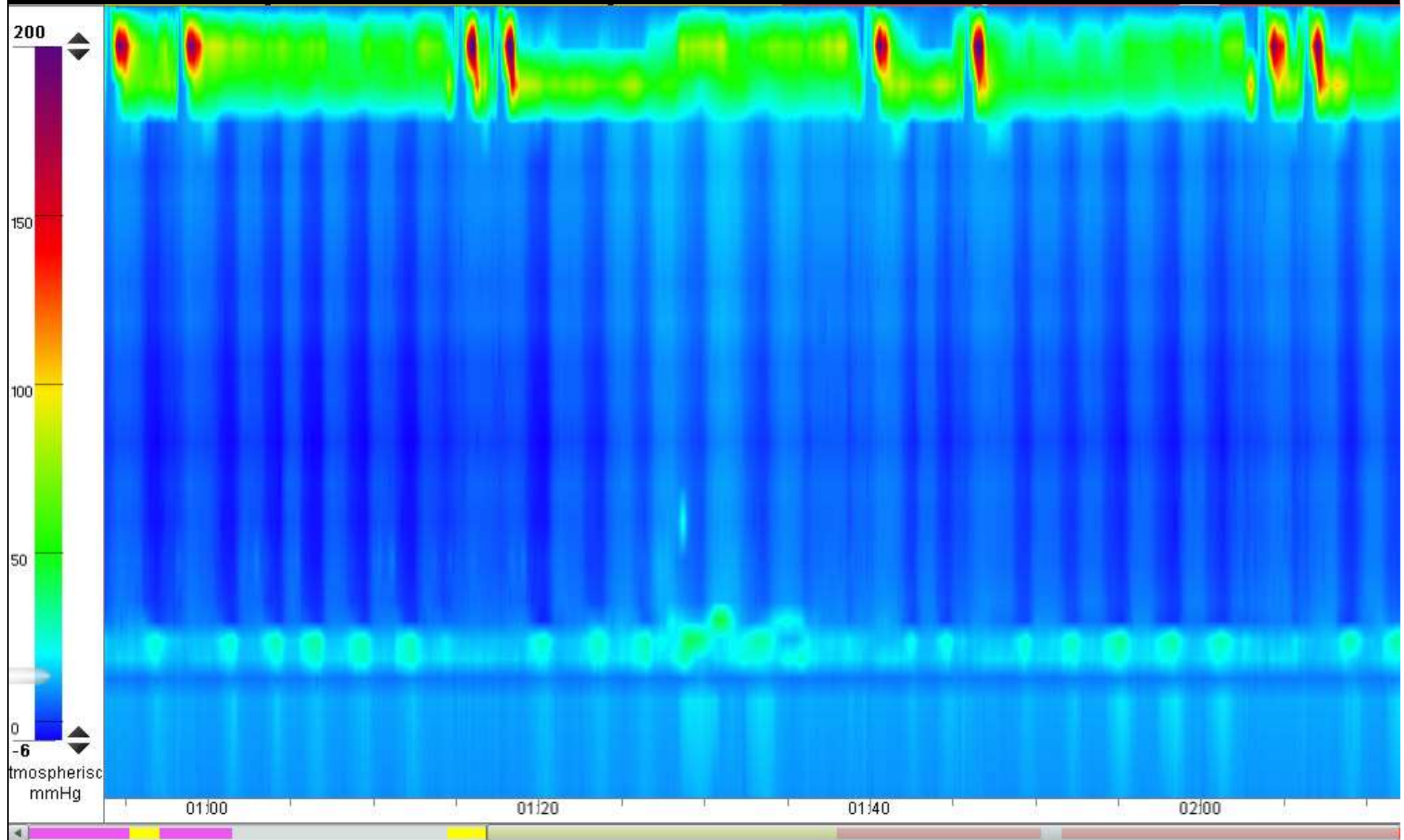
'Myopathy'



