A Neonate with Failure to Pass Meconium

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Objectives

- Discuss the radiologic findings that suggest low intestinal obstruction in a neonate
- Generate a differential for low intestinal obstruction in a neonate and discuss how imaging can be used to narrow that differential
- Consider the complications of our patient's current presentation and how they may present on imaging
- Review some common later sequelae of our patient's condition and describe their radiologic appearance

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Meet Our Patient

- Baby J is a 1-day-old full-term baby boy who has had abdominal distension since birth.
- He has thus far failed to pass meconium and he has had one episode of bilious emesis.

- What are you worried about?
- What tests or studies are indicated?

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Causes of Obstruction

- **High intestinal obstruction (proximal to ileum)**
  - Presents predominantly with vomiting, usually pass meconium from patent distal bowel
  - Radiograph may show distension of the stomach, duodenum, and/or jejunum with few (if any) distended bowel loops*
    - *The only proximal obstruction with multiple bowel loops is midgut volvulus, in which the bowel dilates due to to ischemia or infarction. In this case, the infant would appear very ill.

- **Low intestinal obstruction (ileum and distal to ileum)**
  - Fail to pass meconium, vomiting not predominant presentation
  - Radiograph shows multiple dilated loops of bowel
  - Consistent with our patient's clinical and radiographic presentation!

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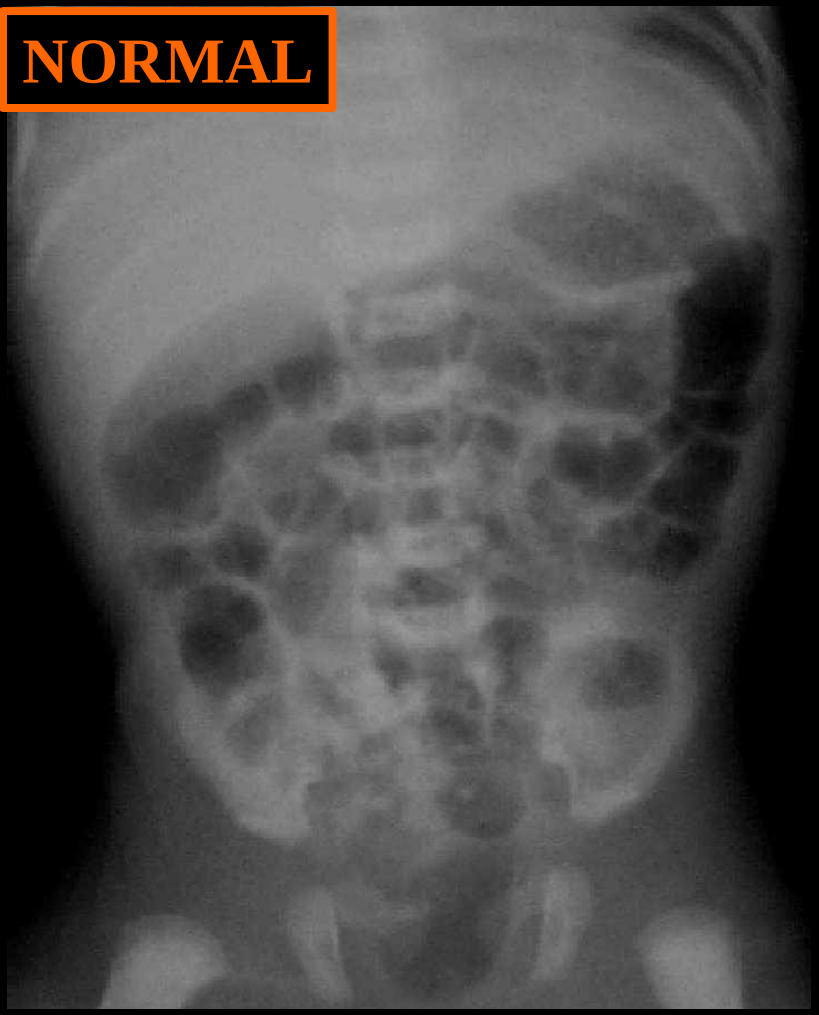
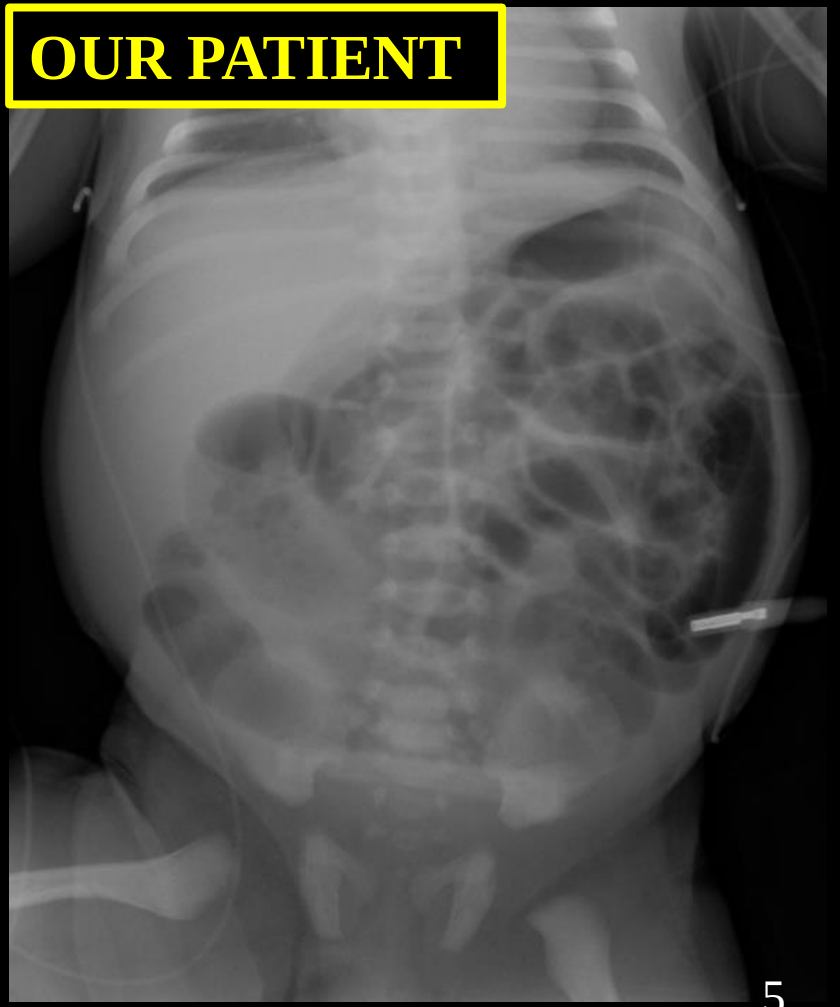
Our Patient: Abdominal Radiograph

NORMAL



http://www.medicine.cmu.ac.th/dept/radiology/pedrad/normal.htm

OUR PATIENT



Index patient radiograph courtesy of Dr. Walters, BCH

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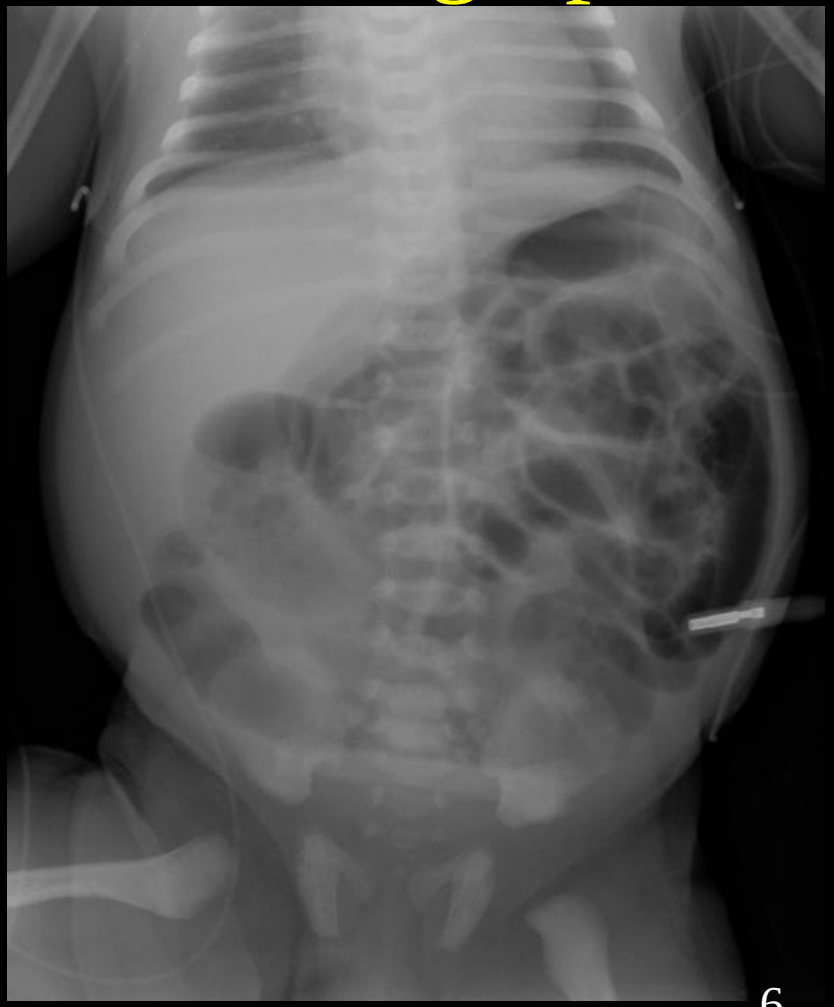
Our Patient: Review of Abdominal Radiograph

- Describe the findings



- What do these findings suggest?

