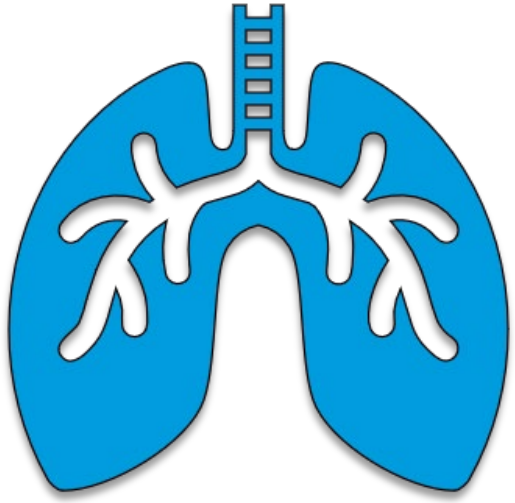
MAYO CLINIC



It’s Not Just the Lungs?

Non-Pulmonary Complications of Cystic Fibrosis

Ashley Otto, PharmD, BCPS

Pharmacy Grand Rounds
January 5, 2021

LEARNING OBJECTIVES

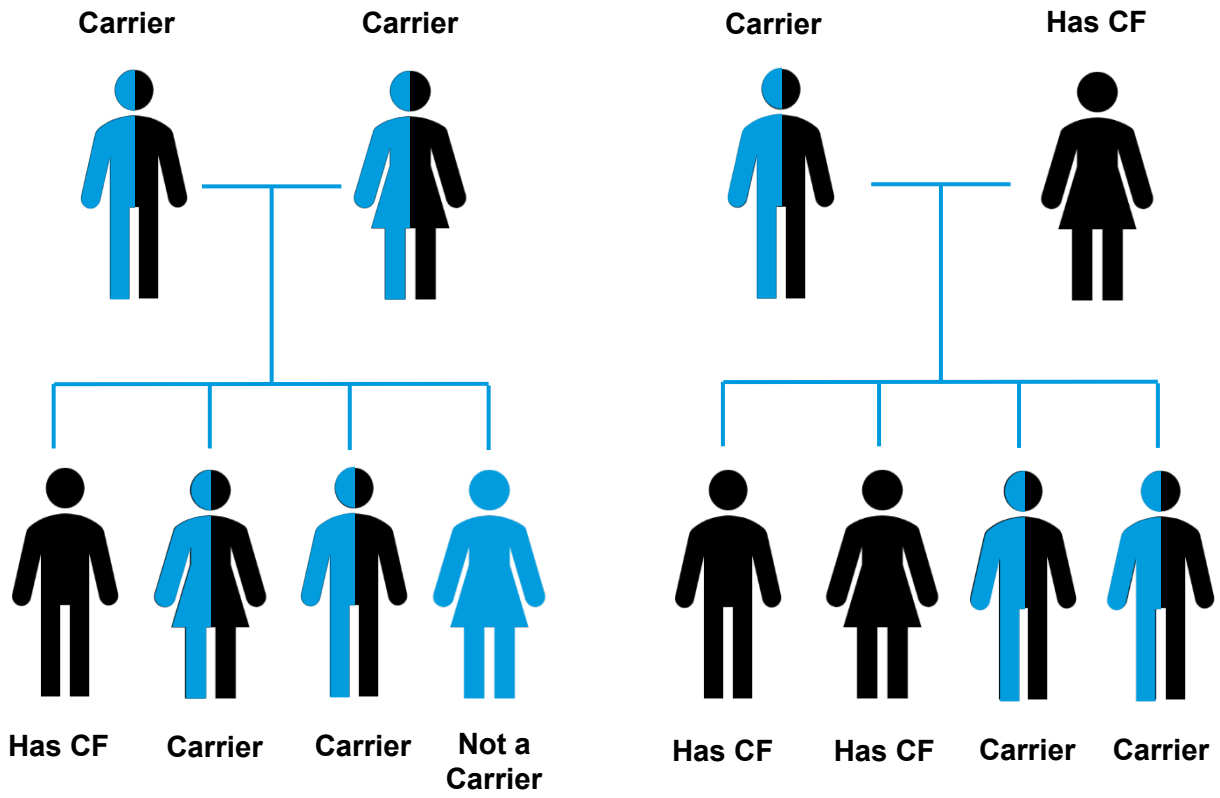
Describe the pathophysiology of no-pulmonary clinical manifestations of cystic fibrosis (CF)

