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* بیمار پسر ۲ سال و ۷ ماهه ای است. که مشکل وی از بدو تولد با عدم دفع مکونیوم و اتساع شکم شروع شده. در گرافی ساده ی شکم انسداد روده مطرح شد اما پس ار باریم انما بیمار مدفوع کرد. اتساع شکم و یبوست وی ادامه داشت ولی هر بار با تحریک دفع مدفوع داشته است. در باریم انمای وی در ۸ ماهگی اتساع شدید کادر کولون و در ترانزیت روده ی باریک و سی تی اسکن شکم اتساع معده روده ی باریک و بزرگ با نامنظمی مخاط بدون شواهد انسداد مکانیکی ملاحظه شد.

* در بیوپسی رکتوم گانگلیون عصبی وجود داشت و بیماری هیرشپرونگ رد شد. پس از آن بیمار به طور مکرر دچار حملات استفراغ صفراوی و اتساع شکم و گاه اختلال در دفع ادرار میشد مورد مایع درمانی وریدی TPN آنتی بیوتیک مصرف پروکینتیک های مختلف از جمله سیساپراید دومپریدون اریترومایسین و اکترؤتاید قرار میگرفت.

* بیمار در سن ۲ سال و ۴ ماهگی دچار حمله ی شدید انسداد روده گشت بطوری که تب و بد حالی پیدا کرده و جهت جلوگیری از پرفوراسیون و پریتونیت ناچار به انجام ایلئوستومی شدیم. از آن به بعد به دلیل از دست دادن حجم زیاد مایعات از محل ایلئوستومی بیمار قادر به ترخیص نمیشود. آمپول بوتاکس به ۴ طرف پیلور وی طی اندوسکوپی تزریق شد.

Chronic intestinal pseudo obstruction syn.

*

Sym Para sym

↘ ↙

ENS

neurons – glial cells- Icc

↓

GI muscles

↓

motility

Icc_s are gut pacemaker and modulator between enteric neurons and gut muscles

Onset
Congenital
Acquired
Acute
Gradual
Presentation
Megacystis-microcolon intestinal hypoperistalsis syndrome
Acute neonatal bowel obstruction, with or without megacystis
Chronic vomiting and failure to thrive
Chronic abdominal distention and failure to thrive
Cause
Sporadic
Familial
Toxic
Ischemic
Viral
Inflammatory
Autoimmune
Area of Involvement
Entire gastrointestinal tract
Segment of gastrointestinal tract
Megaduodenum
Small bowel
Colon
Pathology
Myopathy
Neuropathy
Absent neurons
Immature neurons
Degenerating neurons
Intestinal neuronal dysplasia
No microscopic abnormality

Features of chronic intestinal pseudo-obstruction in pediatric patients

Primary pseudo-obstruction
- Visceral myopathy: sporadic or familial
- Visceral neuropathy: sporadic or familial

Secondary pseudo-obstruction: related or associated recognized causes
- Muscular dystrophies
- Scleroderma and other connective tissue diseases
- Postischemic neuropathy
- Postviral neuropathy
- Generalized dysautonomia
- Hypothyroidism
- Diabetic autonomic neuropathy
- Drugs: anticholinergics, opiates, calcium channel blockers, many others
- Severe inflammatory bowel disease
- Organ transplantation
- Amyloidosis
- Chagas' disease
- Fetal alcohol syndrome
- Chromosome abnormalities
- Multiple endocrine neoplasia IIB
- Radiation enteritis

Causes of chronic pseudo-obstruction in children

Cases of sec. CIP

- **Toxic : ketamine**

  Carbamazepine

  Clonidine

  Opiates

  Ca- blockers

  Vinblastine

  Anti cholinergics

- **Immune: celiac**
  SS
  lupus
  Guillain – barre
- **Tumors : neuroblastoma**
  pheochromocytoma
  Thymoma
- **Striated myopathy: myotonic dystrophy**
  Duchenne
  mitochondrial myopathy
- **Neuropathy : DM – amyloidosis – familial dysautonomia**