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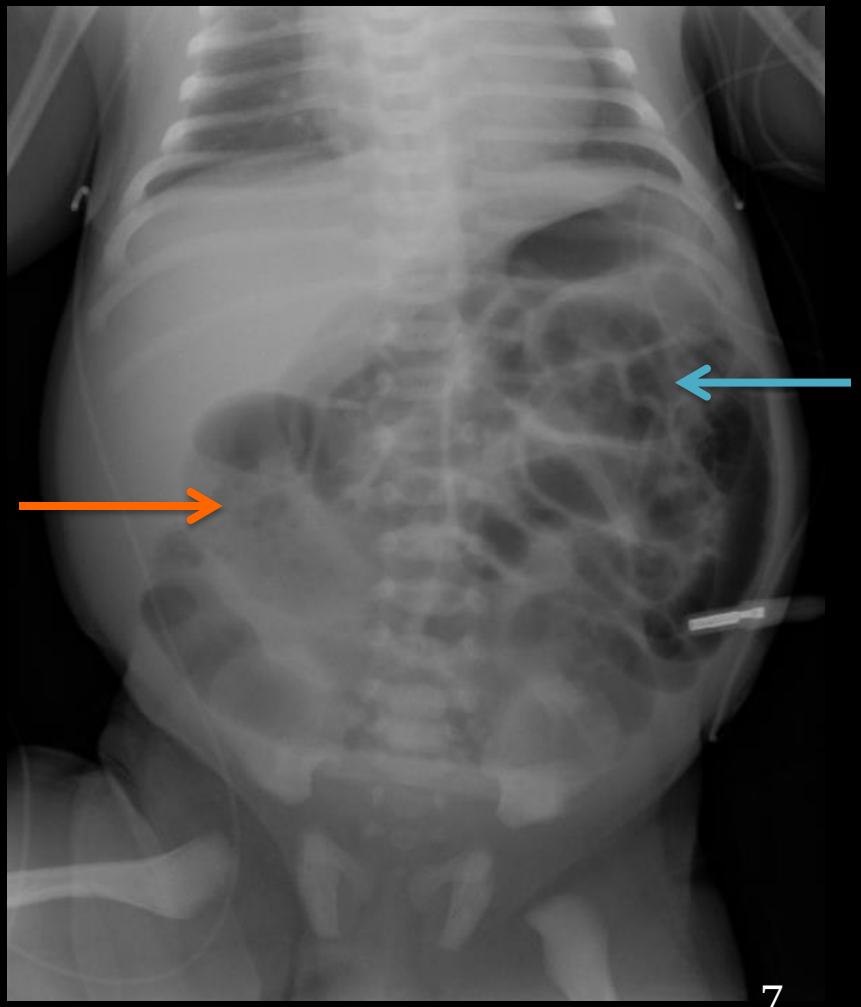
Our Patient: Analysis of Abdominal Radiograph

- Describe the findings
  - Diffuse distension of the bowel.
  - "Bubbly" or "frothy" appearance of bowel contents in RLQ
  - No air-fluid levels
  - No free air in peritoneum. No air in the rectum (confirm with prone cross-table lateral film).
  - In general, it is difficult to distinguish large and small bowel in infant radiographs because the mucosal folds may not yet have developed. Haustra in particular do not become prominent until ~6 mos.
- What do these findings suggest?



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Interim Summary of Patient's Course

- 1-day-old neonate with abdominal distension, failure to pass meconium, and one episode of bilious emesis, who was found on abdominal radiograph to have multiple dilated loops of bowel suggestive of low intestinal obstruction. Other than his abdominal distension, the infant appears well, and his physical exam does not show evidence of anal atresia or anorectal malformation.
  - What is your differential diagnosis?
  - What further workup is indicated?

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Differential Diagnosis

- What is your differential diagnosis?
  - Hirschsprung's disease
  - Ileal atresia
  - Meconium ileus
  - Small left colon (meconium plug) syndrome

- What further workup is indicated?
  - Contrast enema will help to narrow differential.

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Neonatal Contrast Enema

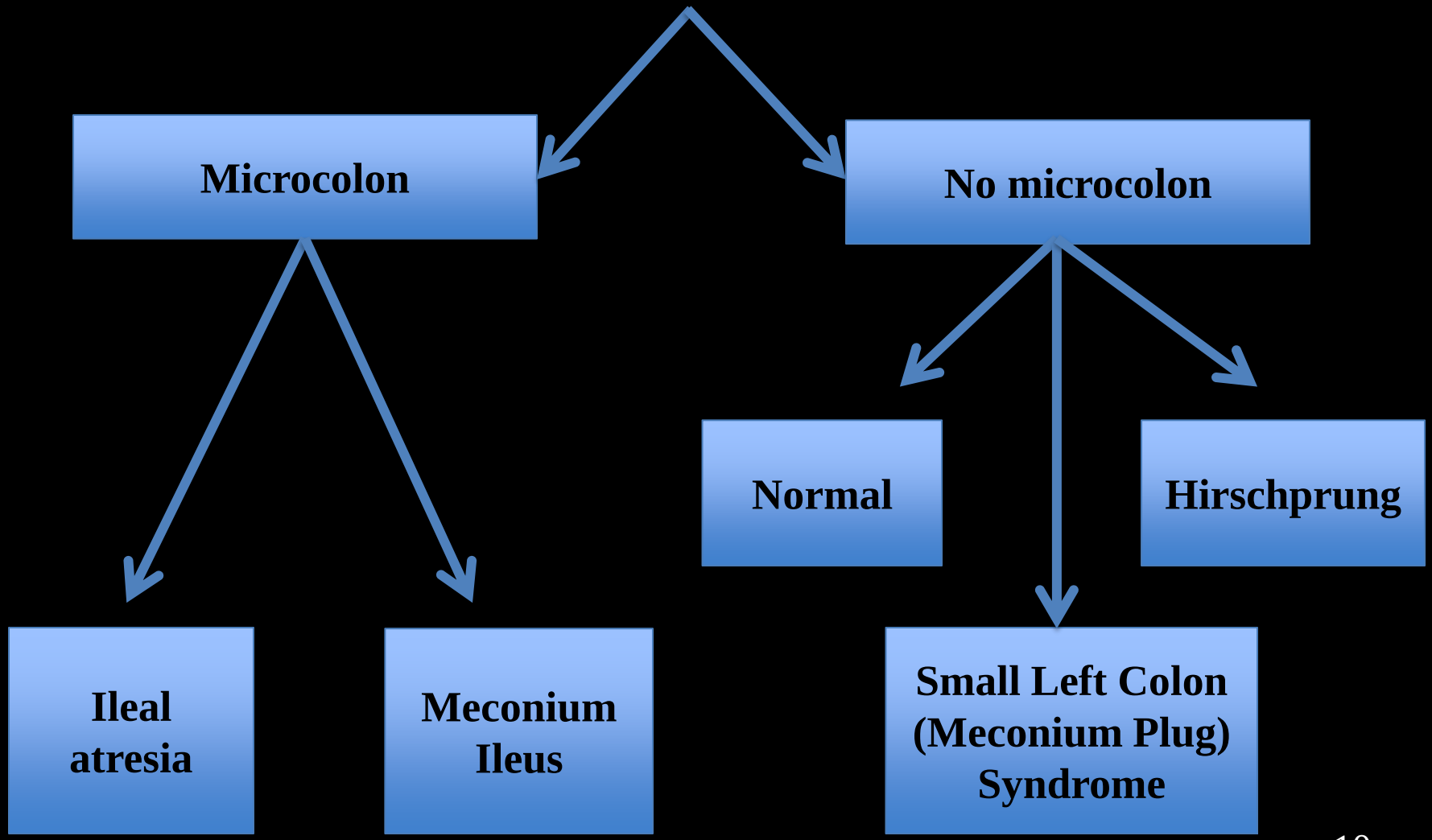

Chart adapted from: Donnelly, L. F. (2009). *Pediatric Imaging: The Fundamentals*. Saunders/Elsevier, p. 95

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Our Patient: Water Soluble Contrast Enema

- Pause to review the image and then continue for a discussion of the findings.