Review guideline recommendations and primary literature to manage non-pulmonary CF complications

Select appropriate treatment options for patients with non-pulmonary CF complications

Cystic Fibrosis Epidemiology

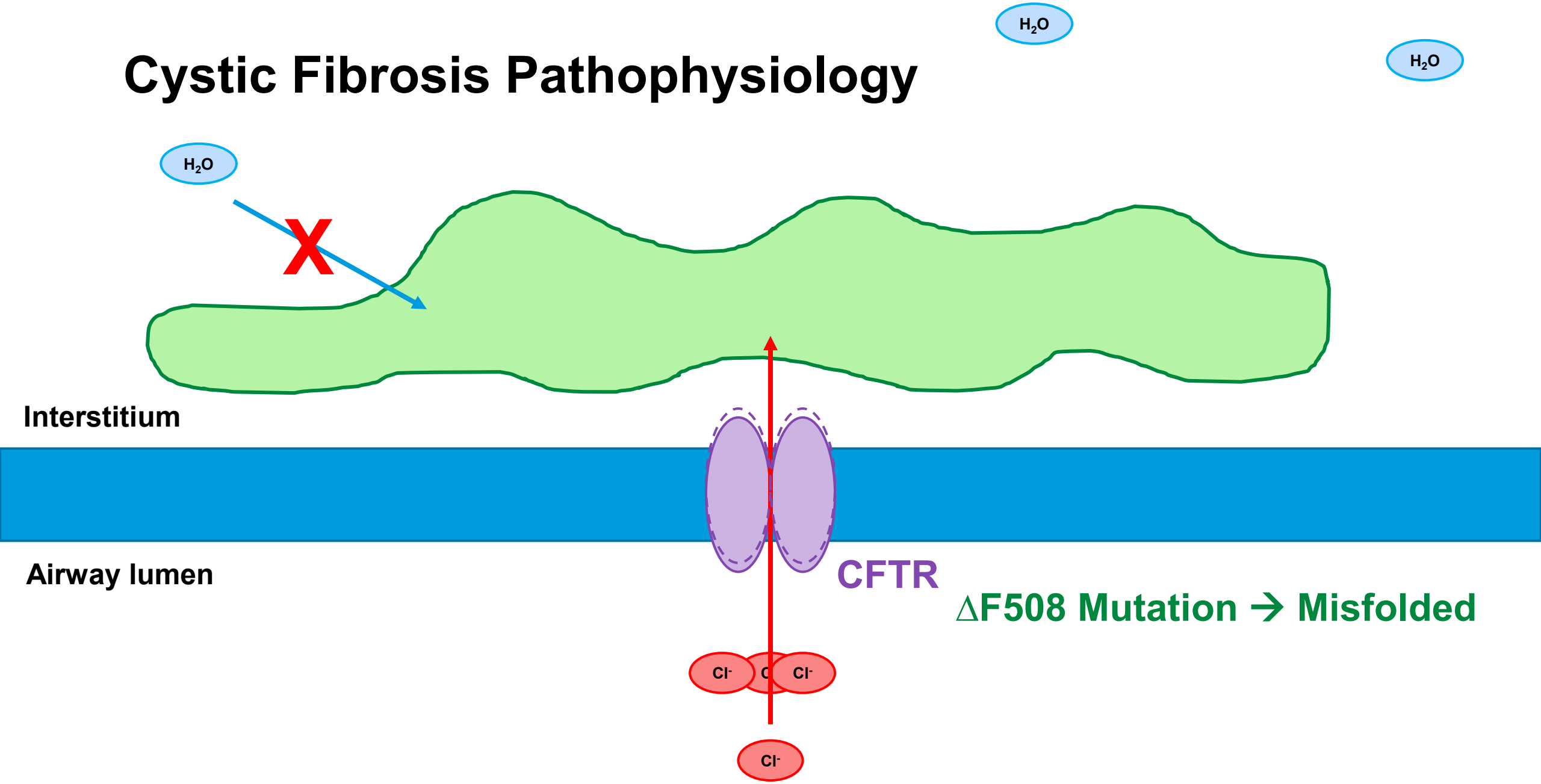
- Autosomal recessive genetic disorder
- Affects 70,000 people worldwide, of which > 30,000 are living in the United States alone
- Approximately 1,000 new cases diagnosed each year
- > 50 % of patients living with CF are > 18 years old



CF: cystic fibrosis

Cystic Fibrosis Foundation. About Cystic Fibrosis. Available at: https://www.cff.org/What-is-CF/About-Cystic-Fibrosis/. Accessed November 5, 2020.