systemische sclerose

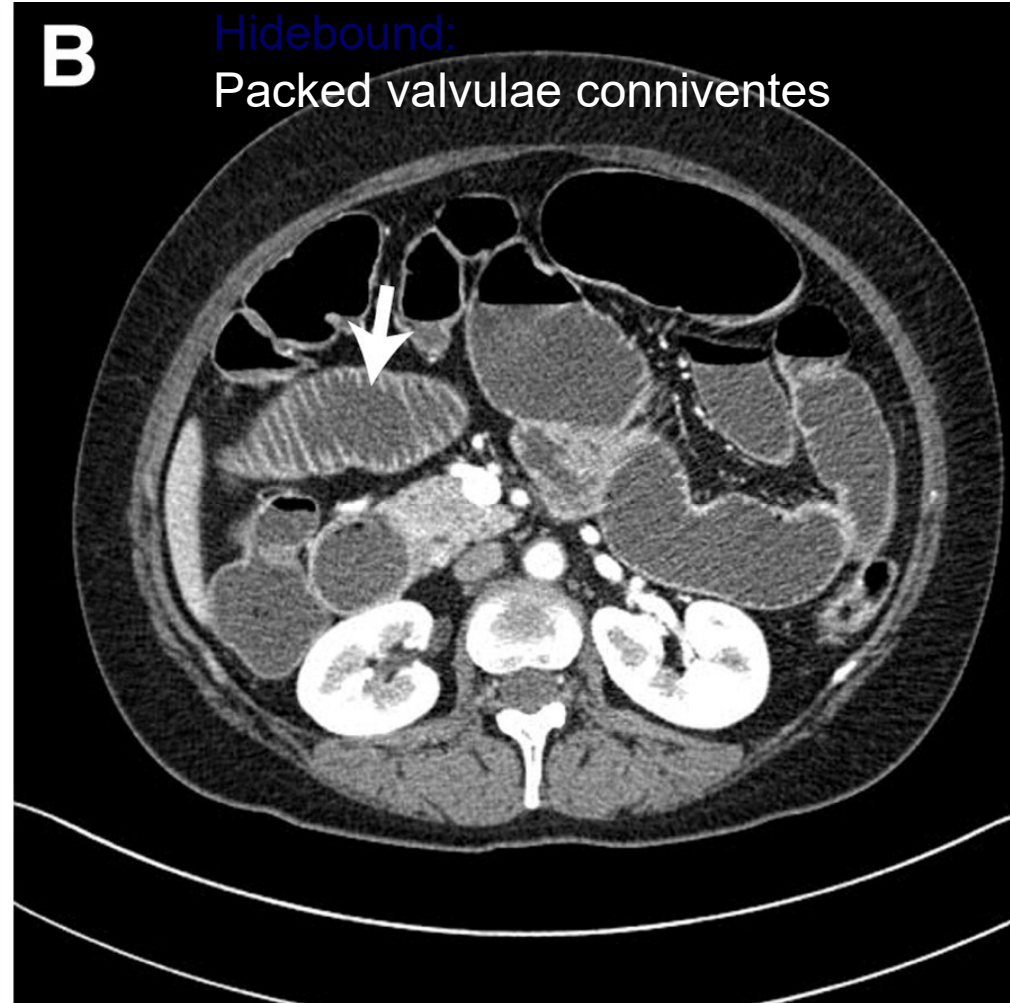
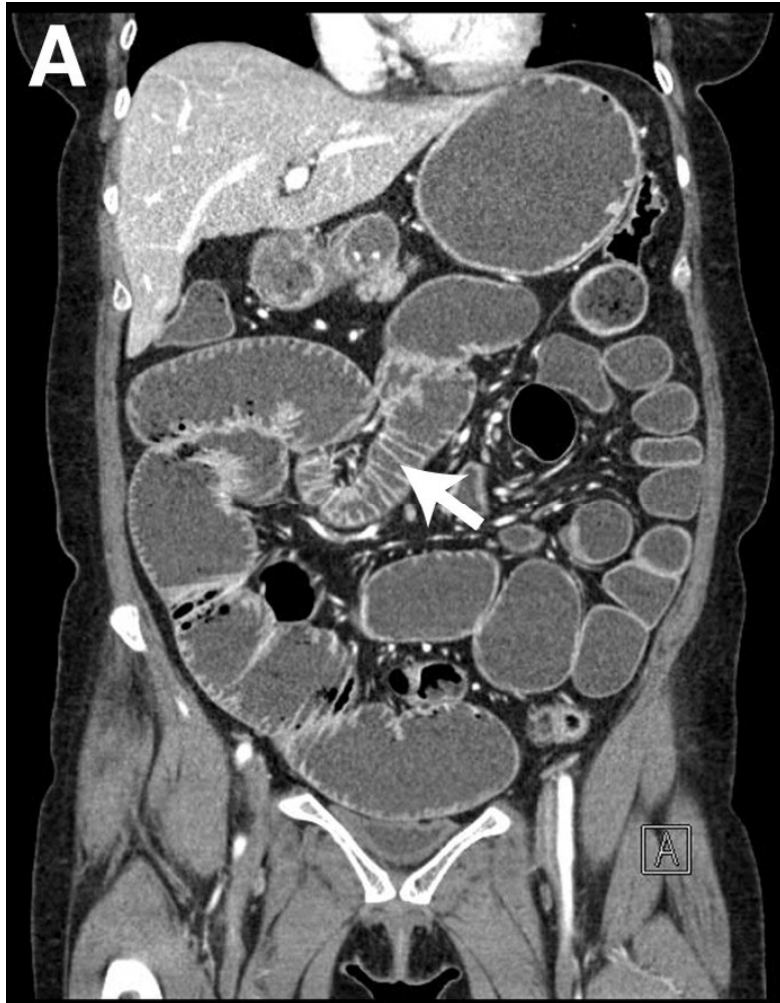