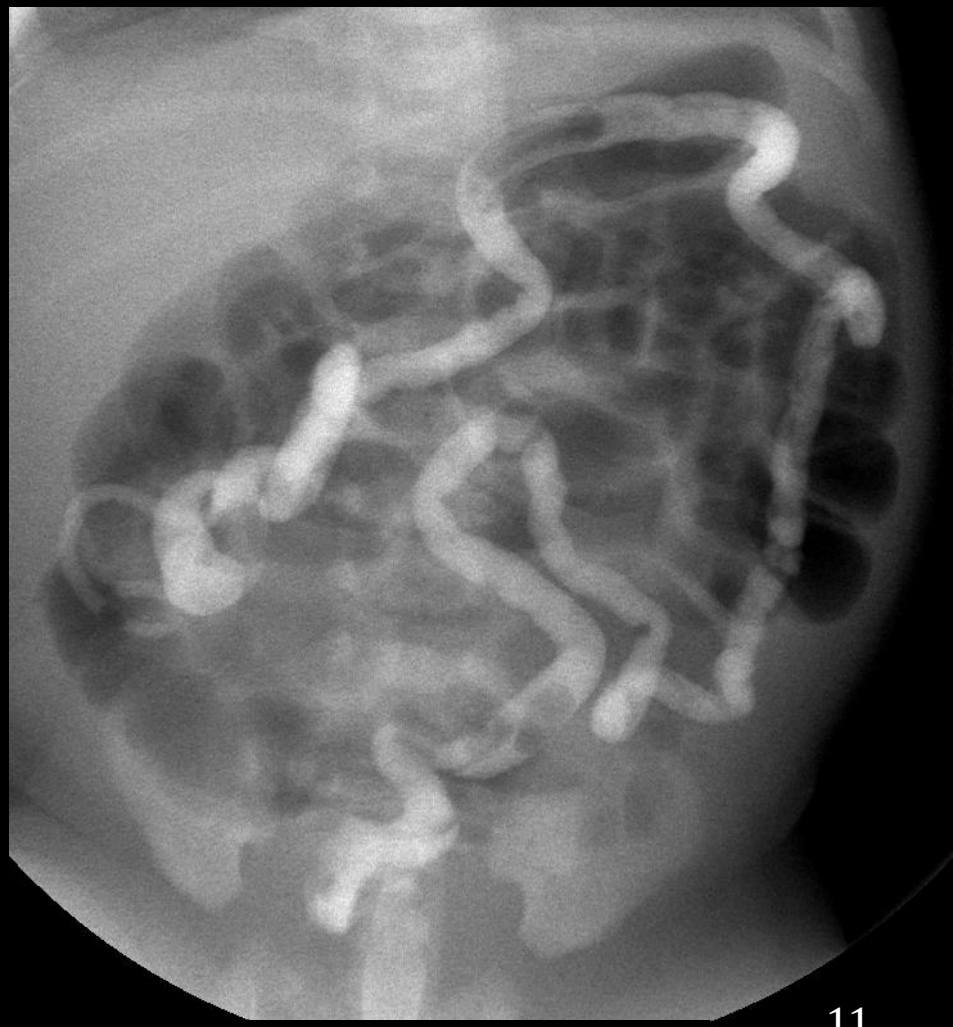

Image courtesy of Dr. Walters, BCH

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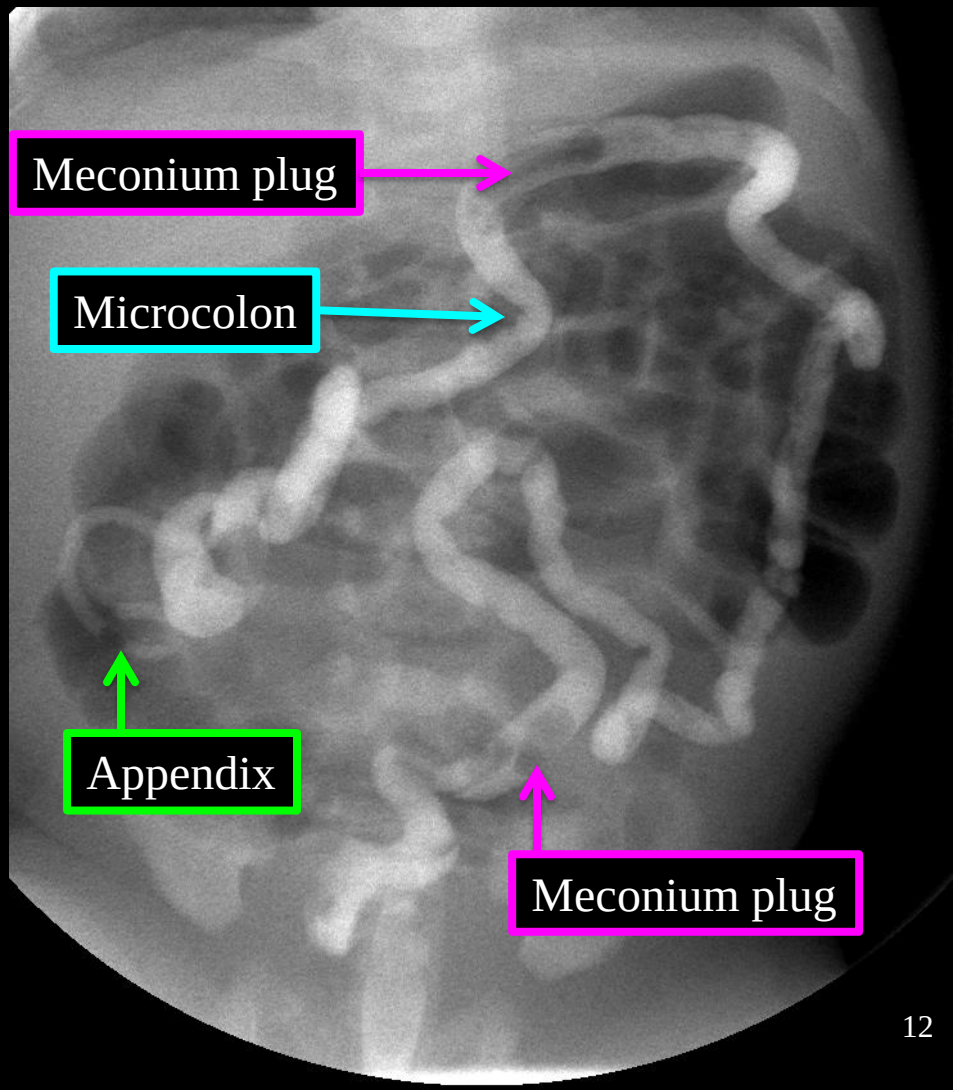
Our Patient: Analysis of Water Soluble Contrast Enema

- Note the narrow caliber of this colon (referred to as a microcolon). The colon has this appearance because it has not yet been used. Also note the that the terminal ileum is distended. There are also multiple filling defects likely representing meconium plugs.
- This image is highly suggestive of meconium ileus.



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Choice of Contrast for Enema

- Neonatal contrast enemas are typically performed using dilute, ionic, water-soluble agents.
- Gastrograffin is hypertonic, which causes increased small bowel secretions and thus facilitates passage of meconium; however, non-diluted gastrograffin may cause excessive fluid loss through the bowel wall, resulting in electrolyte disturbances, so diluted gastrograffin is the enema of choice.
- This enema is frequently therapeutic, successfully breaking up the meconium plugs and clearing the obstruction in 50-60% (Kao et al) of patients. If enema fails, surgical incision into site of obstruction and irrigation with acetylcysteine to dislodge meconium pellets is indicated.