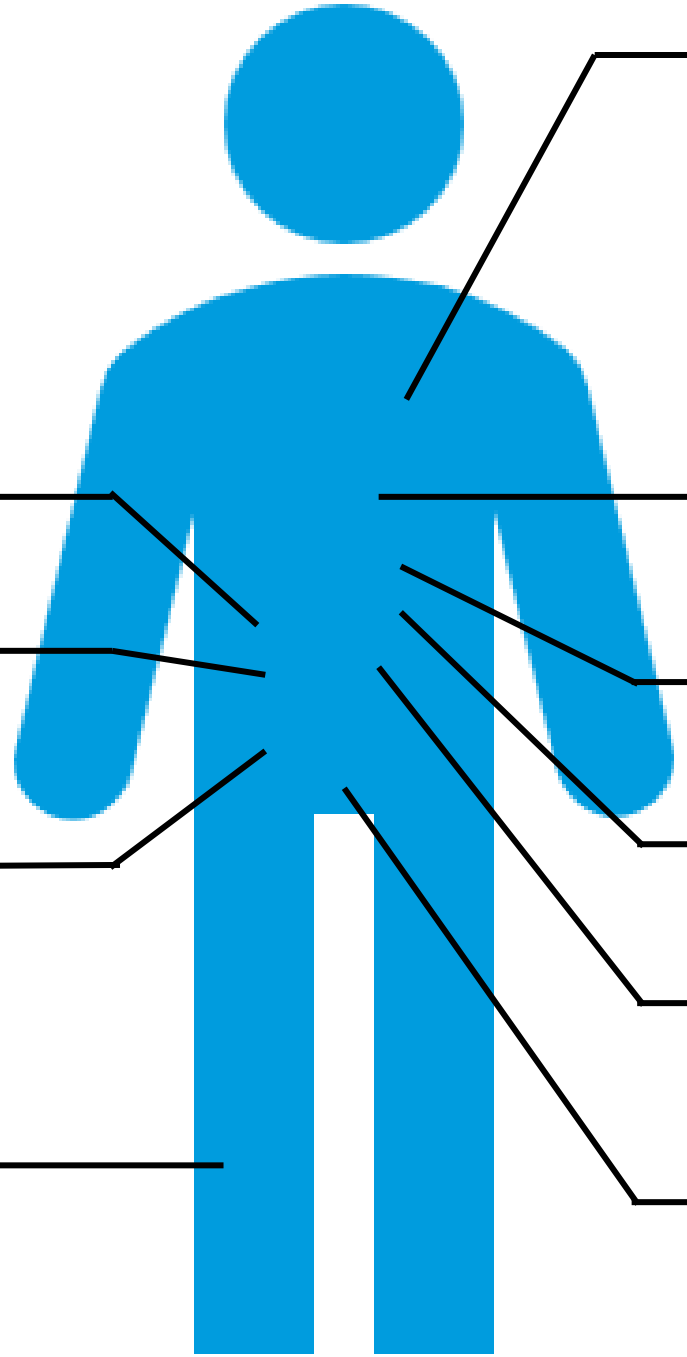
Cystic Fibrosis Pathophysiology



N Engl J Med 2005;352:1992-2001

CFTR: cystic fibrosis transmembrane conductance regulator

Complications of Cystic Fibrosis



General
- Growth failure (malabsorption)
- Vitamin deficiencies (A, D, E, K)

Liver
- Hepatic steatosis
- Portal hypertension

Gallbladder
- Biliary cirrhosis
- Neonatal obstructive jaundice
- Cholelithiasis

Intestines
- Meconium ileus, peritonitis
- Rectal prolapse
- Distal intestinal obstruction syndrome
- Intussusception
- Fibrosis colonopathy (strictures)

Bone
- Hypertrophic osteoarthropathy
- Arthritis
- Osteoporosis

Lungs
- Bronchiectasis, bronchitis
- Pneumonia, atelectasis
- Hemoptysis
- Pneumothorax
- Reactive airway disease
- Cor pulmonale
- Mucoid impaction