Complete aperistalsis, low LES pressure:
consistent with scleroderma



The “hidebound” bowel sign

Hidebound:
adjective denoting malnourished
condition of cattle



pneumatosis intestinalis
in a patient with systemic sclerosis



Mw D., 64 jaar

Sinds 1 jaar:
Progressieve buikklachten
Ileus
Geen obstructie aantoonbaar
TPV-afhankelijk

Antroduodenale manometrie:
neuropathie



Welke antistoffen zou u aanvragen ?

ROOD: Anti-Hu

GROEN: Anti-ANNA-1

Mw D., 64 jaar

Sinds 1 jaar:
Progressieve buikklasten
Ileus
Geen obstructie aantoonbaar
TPV-afhankelijk

Antroduodenale manometrie:
neuropathie

Anti-Hu +++++

Paraneoplastische CIPO



Welke tumor is het meest waarschijnlijk?

ROOD: kleincellig longcarcinoom

GROEN: ovariumcarcinoom

Aan welke aandoening lijdt deze vrouw?

ROOD: Neurofibromatose type 1

GROEN: MEN-2B



MEN 2B



Eversion of upper eyelid
Prominent eyebrows



“Blubbery” lips



MEN 2B

Multiple endocrine neoplasia

- Benigne tumoren mondholte
- Medullair schildkliercarcinoom
- Feochromocytoom
- Obstipatie
- CIPO