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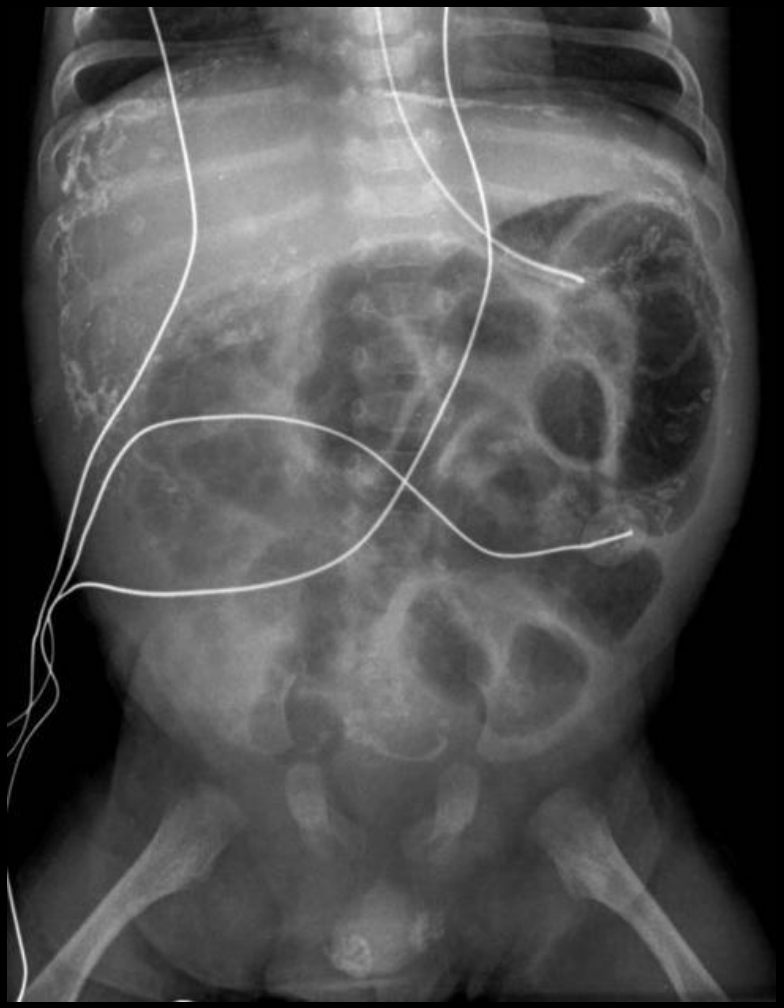
Complications of Meconium Ileus

- Meconium peritonitis
  - Refers to in utero bowel perforation and leakage of meconium into fetal peritoneal cavity
  - Notice the intraperitoneal calcifications
- Ileal stenosis
- Ileal atresia(s)
- Volvulus
- Pseudocyst formation
- Bowel perforation

- Any one of the above complications changes the classification of the meconium ileus from “simple” to “complex”.
- ~40% of meconium ileus is complex and may require surgery.



http://radiopaedia.org/articles/meconium-peritonitis

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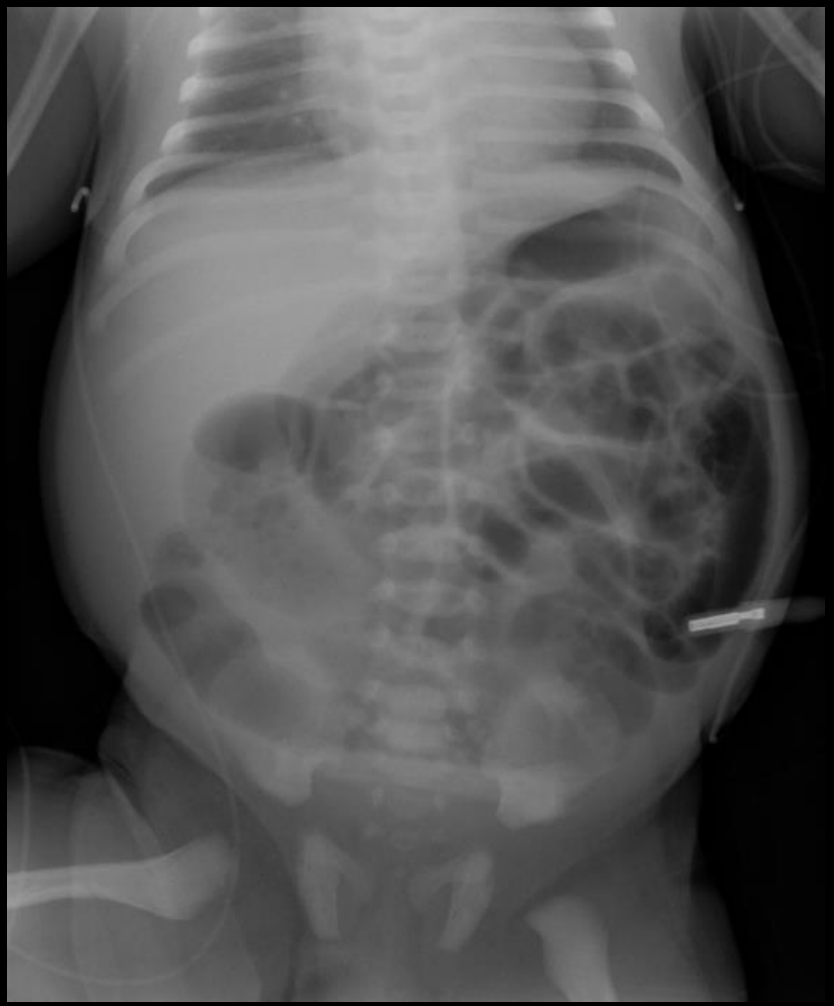
Meconium Ileus

- We now know that the bubbly appearance in the right lower quadrant was due to air mixed with meconium, creating a "frothy" or "soap-bubble" appearance. There are no air fluid levels because the meconium is thickened.
- Our patient's enema proved to be of therapeutic benefit, and surgery was not required. He then underwent further testing and was found to have CF.



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Review of Cystic Fibrosis

- Meconium ileus is highly associated with cystic fibrosis
  - 10-20% of patients with CF first present with meconium ileus.
  - ~ 90% of patients with meconium ileus are found to have CF.
- Newborns are screened for CF in all 50 states, but these results are usually not available until ~2 weeks after birth.
- CF is an autosomal recessive disease caused by mutations in CFTR, which is a chloride channel in all exocrine tissues. Disturbances in electrolyte transport result in thick, mucosal secretions that cause progressive dysfunction in multiple organ systems, most notably lungs, pancreas, intestine, liver, and reproductive tract. These mutations also result in abnormal electrolyte concentrations in sweat, which is useful for diagnosis.

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Sequelae of CF

- As our patient ages, he will likely develop many of the characteristic pathologies of CF. We will now review some common radiographic presentations of CF in the lungs, pancreas, and sinuses, by viewing some companion patients.

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Pulmonary Manifestations of CF

- The thick secretions of CF lead to inadequate clearance of pathogens, which results in recurrent respiratory infections and chronic inflammatory response, which in turn causes breakdown of the airway walls, leading to floppy, collapsible airways that contribute to air trapping.
- The combination of collapsible airways and thick mucus secretions that plug airways results in an obstructive pattern of lung disease.
- Obstructive lung disease is characterized by increases in TLC and RV, as well as decreases in FEV1, FVC, and the ratio of FEV1/FVC.