Heart
- Right ventricular hypertrophy
- Pulmonary artery dilation

Spleen
- Hypersplenism

Stomach
- Gastroesophageal reflux disorder

Pancreas
- Pancreatitis
- Insulin deficiency
- Diabetes, hyperglycemia

Reproductive
- Infertility
- Amenorrhea
- Delayed puberty

Adapted from: www.cancertherapy.com/hospital-medicine/cystic-fibrosis/article/602444

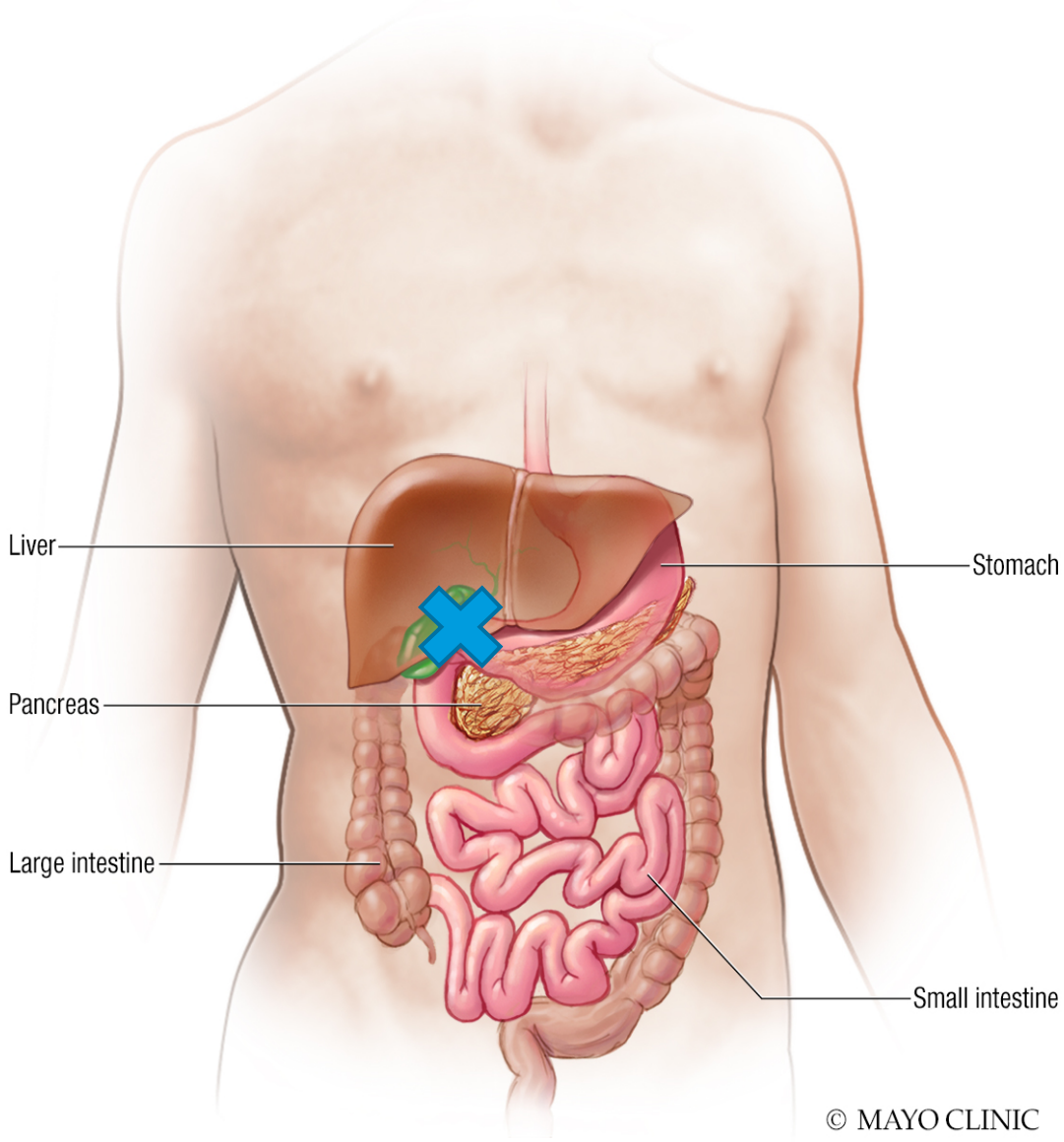
Cystic Fibrosis – Related Liver Disease (CFLD)

Cystic Fibrosis – Related Liver Disease (CFLD)

Loss of CFTR function in cholangiocytes

Biliary retention leads to obstruction and biliary sludge

Periportal fibrosis → portal hypertension → multi-lobular cirrhosis



CFTR: cystic fibrosis transmembrane conductance regulator

J Pediatr Gastroenterol Nutr 1999; 28 Suppl 1:S1-S13. | *J Cyst Fibros 2011;10(2):S29-36.* | *J Cyst Fibros* 2013;12(2):116-24.