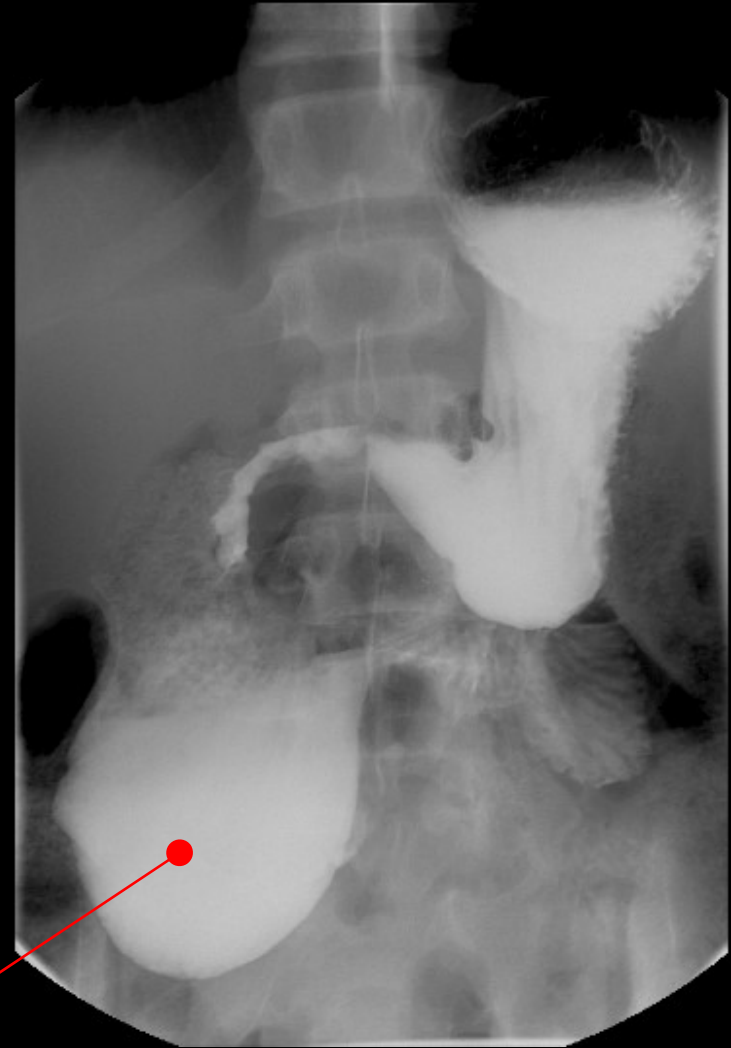
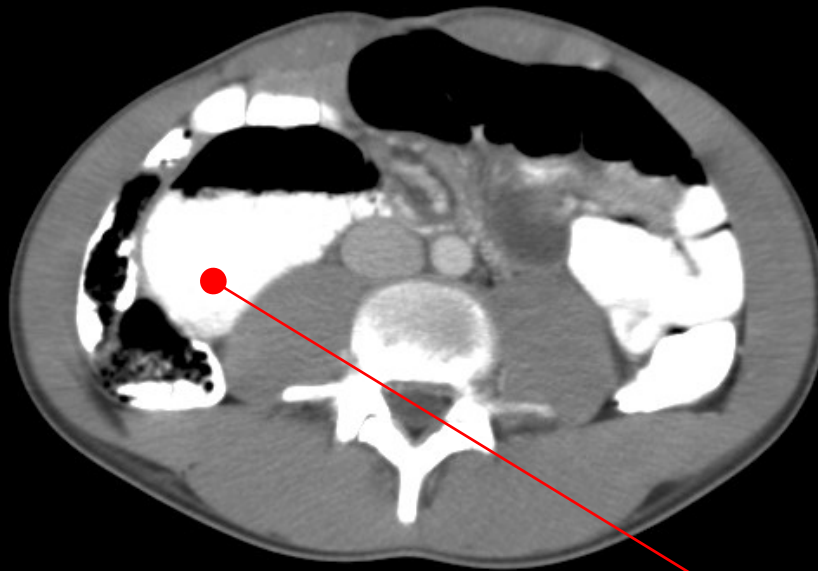


Mw V, 31 jaar, MEN 2B
Status na subtotale colectomie

Hugo, 18 jaar

“mijn tante heeft precies dezelfde klachten”