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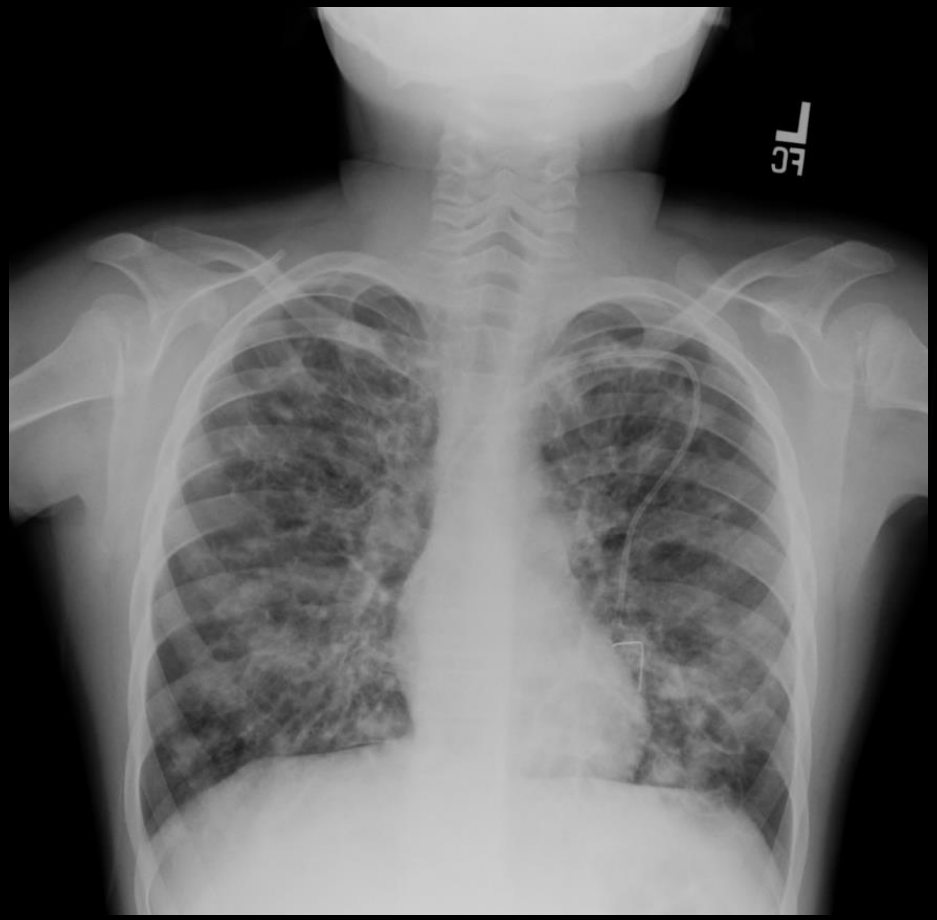
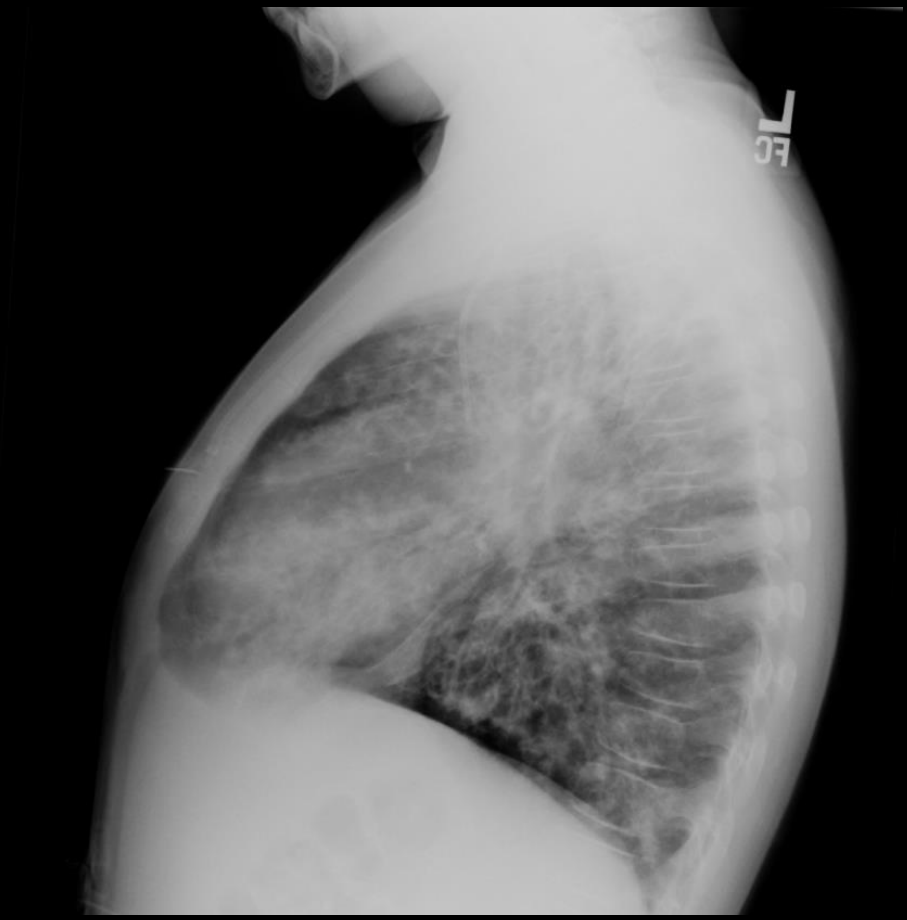
Companion Pt 1: AP and Lateral CXR of a 10 yo M with CF





Images courtesy of Dr. Walters, BCH

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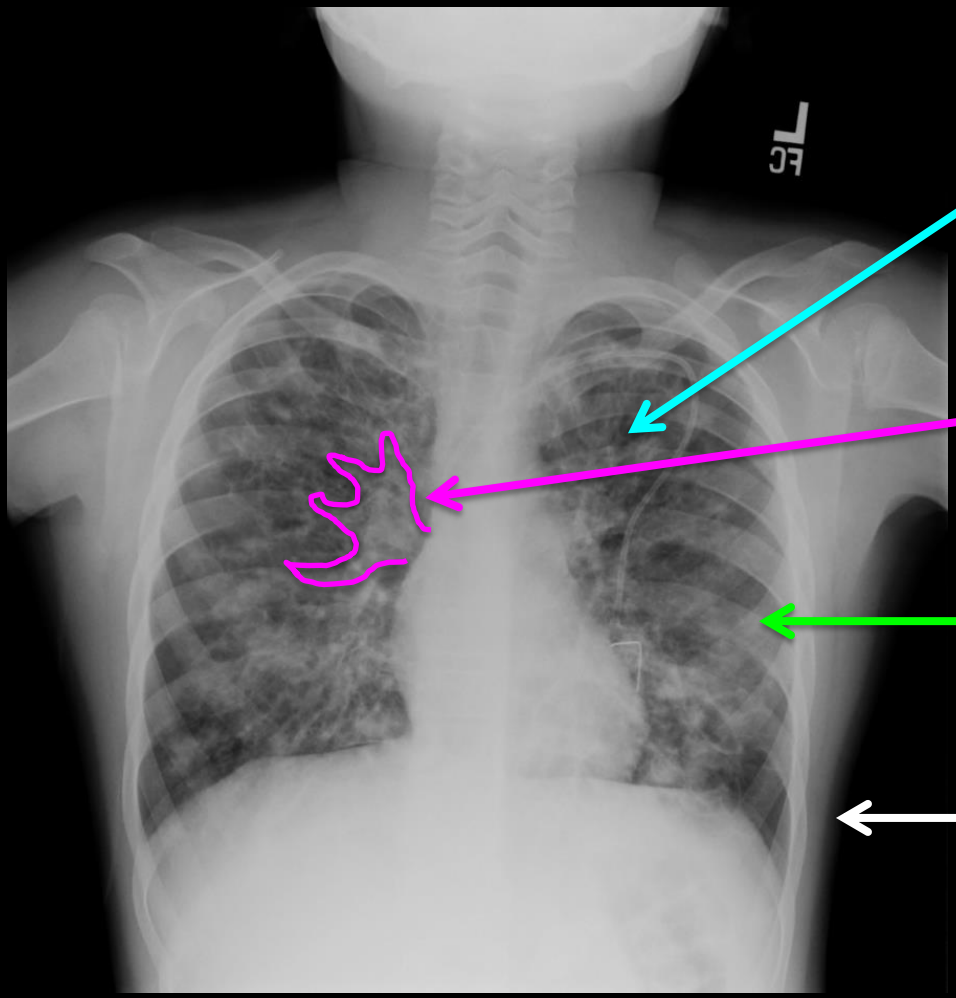
Companion Pt 1: Analysis of AP CXR



Increased bronchovascular markings

Diffuse, severe bronchiectasis with dilated, thick-walled bronchi in an upper lobe predominant pattern.

Nodular opacities scattered throughout lung fields represent mucus plugging

Hemidiaphragms are flattened, which is evidence of hyperinflation with air trapping