Cystic Fibrosis – Related Liver Disease (CFLD)

Presentation

- Hepatomegaly +/- splenomegaly
- Jaundice
- Abdominal distention and ascites

Manifestations

Hepatic Complication	Approximate Frequency
Neonatal cholestasis	Infrequent
Hepatic steatosis	20 – 60%
Focal biliary cirrhosis	11 – 70%
Multi-lobular cirrhosis	5 – 15%
Synthetic liver failure	Infrequent

J Pediatr Gastroenterol Nutr 1999; 28 Suppl 1:S1-S13. | *J Cyst Fibros* 2013;12(2):116-24. | *J Cyst Fibros* 2017;16:S50-61.

CFLD Classification

CFLD **WITH** cirrhosis / portal hypertension

Diagnosis based on the following

- Clinical exam
- Imaging
- Histology
- Laparoscopy

CFLD **WITHOUT** cirrhosis / portal hypertension

Must have at least ONE of the following

- Persistent AST, ALT and GGT >2x ULN
- Intermittently elevated AST, ALT or GGT
- Steatosis
- Fibrosis
- Cholangiopathy
- US abnormalities inconsistent with cirrhosis

CFLD: cystic fibrosis-related liver disease AST: aspartate aminotransferase
ALT: alanine aminotransferase GGT: gamma-glutamyltransferase
US: ultrasound

J Pediatr Gastroenterol Nutr 1999; 28 Suppl 1:S1-S13. | *J Cyst Fibros* 2013;12(2):116-24.

CFLD Management

Liver and Biliary Tract Disease in Cystic Fibrosis Guidelines (1999)

Cystic Fibrosis-Related Cirrhosis Consensus Statement (2017)

Referral	Nutrition	Liver Disease Reduction	Ursodeoxycholic Acid (UDCA)
Hepatology	Increase in daily caloric intake by 50%	Hepatitis A and B immunization	?
Liver biopsy versus imaging	Evaluation and repletion of fat-soluble vitamins	Avoidance of alcohol and hepatotoxic substances	

J Pediatr Gastroenterol Nutr 1999; 28 Suppl 1:S1-S13. | *J Cyst Fibros* 2013;12(2):116-24. | *J Cyst Fibros* 2017;16:S50-61.

Ursodeoxycholic acid (UDCA) / Ursodiol

Proposed Mechanism of Action - CFLD

- Increase hepato- and cholangiocellular secretion → increased bile flow

Dosing

- 15 – 20 mg/kg/day in divided doses (MAX 450 mg/day)

Primary Biliary Cirrhosis:
10 – 15 mg/kg/day

Adverse Reactions (>10%)

- GI (diarrhea, constipation, nausea, dyspepsia), headache, dizziness

Contraindications for Use

- Allergy to bile acids

J Cyst Fibros 2017;16:S50-61. | Ursodiol [prescribing information]. Irvine, CA. Allergan USA Inc. 2018.

UDCA – Effectiveness in CFLD

	Design	Population	Intervention	Results
Colombo C, et al 1996[1]	Multicenter, randomized, double-blind, placebo-controlled (n=55)	CFLD • Male, 13.8 years	UDCA 15 mg/kg/day + taurine 30 mg/kg/day (n=15) vs. UDCA + placebo (n=15) vs. Taurine + placebo (n=12) vs. Placebo + placebo (n=12) X 1 year	• UDCA ↓GGT (p= 0.06) • Stable clinical condition with UDCA via Shwachman-Kulczycki Score (p= 0.02) • No severe side effects reported

UDCA: ursodeoxycholic acid CFLD: cystic fibrosis-related liver disease
GGT: gamma-glutamyl transferase

1. *Hepatology* 1996;23:1484-90.