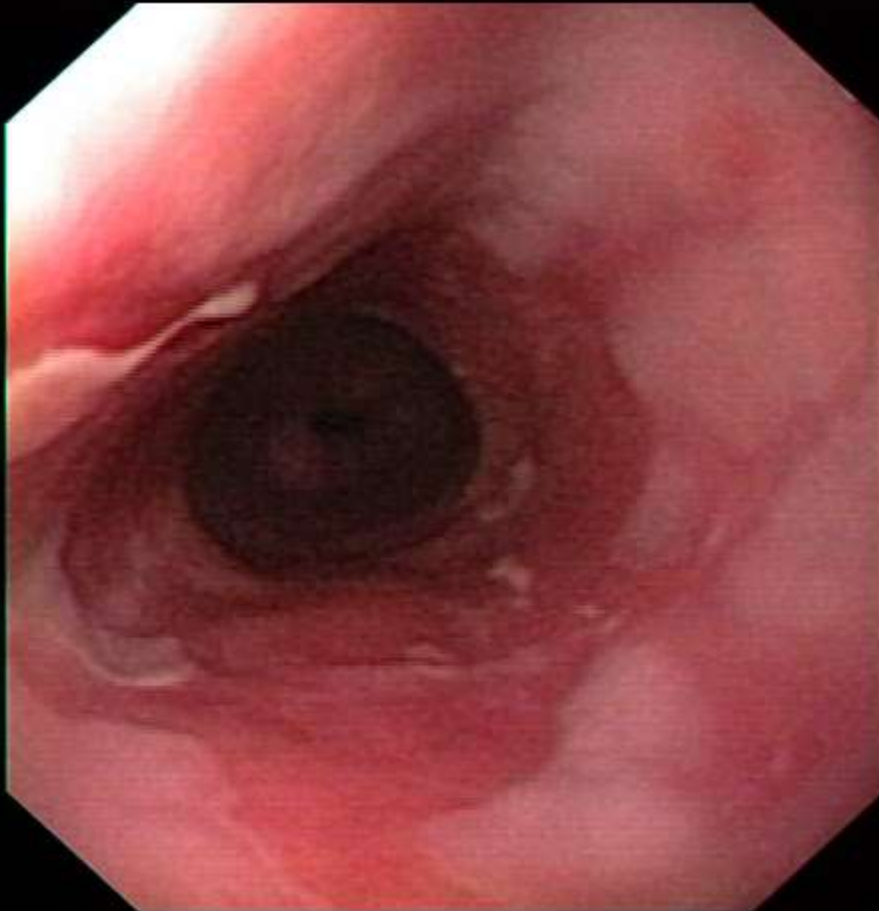
- refluxziekte
- Barrett
- antireflux-chirurgie?
- aperistalsis slokdarm