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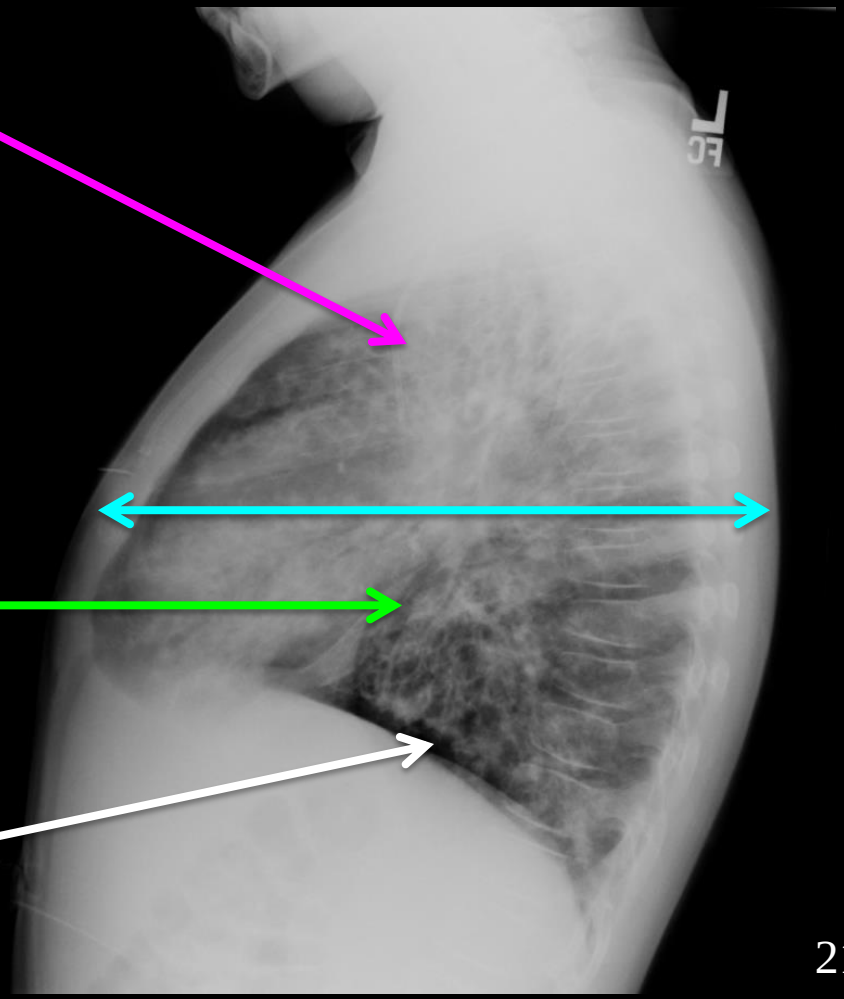
Companion Pt 1: Analysis of Lateral CXR

Diffuse bronchiectasis with upper lobe predominance

Increased AP diameter due to hyperinflation (blue arrow)

Nodular opacities representing mucous plugs

Increased lucency at bases represents air trapping



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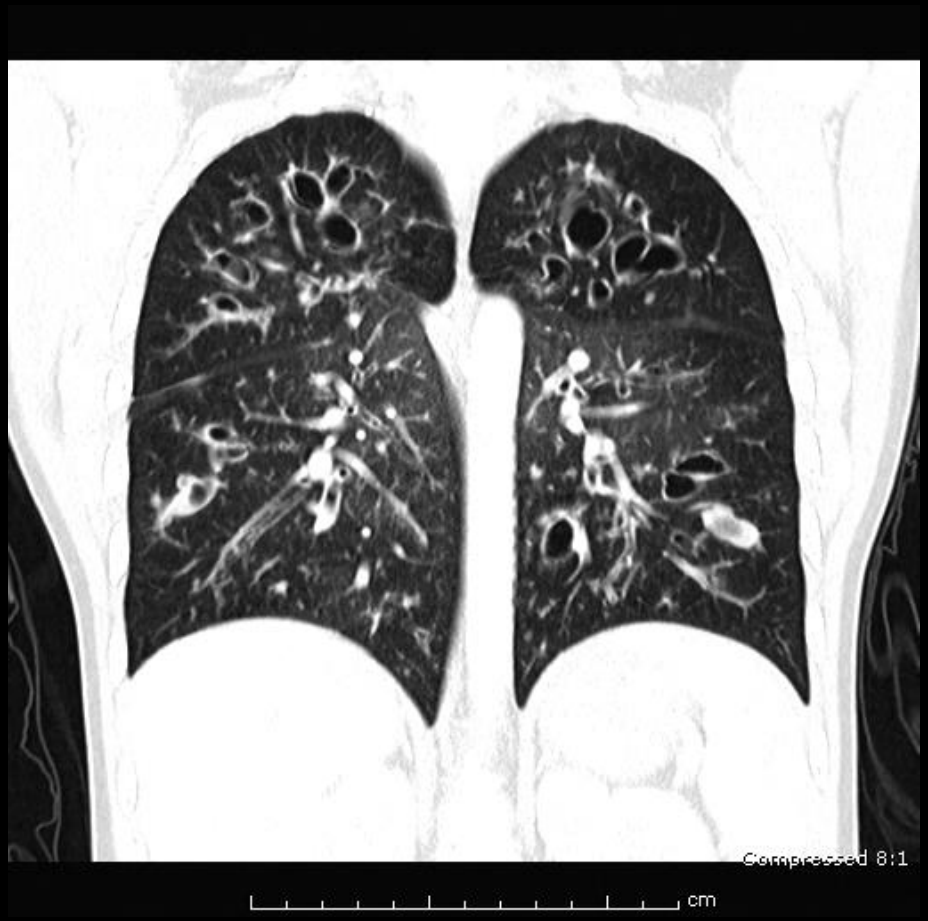
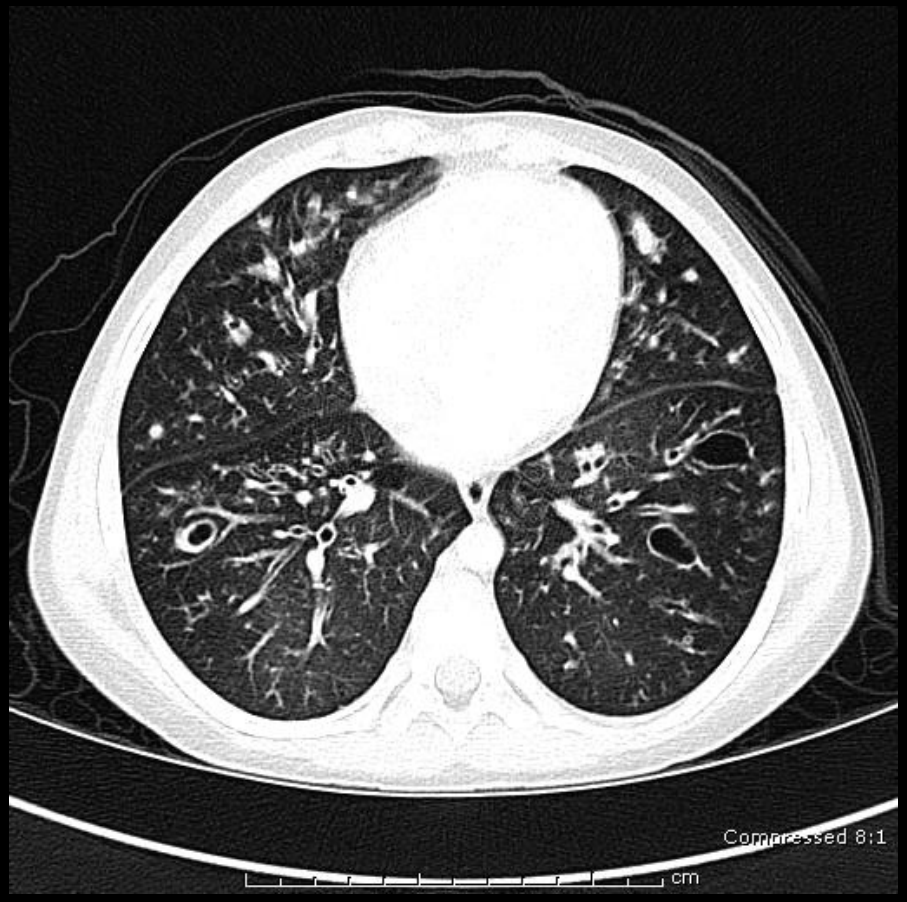
Companion Pt 1: Coronal and Transverse Chest CT





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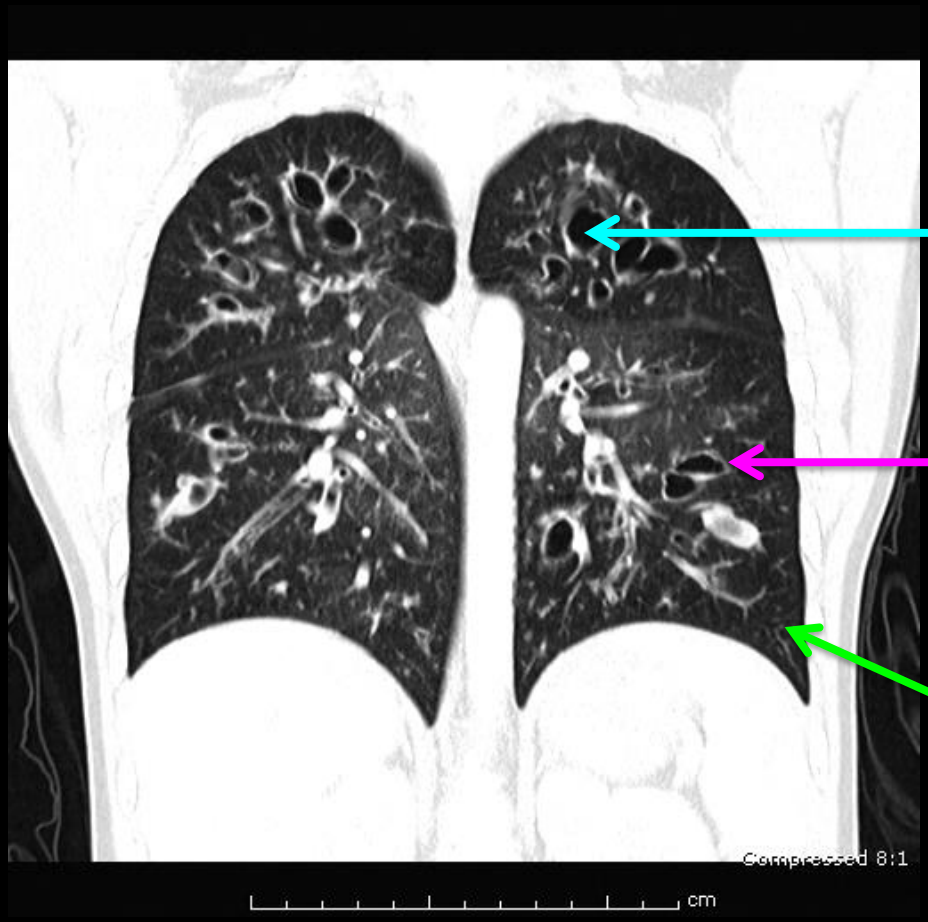
Companion Pt 1: Analysis of Coronal Chest CT