UDCA – Effectiveness in CFLD

	Design	Population	Intervention	Results
Colombo C, et al 1996[1]	Multicenter, randomized, double-blind, placebo-controlled (n=55)	CFLD • Male, 13.8 years	UDCA 15 mg/kg/day + taurine 30 mg/kg/day (n=15) vs. UDCA + placebo (n=15) vs. Taurine + placebo (n=12) vs. Placebo + placebo (n=12) X 1 year	• UDCA ↓GGT (p= 0.06) • Stable clinical condition with UDCA via Shwachman-Kulczycki Score (p= 0.02) • No severe side effects reported
van de Meeberg PC, et al 1997[2]	Single center, randomized (n=30)	CFLD • Male, 18.3 years	UDCA 10 mg/kg/day (n=17) vs. UDCA 20 mg/kg/day (n=13) X 1 year	• High-dose UDCA: ↓ AP (p< 0.01), GGT (p< 0.002), AST (p< 0.01), ALT (p< 0.02)

UDCA: ursodeoxycholic acid
GGT: gamma-glutamyl transferase
AST: aspartate aminotransferase
CFLD: cystic fibrosis-related liver disease
AP: alkaline phosphatase
ALT: alanine aminotransferase

1. *Hepatology* 1996;23:1484-90. | 2. *Scand J Gastroenterol* 1997;32:369-73.

UDCA – Safety in CFLD

	Design	Population	Intervention	Results
Siano M, et al 2010[1]	Single center, retrospective cohort (n=26)	CF with MI • 8.4 years • 1989-2008	Early UDCA (n=14) vs. CFLD-onset UDCA (n=12) *Mean dose: 15 mg/kg/day	• Prevalence of chronic liver disease: CFLD vs Early (40% vs 0%, p<0.05) • No significant differences seen in glucose control or GI side effects

CF: cystic fibrosis
MI: meconium ileus
UDCA: ursodeoxycholic acid
CFLD: cystic fibrosis-related liver disease

1. *Digestive and Liver Disease* 2010;42:428-31

UDCA – Safety in CFLD

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Colombo C, et al 2016[2]	Single, center, prospective, observational (n=20)	CFLD • 16 years • Cirrhosis (n=13)	UDCA 20 mg/kg/day Duration: at least 2 years (median = 8 years)	• Primary serum bile acid: UDCA • No significant changes in lithocholic acid concentrations observed

UDCA 15 – 20 mg/kg/day is safe and effective for the management of CFLD

CF: cystic fibrosis
MI: meconium ileus
UDCA: ursodeoxycholic acid
CFLD: cystic fibrosis-related liver disease

1. *Digestive and Liver Disease* 2010;42:428-31. | 2. *J Pediatrics* 2016;177:59-65.

Audience Participation Question

Which of the following describes the pathophysiology of cystic fibrosis-related liver disease (CFLD)?