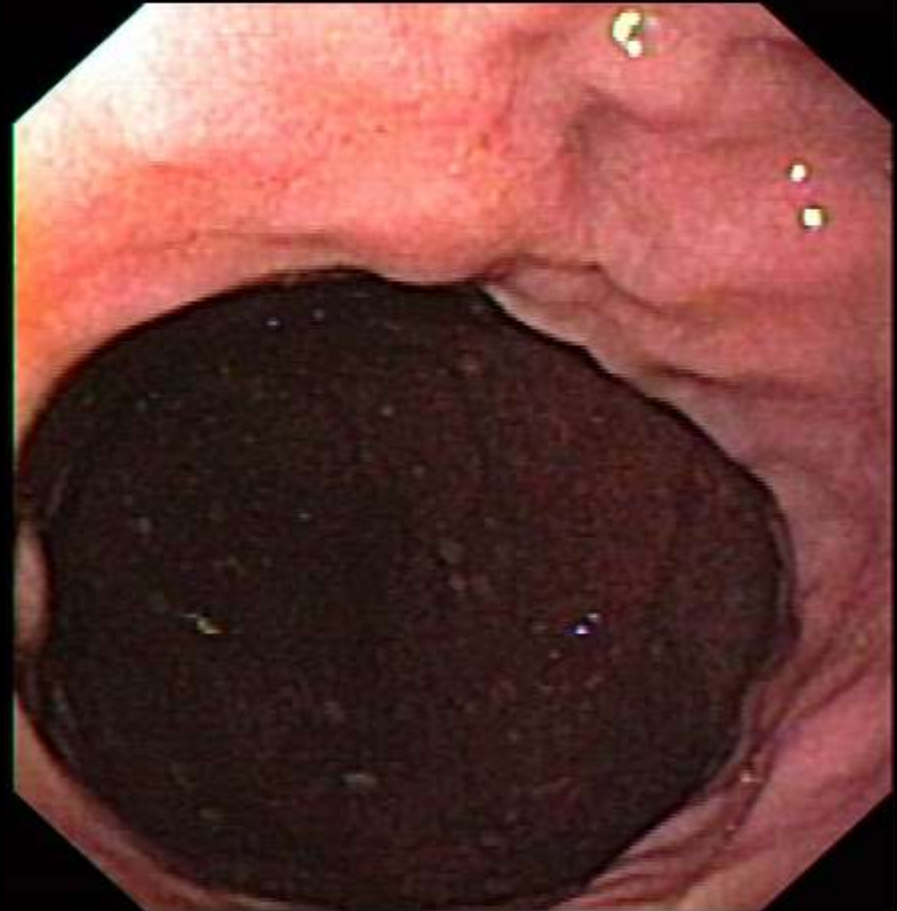
megaduodenum

Hugo's tante, 31 jaar

Barrett



megaduodenum



Familial visceral myopathy with pseudo-obstruction, megaduodenum, Barrett's esophagus and cardiac abnormalities

Mungan Z *et al.* Am J Gastroenterol 2003;98:2556-2560



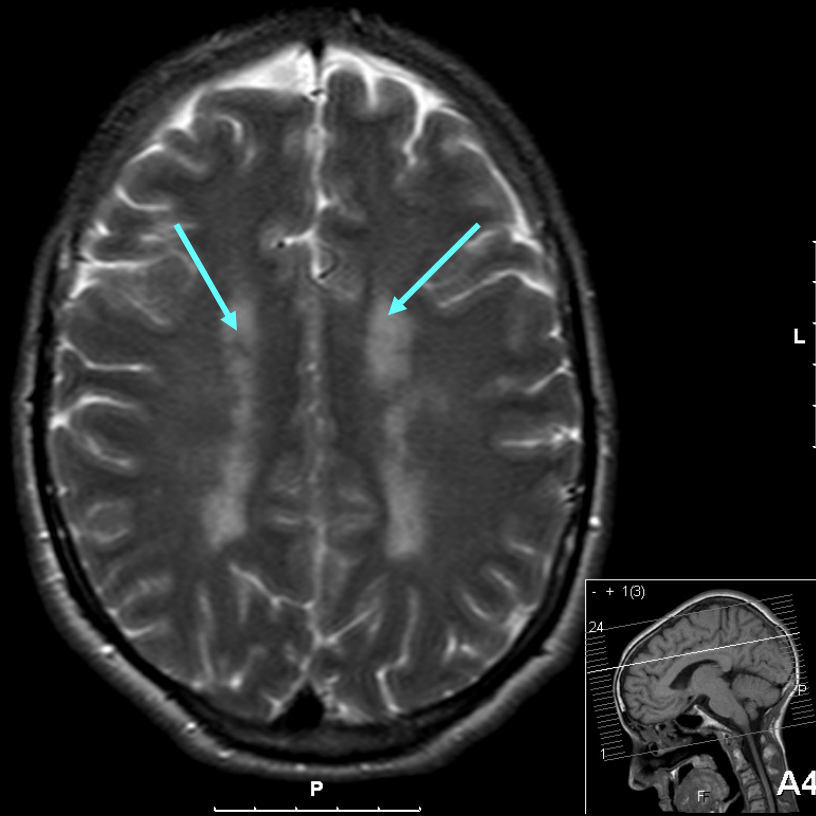
3 children from consanguineous parents:

- Gastric atony
- Megaduodenum
- Esophageal aperistalsis
- Long-segment Barrett's esophagus
- Cardiac abnormalities:
 - Trivial supra-valvular pulmonary stenosis (2)
 - Membranous VSD (1)

Mr B, 39 years



periventricular white matter changes (leukoencephalopathy)