- **Mis: Angioedema**
  postradiation
  kawasaki

- **Metabolic : electrolyte imbalance**

  hypothyroidism

  hypoparathyroidism

  ↓

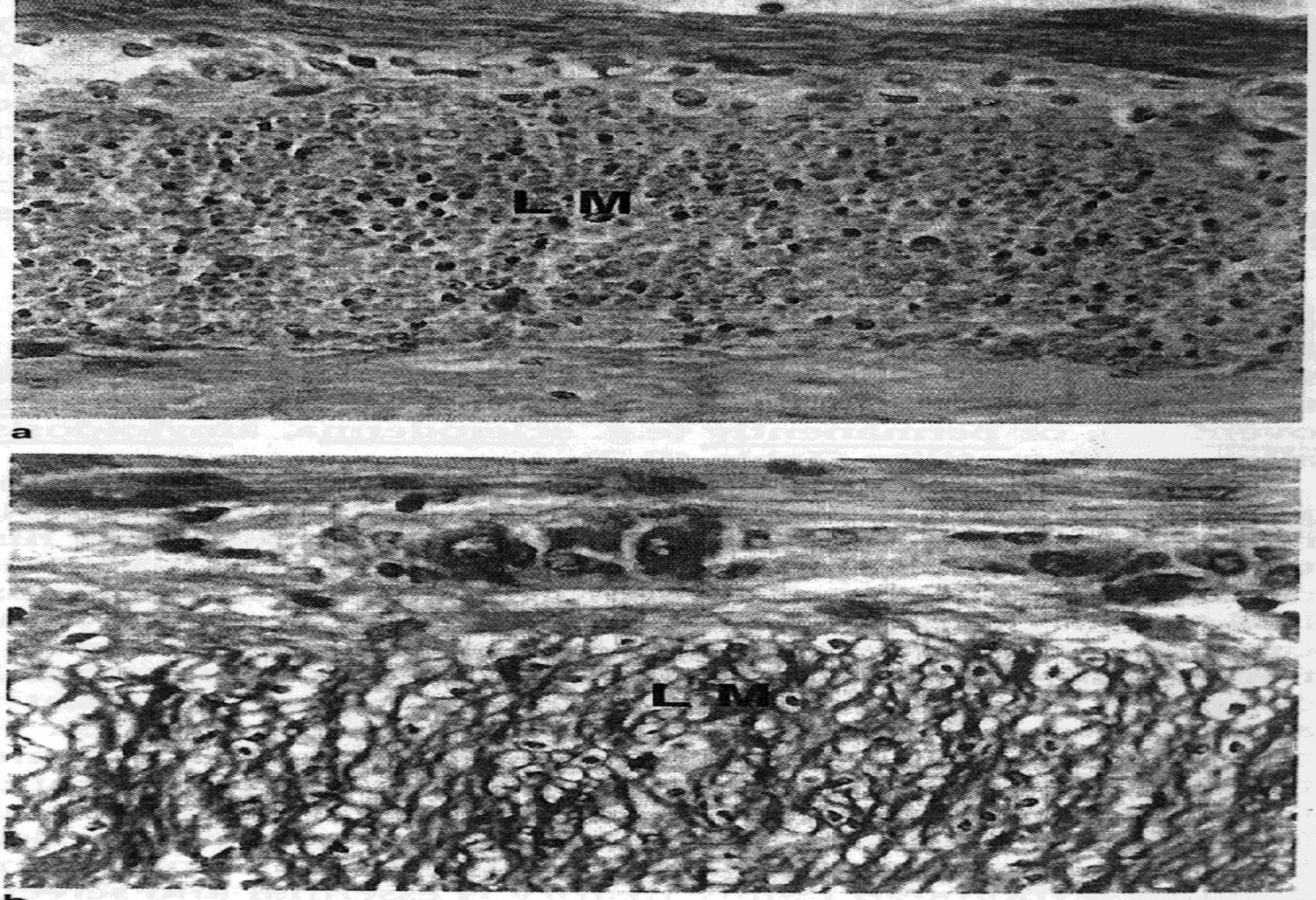
  carnitine

  ↓

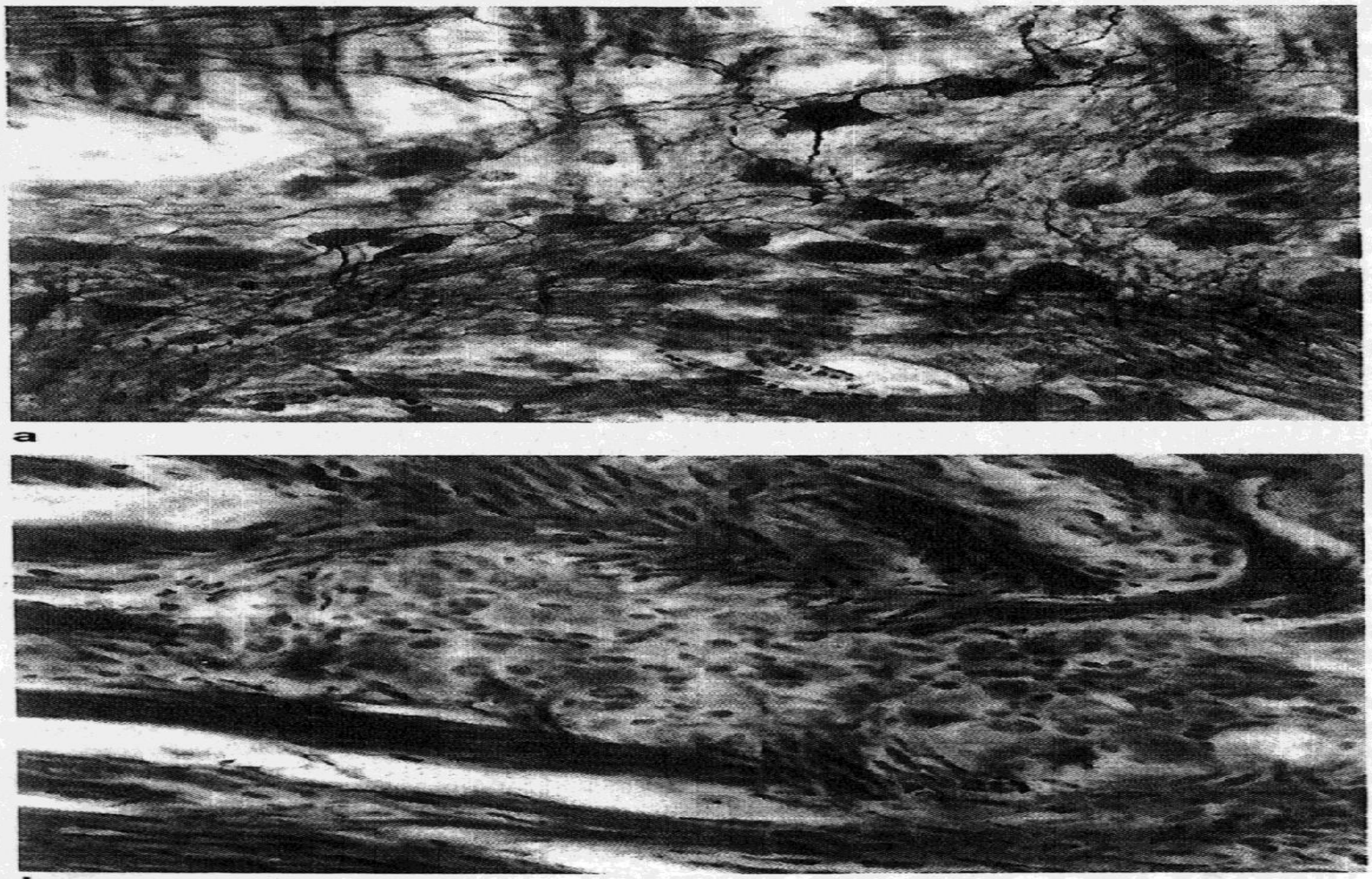
  vit. E (brown bowel syn)

- **Infections : CMV – EBV – HSV – rotavirus**

  Lyme -HIV



Visceral myopathy. (a) Longitudinal muscle cut in cross section from the small intestine of a control infant. (b) Longitudinal muscle from an infant with visceral myopathy shows classic vacuolar degeneration. Note the normal neurons in the myenteric plexus above the longitudinal muscle. ×136. (Courtesy of Michael D. Schuffler.)



Maturational arrest of myenteric plexus. (a) Ganglionic area of myenteric plexus from the small intestine of a control infant. Note the numerous argyrophilic neurons and axons. (b) Ganglionic area of myenteric plexus from the small intestine of an infant with chronic intestinal pseudo-obstruction caused by maturational arrest. Note the absence of argyrophilic neurons and axons. The ganglion is filled with numerous cells, which are probably glial cells and immature neurons. ×544. (Courtesy of Michael D. Schuffler.)