A. Gain of CFTR function , biliary obstruction , subsequent fibrosis and cirrhosis

B. Loss of CFTR function , biliary obstruction , subsequent fibrosis and cirrhosis

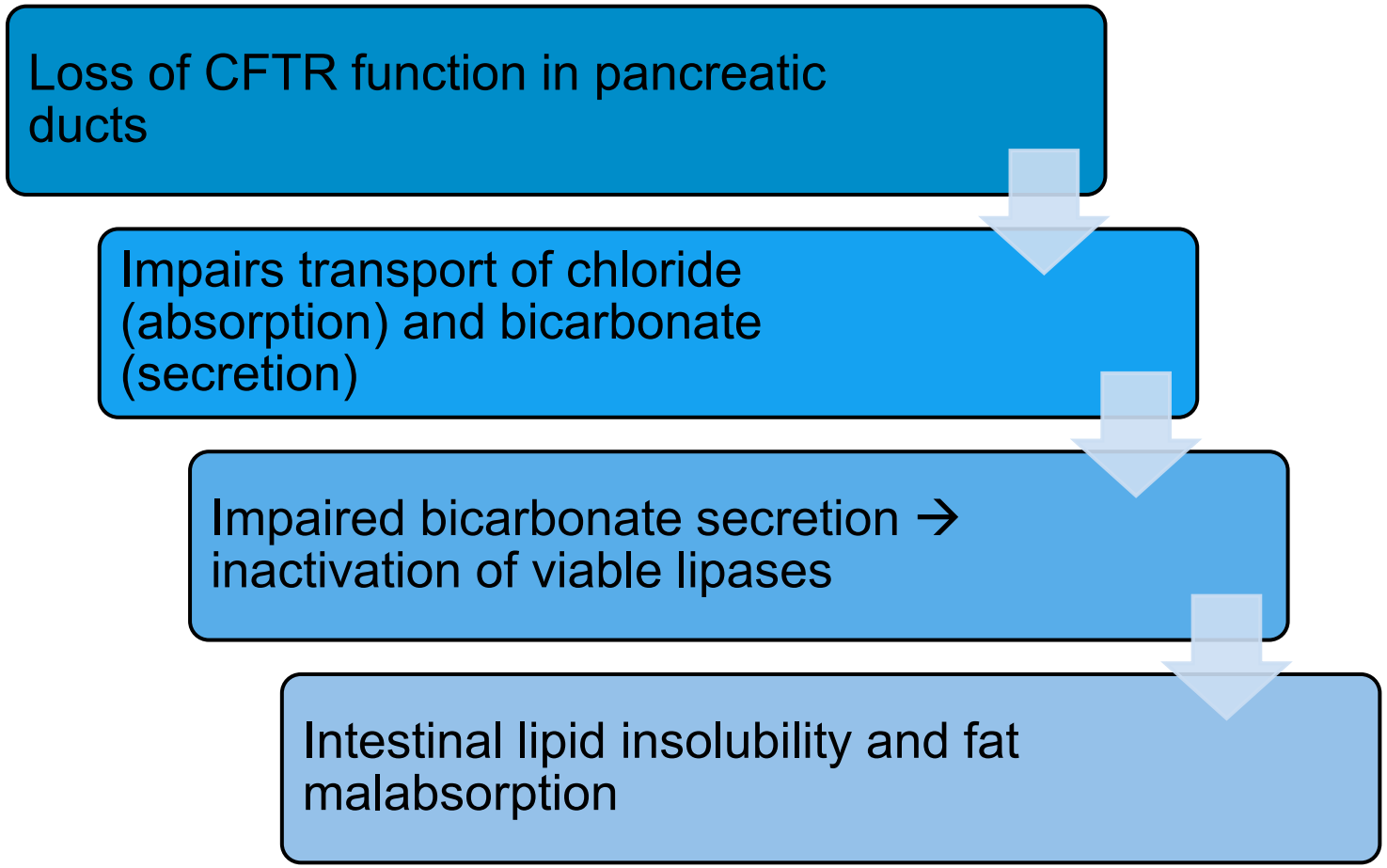
C. Loss of CFTR function , increase in biliary secretion , subsequent fibrosis and cirrhosis

D. Gain of CFTR function , biliary obstruction , improvement in overall liver function

CFTR: cystic fibrosis transmembrane conductance regulator

Pancreatic Insufficiency

Pancreatic Insufficiency



Presentation

- Weight loss
- Gas, bloating
- Dyspepsia
- Steatorrhea (loose, oily stool)

CFTR: cystic fibrosis transmembrane conductance regulator

J Cyst Fibros 2017;16:S70-8. | *Neth J Med* 1992;41:97-100. | *J Pathol* 2016;238:311-20.

Pancreatic Insufficiency

Pancreatic Insufficient

- 2 severe CFTR Mutations
- Classes I, II, III, VI
- Early diagnosis (birth)

Pancreatic Sufficient

- 2 mild CFTR Mutations
- Classes IV and V
  OR 1 severe and 1 mild
- Able to grow and maintain health without supplemental enzyme therapy

CFTR: cystic fibrosis transmembrane conductance regulator

J Cyst Fibros 2017;16:S70-8.

Pancreatic Insufficiency Management

Pancreatic Enzyme Therapy in Cystic Fibrosis Guidelines (1995)

Pancreatic Insufficiency in Cystic Fibrosis Consensus Statement (2017)

Diagnosis

72h stool fat estimation (gold standard) > 7g fat/ 24hr

Fecal elastase-1 (most common) < 100 mcg/g

Nutrition

Evaluation and repletion of fat-soluble vitamins

Pancreatic Enzyme Replacement Therapy (PERT)

For those who classify as “pancreatic insufficient”

J Cyst Fibros 2017;16:S70-8.

Pancreatic Enzyme Replacement Therapy (PERT)

FDA-Approved PERT Products

- Creon®
- Pancreaze®
- Ultresa®
- Zenpep®
- Pertzye®

Formula Characteristics

- Enteric-coated, gelatin capsule
- Porcine-derived
- Dissolves at pH 5 - 5.5 (more distal)
- No acid suppression required

Dosage Strength and Administration

- Strength: units of lipase per capsule in 1000s
- Should be taken during or just after a meal
- May open capsule and mix with small amount of soft, acidic food (i.e. applesauce)

J Cyst Fibros 2017;16:S70-8.