MNGIE

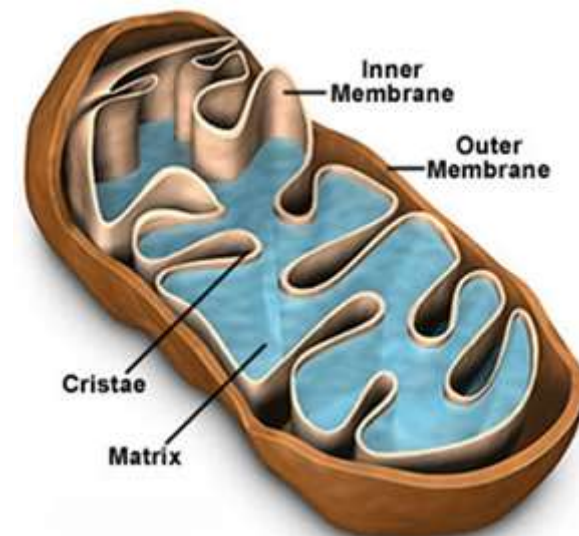
Mitochondrial Neurogastrointestinal Encephalomyopathy

- autosomal recessive disease
- caused by mutation in the ECGF1 gene
encoding thymidine phosphorylase (TP)
located on chromosome 22q13.32-qter
- leading to accumulation of thymidine and deoxyuridine
- → defects in mitochondrial DNA

Onset of symptoms < age 20 in 60%
Death in third or fourth decade

Therapeutic options

- hemodialysis
- peritoneal dialysis
- allogeneic stem cell transplantation



'Enteric dysmotility'

Wingate D et al. J Gastroenterol Hepatol 2002;17;S1-S4

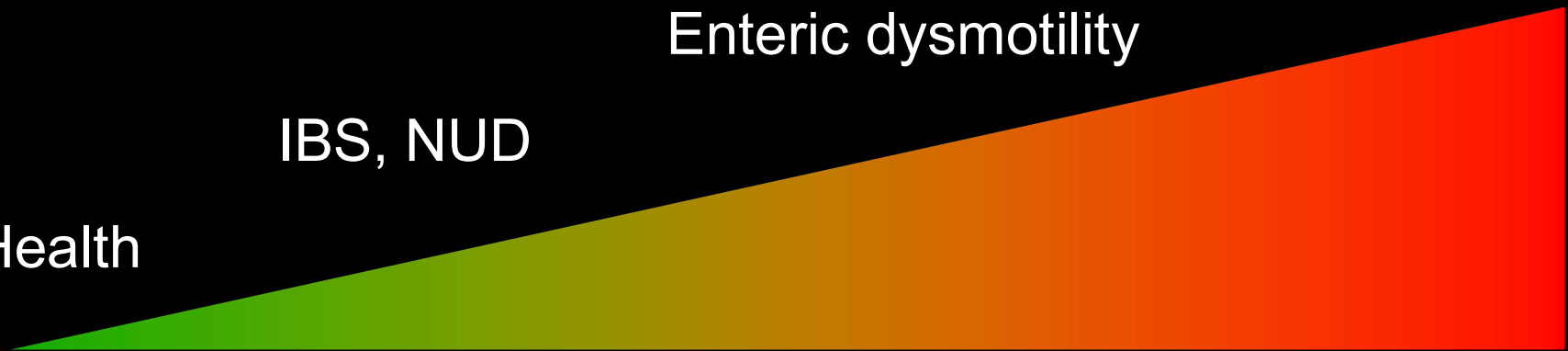
- Intermittent abdominal distension, cramping, nausea, vomiting, weight loss
- No organic cause
- No bowel distension, no air-fluid levels
- Manometric abnormalities as in pseudoobstruction

Health

IBS, NUD

Enteric dysmotility

Pseudo-obstruction



Chronic Intestinal Pseudo-obstruction

Treatment

Nutritional support

- oral
- nasogastric tube / PEG
- nasojejunal tube / PEJ / surgical jejunostomy
- parenteral