Severe bronchiectasis with upper lobe predominance

Peribronchial cuffing

"Tree in bud sign" suggesting bronchiectasis with mucus plugging

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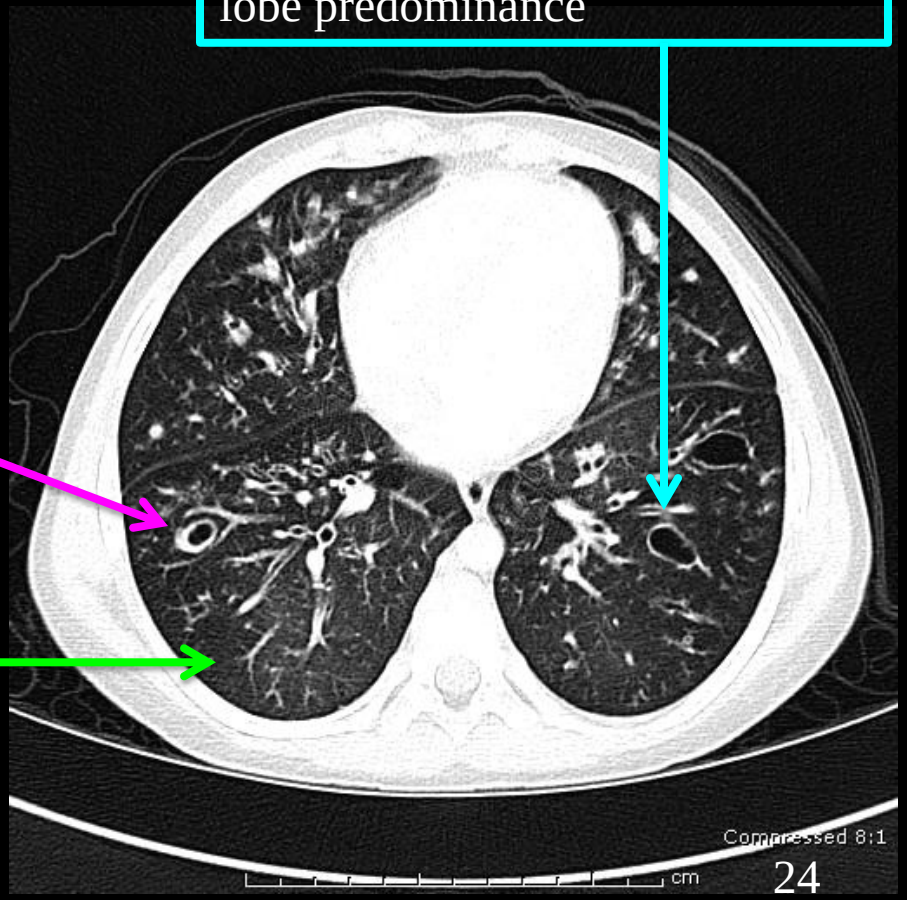
Companion Pt 1: Analysis of Transverse Chest CT

Severe bronchiectasis with upper lobe predominance

Peribronchial thickening

"Tree in bud sign" suggesting bronchiectasis with mucus plugging



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Pancreatic Manifestations of CF

- CF is associated with both pancreatic insufficiency and pancreatitis:
  - Due to the malfunction of the CFTR chloride channel, some CF patients are unable to secrete pancreatic enzymes, which need to be replaced in order to prevent malabsorption of fat, protein, and fat-soluble vitamins.
  - In some patients, thickened pancreatic secretions block the pancreatic ducts, leading to acute or chronic pancreatitis, the latter of which may eventually lead to total fatty replacement of the pancreas, as shown on the following slide.

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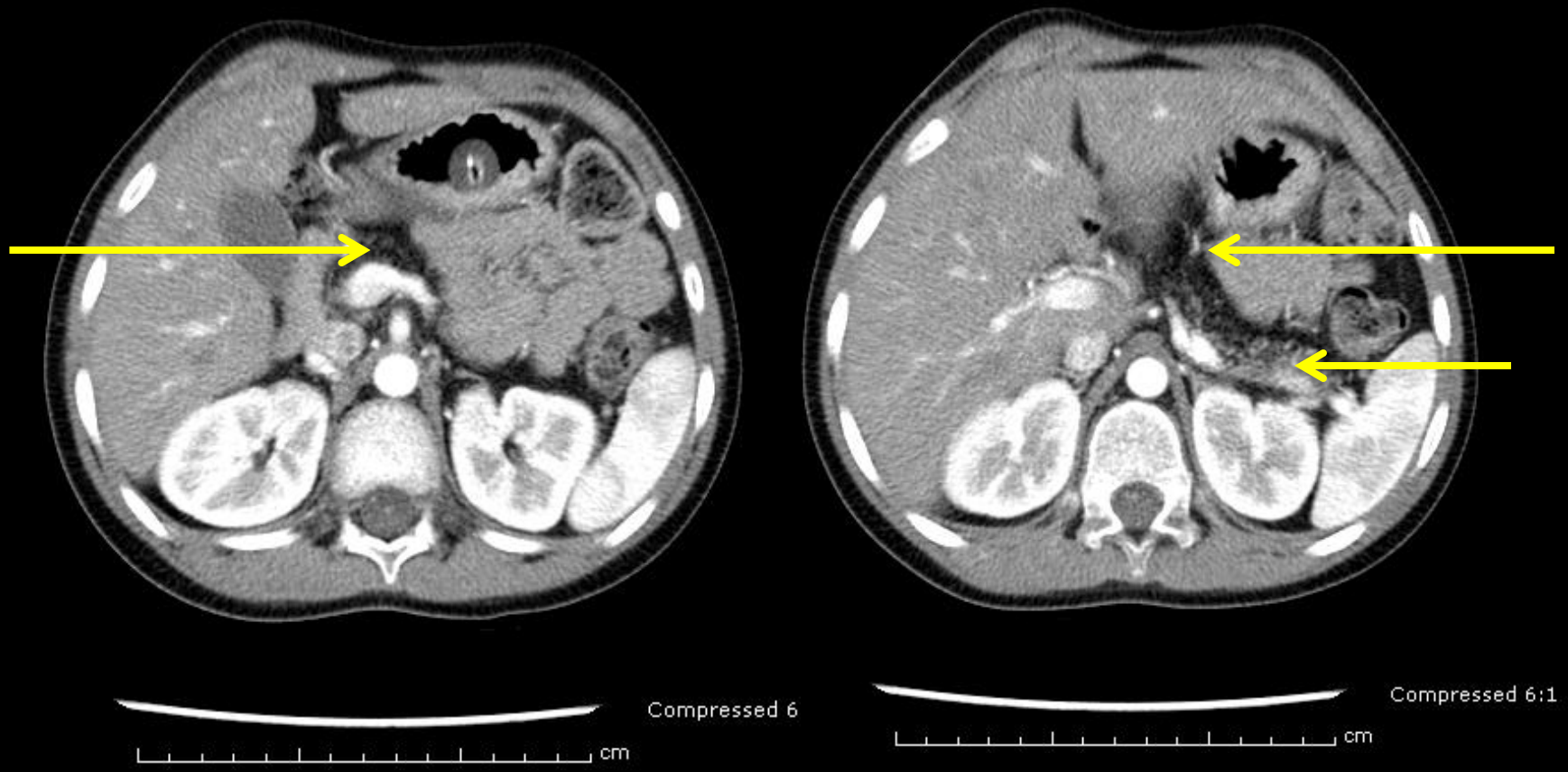
Companion Pt 2: Total Fatty Replacement of the Pancreas



Total fatty replacement of the pancreas is indicated by yellow arrows.
Images courtesy of Dr. Walters, BCH.