*Clinical manifestations

- Prenatal : In 20%of cases

  MCMC IHS

  The most common: megacyctis 88%

  hydronephrosis: 53%

  ↑AF 34%

  gastric dilation 10%

- Neonatal : severe abd. distention

  bilious vomiting

  air fliud levels in abd XR

  diarrhea secondary to bact. Overg

* should be differentiated from motility

  immaturity in premature infants

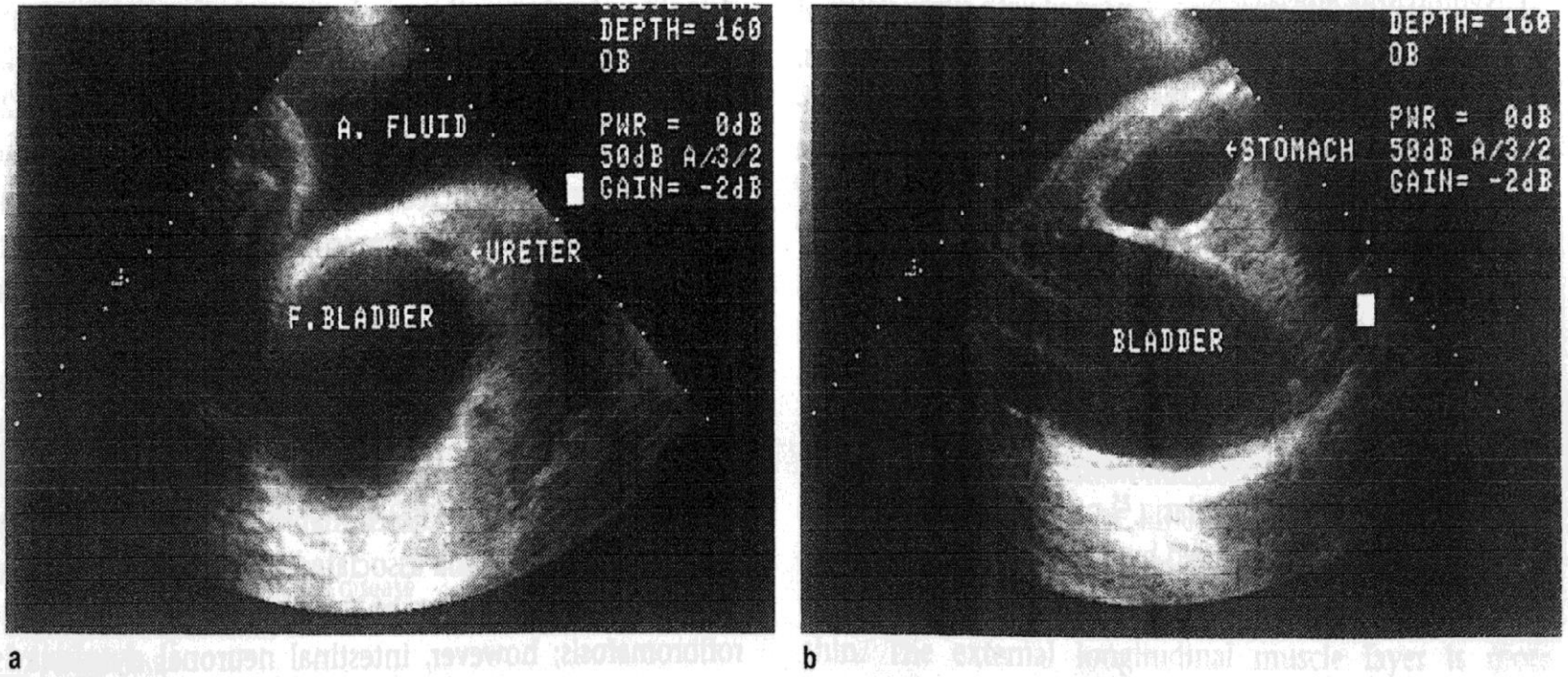


Figure 42.3: Ultrasound of infant with pseudo-obstruction diagnosed *in utero*. There is polyhydramnios as well as distention of the stomach and urinary bladder. (Courtesy of Radha Cherukuri.)