Antibiotics

- tetracyclin / ciprofloxacin / metronidazole (rotating)
- drug-free intervals

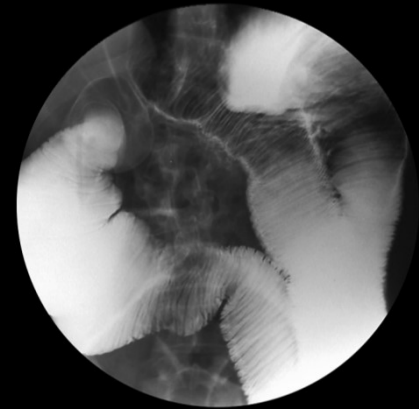
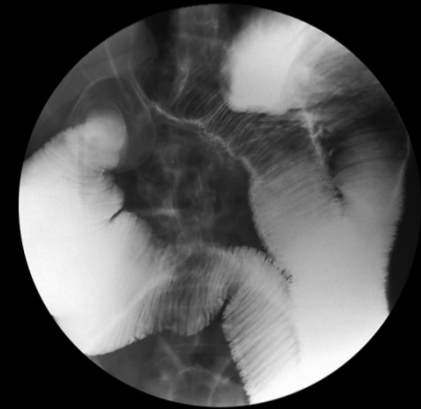


Table 1 Antibiotics to be considered in the treatment of small intestine bacterial overgrowth

Agent	Dose
Tetracycline	250 mg 4 times daily
Doxycycline	100 mg twice daily
Minocycline	100 mg twice daily
Amoxicillin-clavulanic acid	875 mg twice daily
Cephalexin +	250 mg 4 times daily
Metronidazole	250 mg 3 times daily
Ciprofloxacin	500 mg twice daily
Norfloxacin	400 mg twice daily
Chloramphenicol	250 mg 4 times daily
Rifaximin	400 mg 3 times daily
Nitazoxanide	500 mg twice daily

Chronic Intestinal Pseudo-Obstruction

Treatment



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- oral
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Prokinetic agents

- domperidone/ metoclopramide
- erythromycine 250 mg tid orally
- octreotide 50 µg s.c. a.n.
- pyridostigmine 30 mg tid orally
- prucalopride 2 mg once daily orally

Octreotide induces interdigestive motility patterns in small bowel of scleroderma patients

Patient with scleroderma

Basal



Soudah HC *et al.* N Engl J Med 1991;325:1461-1467

After 100 μ g of
octreotide

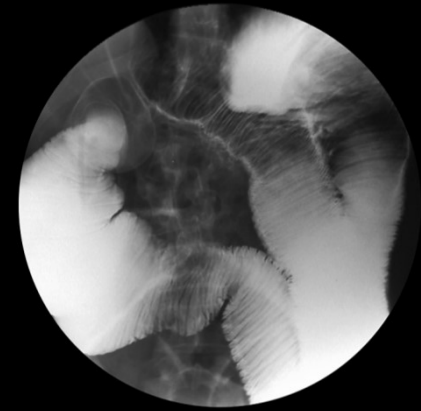


“The Verne Cocktail”

octreotide 50 μ g subcutaneously at bedtime
erythromycin 200 mg suspension per os tid

Verne GN *et al.* Dig Dis Sci 1995;40:1892-1901

Chronic Intestinal Pseudo-obstruction



Surgery

- Resection or bypass of localized small bowel disease
- Colectomy
- Placement of feeding tube
- Placement of venting tube
- Full-thickness biopsy
- Small bowel transplantation

Review artikel

Chronic Intestinal Pseudo-Obstruction

Gabbard SL and Lacy BE

Nutrition in Clinical Practice 2013;28(3):307-316

Om te onthouden

- Denk bij onbegrepen intermitterende/chronische ileus aan CIPO!
- Proeflaparotomie is niet ouderwets
- Laat bij de proeflaparotomie een full-thickness biopsie nemen
- Probeer de patiënt te behoeden voor nodeloze chirurgie
- Behandeling van bacteriële overgroei is vaak effectief