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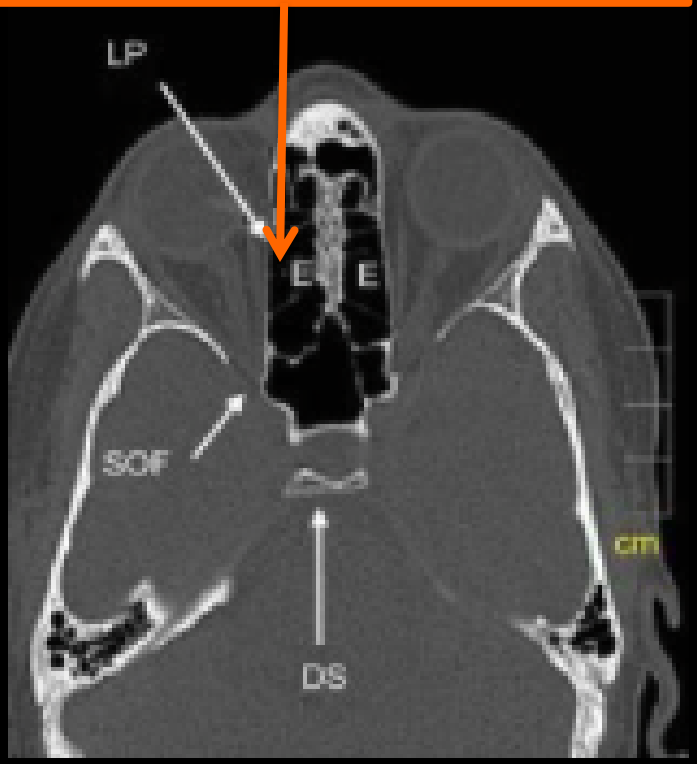
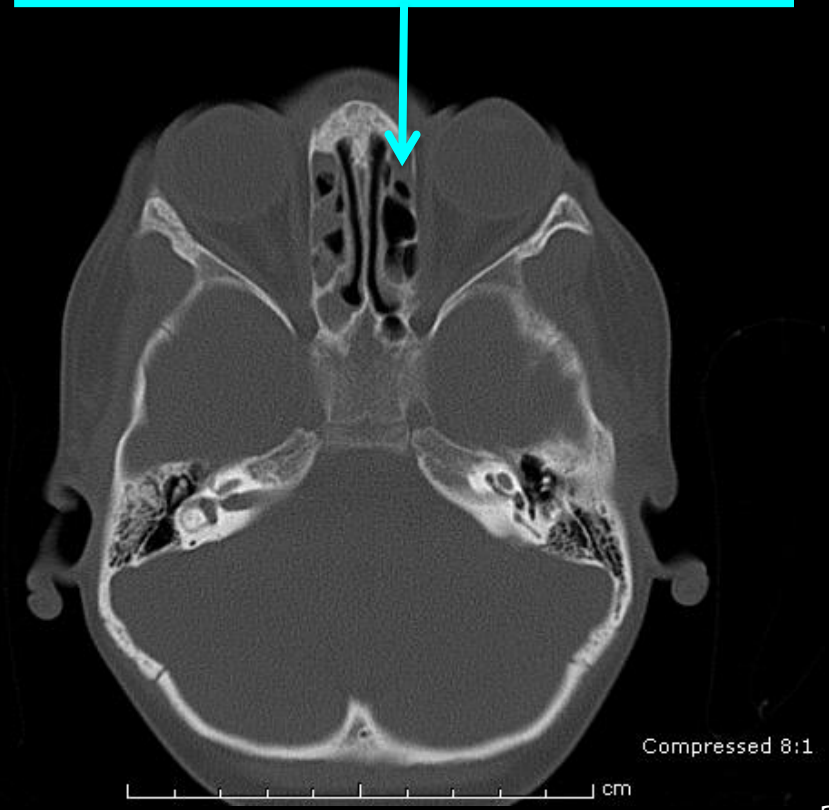
Companion Pt 3: Ethmoid Air Cell Opacification

NORMAL Ethmoid Air Cells



Opacified Ethmoid Air Cells



http://www.jaypeejournals.com/eJournals/ShowText.aspxID=3093&Type=FREE&TYP
=TOP&IN=_eJournals/imagesJPLOGO.gif&IID=238&isPDF=NO

Image courtesy of Dr. Walters, BCH

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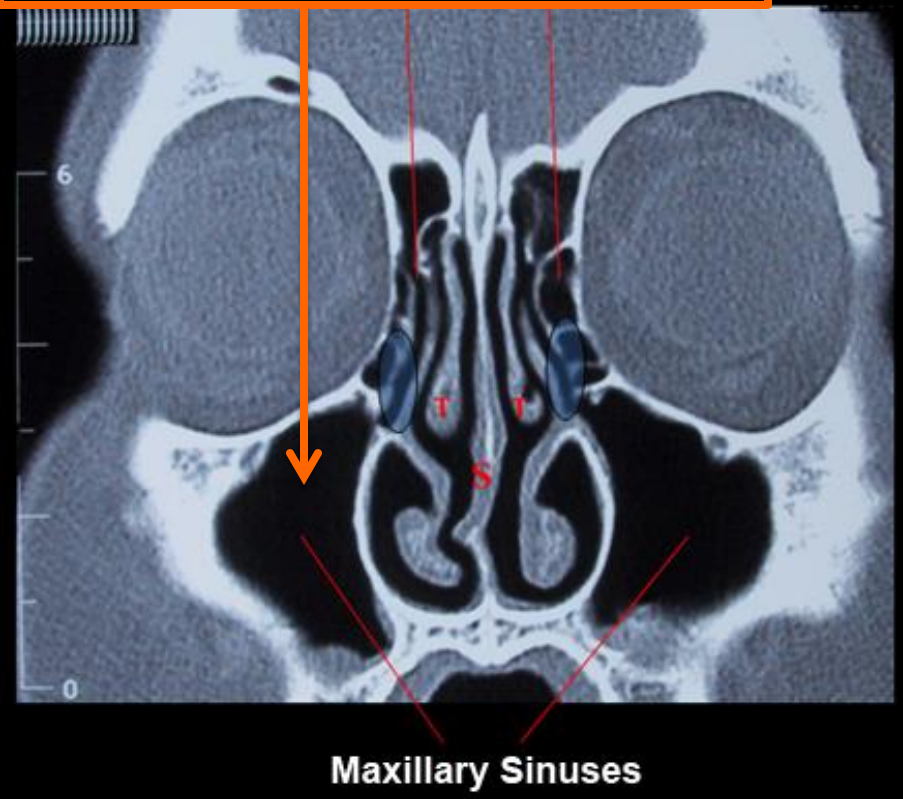
Companion Pt 3: Bilateral Maxillary Sinus Opacification

NORMAL Maxillary Sinus



http://care.american-rhinologic.org/sinus_anatomy

Opacified Maxillary Sinus

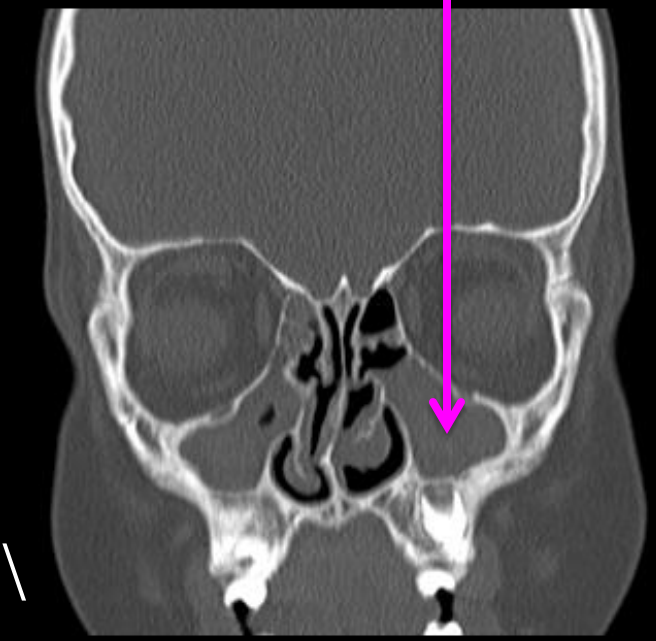


Image courtesy of Dr. Walters, BCH

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- Dr. Michele Walters

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