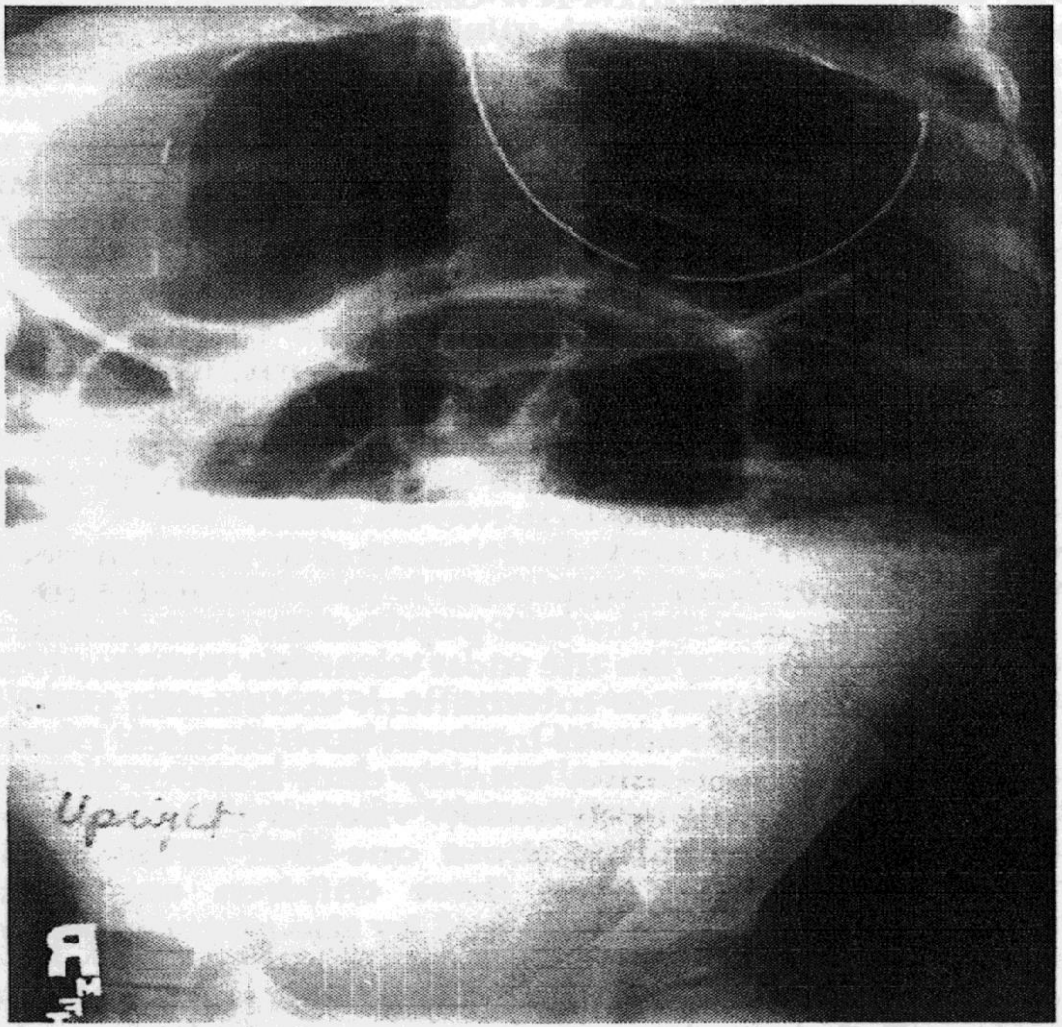


Figure 42.4: Upright abdominal radiograph in a 4-year-old boy with hollow visceral myopathy. Note bowel dilatation and air-fluid levels, catheter in the inferior vena cava and antroduodenal manometry catheter in the stomach and duodenum.

*Infantile or late onset

- **Major (acute) : episodes of GI obstruction**
  water and electrolyte losses
  Exacerbation with intercurrent infections –vaccination – fever – general anesthesia and stress
  Severe pain → malnutrition
  Megacyctis and cystic adynamia uretrohydronephrosis in 56-68%

- **Subacute : constipation 70%**
  abd. Distention
  bilious vomiting
  FTT

*Diagnosis

1. Recurrent vomiting after ladd's operation
2. Gut obstruction sign and symptoms without mechanical problem
3. Cystic dysmotility
4. R/O of hirschsprung and hypothyroidism in a full term neonate with GI obstruction

*3 steps in diagnosis

1. $^{R}/_{O}$ of mechanical obstructions
2. Documentation of GI motility disorder
3. Finding a systemic treatable cause

*X Ray

- Air fluid level in abd.XR
- Microcolon in neonates
- Megacystis
- Contrast study for $^{R}/_{O}$ of extra or intra luminal lesions with dilute nontoxic water soluble contrast
- Bowel distention is the most common finding 60%
- Bowel diverticticulosis
- Pneumatosis cystoides intestinalis
- Int. malrotation

*DD

Aerophagia

Gastroparesis

Functional constipation

Cyclic vomiting syn.

Severe IBS

Bacterial overgrowth

Aerodigestive fistula

Munchhausen syn.

*Manometery

- The most sensitive way for evaluation of gut muscle contractions

  In non dilated loop.

  In relax and non motile patient

- Visceral neuropathy → contractions with normal amplitude but disorganizing and not propagative
- Visceral myopathy → low amplitude contractions but organized

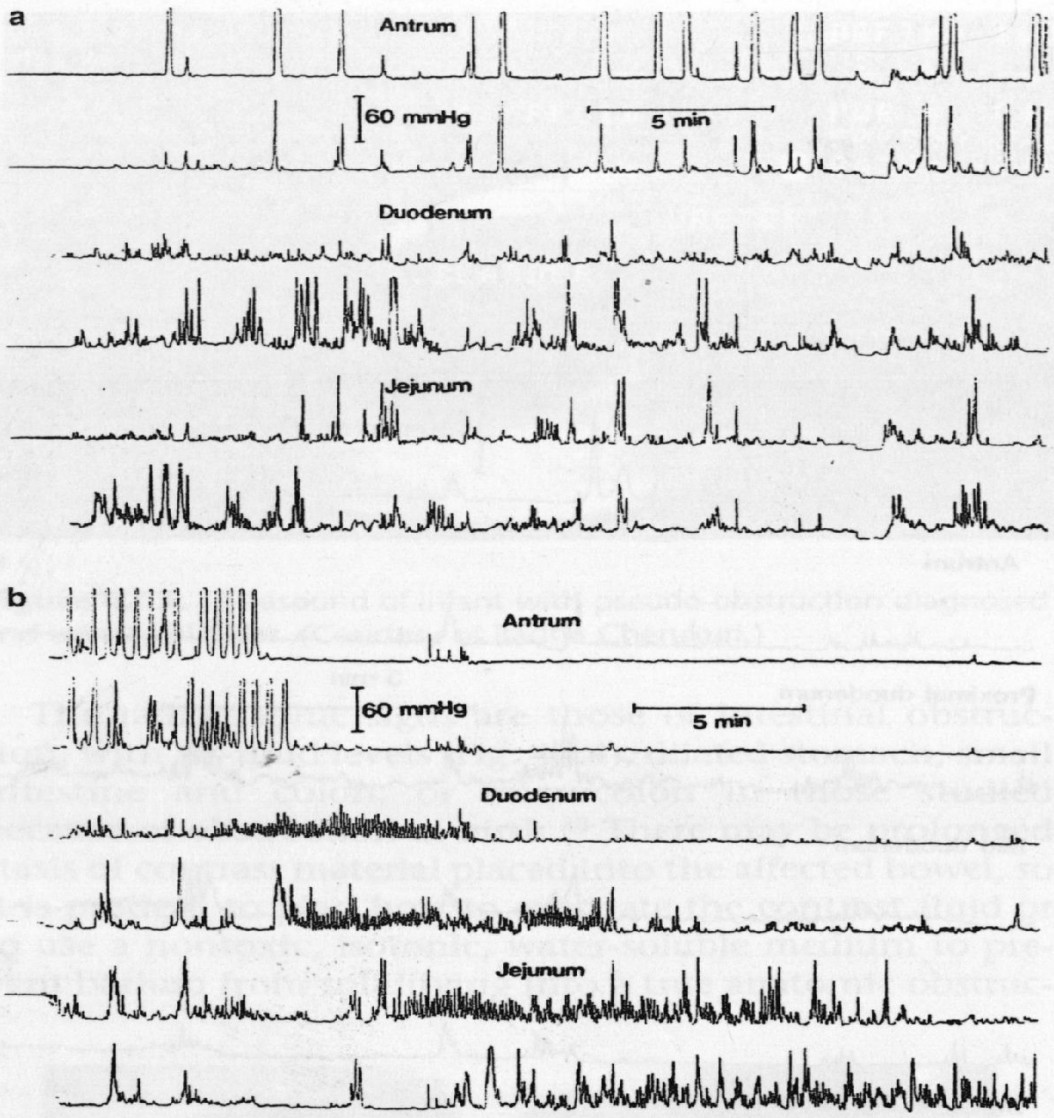
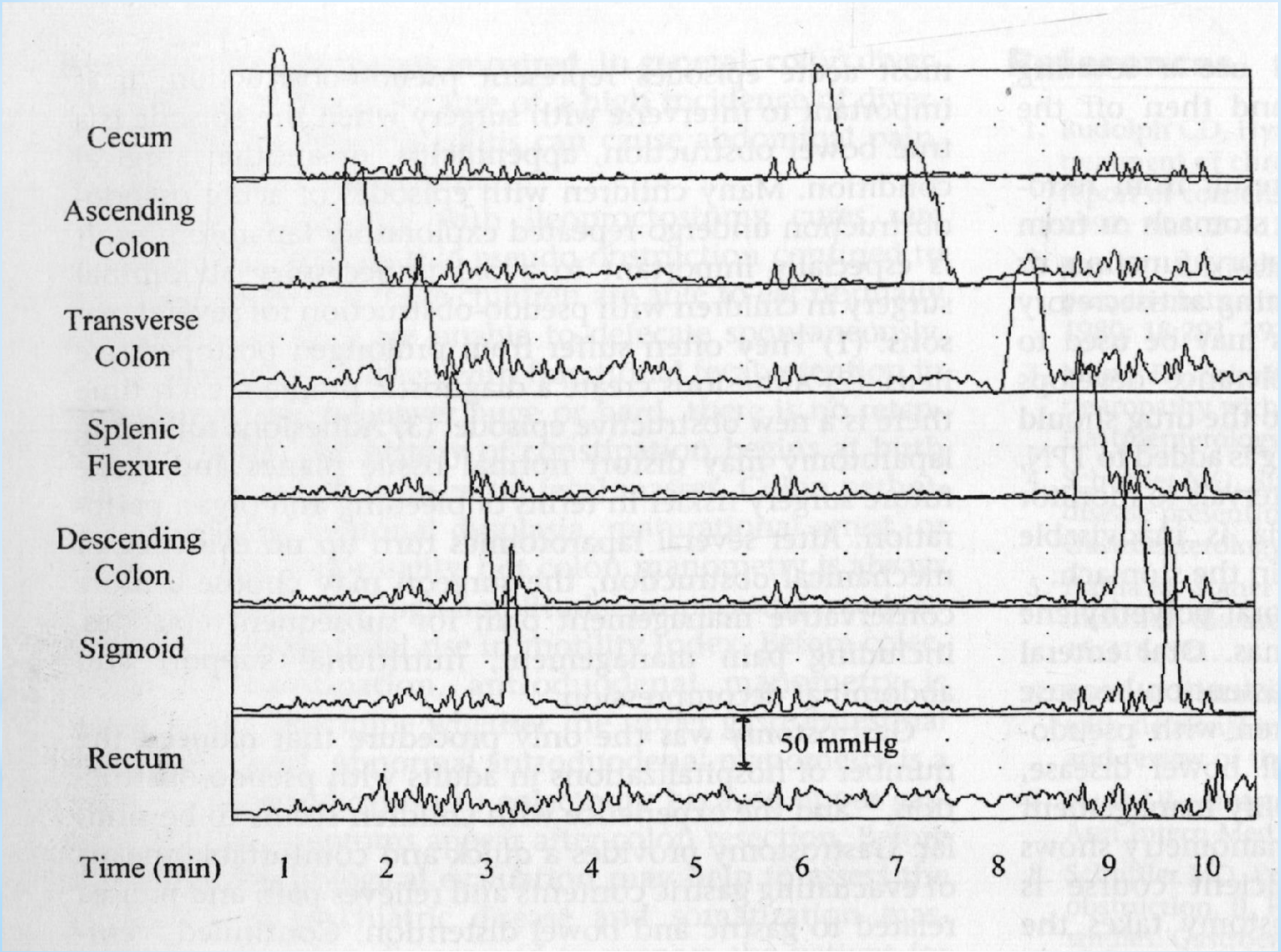


Figure 42.6: (a) Phase 2-like activity, consisting of random, intermittent, variable-amplitude contractions in the gastric antrum and the duodenum. This is also the postprandial pattern that interrupts the migrating motor complex (MMC). (b) Phase 3 of the MMC.


Cecum
Ascending Colon
Transverse Colon
Splenic Flexure
Descending Colon
Sigmoid
Rectum
50 mmHg
Time (min)
1
2
3
4
5
6
7
8
9
10

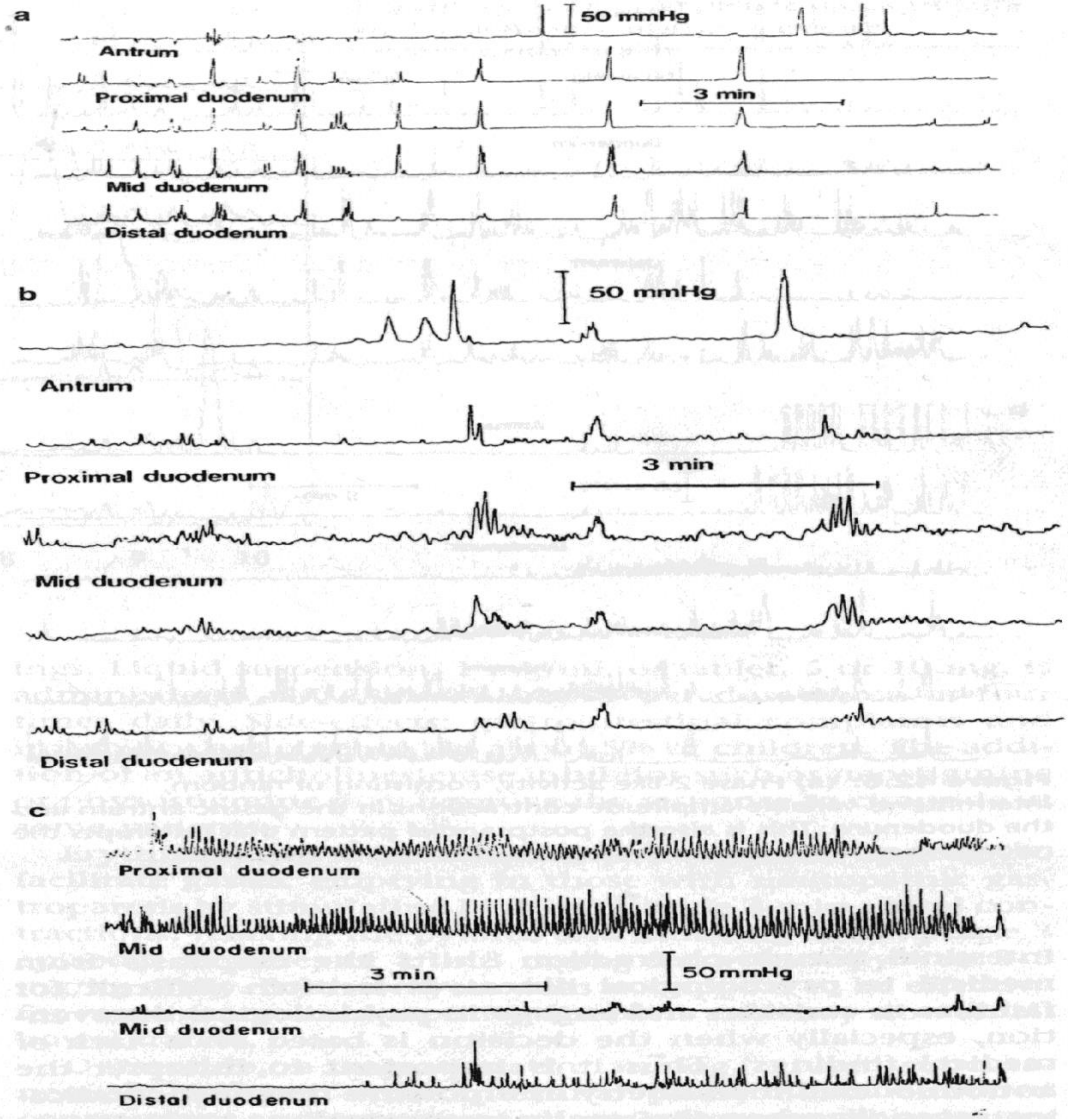


Figure 42.5: Discrete abnormalities in antroduodenal manometry. ...agating high-amplitude duodenal contractions. These ...single ring contractions are associated with irritable bowel disorders when the migrating motor complex is present. (b) Non-propagating, simultaneous, short clusters in monotonous pattern continuing uninterrupted for a 6-h study. (c) Long burst of non-propagating duodenal contractions.

* 50-90% abnormal esophageal manometery
* Inhibitory rectoanal reflex is present in CIP
* antroduodenal manometery is always abnormal and normal test rules out CIP
* Octreotide or erythromycin stimulation test is abnormal
* Gastric emptying scan is abnormal

- ***Pathology***: maturational arrest

  hypo or hyper ganglionosis

  ultrastructural study (mitochonria)

  immunohistochemistery for receptors and transmitters

- ***IND***: 1) hyperplasia of parasympathic and myenteric plexus neurons.

  2) ↑Acetyl cholinesterase + fibers

  3) heterotopic cell bodies

* ***PE***: light reflex

deafnens

motility of globe

(external ophtalmoplegia)

accomodation

Orthostatic hypotension

sweat abnormalities

vertigo

*PE

Skin rash

Palmar erythema

Telangiactasis

Nodules

Scleroderma

Reynoud

Peripheral Neuropathy

- ***Familial dysautonomia*** : 1) no axon flare after histamine

2) no papila in tongue

3) myosis after metacholin

4) no deep tendon reflexes

5) ↓ tears

GI cardinal sign is chronic vomiting

Lab // electrolytes
- TFT
- TTG
- Urine cat
- Plasma VIP
- ESR-C_3- C_4- ANA
- Plasma carinitine and vit. E
- Plasma lactate and pyrovate

Sono and CT of chest and abdomen for neural crest tumors
Brain MRI for leukoencephalopathy
Heart echo (in myopathies)
Rectal full thicknenss biopsy

* Treatment:

* Symptomatic management
* Dietary modification
* Prokinetics
* Tx of complications
* Avoidance of unnecessary surgery

Treatment

- In autoimmune dis : steroids
- NG suction or gastrostomy and gut decompression
- IV therapy
- AB
- Nutritional support (enteral is preferred) TPN infection – liver dis – thrombosis
- Prokinetics
- Opiates – loperamide
- Surgery

- Prokinetics : cisapride
  erythromycin
  octreotide
  betanechol
  tegaserod
  prucalopride
- Surgery : for gastrostomy or jejunostomy
  tapering of bowel
  or bowel, colon resection
  sym. Plexus neurolysis
  cecostomy
  small bowel TX

*Complications

- Ostomal prolapse
- Recurrent pancreatitis
- Diversion colitis
- ↑ileostomy out put
- Gastric perforation
- Gastric bezoar
- TPN comp.
- Secretory mucosa → severe dehydration

*Out come

- Bad prognosis : - neonatal onset
  - urinary disorders
  - need to surgery
  - myopathic CIP
  - absent MMC (phase III)
  - need to TPN> 6 mo
  - $MENII_b$ – thyroid med. Carcinoma