PERT Dosing Strategies

Pancreatic Insufficiency in Cystic Fibrosis Consensus Statement (2017)

Dosing by Body Weight

- 500 – 2,500 units lipase/kg/meal
- Half of this content with snacks

Dosing by Meal Fat Content

- 500 – 4,000 units of lipase per gram of fat ingested per day
- Mimics natural pancreatic enzyme response

Difficult for some patients to calculate

Dosing by Pancreatic Lipase Output

- 480,000 – 960,000 units of lipase secreted during a meal
- 50,000 – 100,000 units of lipase required per meal (> 5-10% of normal levels)

Not typically done in cystic fibrosis

J Cyst Fibros 2017;16:S70-8.

PERT: pancreatic enzyme replacement therapy

PERT Products – Mayo Clinic Rochester

Creon® 3,000 units	Lipase 3,000 units Protease 9,500 units Amylase 15,000 units
Creon® 6,000 units	Lipase 6,000 units Protease 19,000 units Amylase 30,000 units
Creon® 12,000 units	Lipase 12,000 units Protease 38,000 units Amylase 60,000 units
Creon® 24,000 units	Lipase 24,000 units Protease 76,000 units Amylase 120,000 units
Creon® 36,000 units	Lipase 36,000 units Protease 114,000 units Amylase 180,000 units

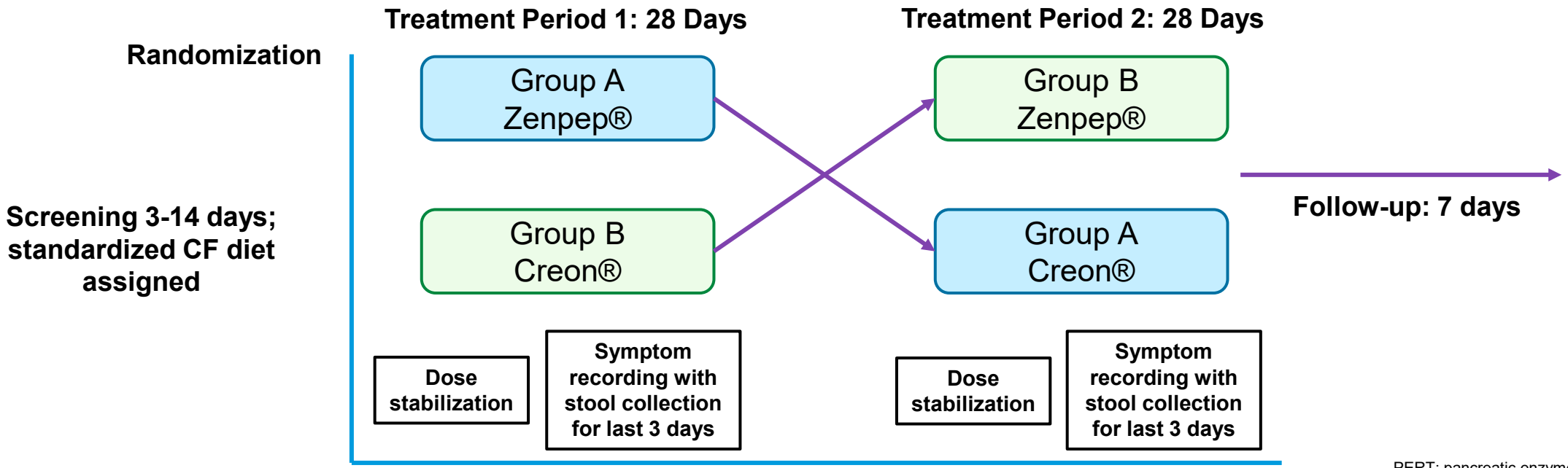
Zenpep® 5,000 units	Lipase 5,000 units Protease 17,000 units Amylase 24,000 units
Zenpep® 10,000 units	Lipase 10,000 units Protease 32,000 units Amylase 42,000 units
Zenpep® 15,000 units	Lipase 15,000 units Protease 47,000 units Amylase 63,000 units
Zenpep® 20,000 units	Lipase 20,000 units Protease 63,000 units Amylase 84,000 units
Zenpep® 25,000 units	Lipase 25,000 units Protease 79,000 units Amylase 105,000 units

Creon [prescribing information]. North Chicago, IL; AbbVie Inc. 2015. | Zenpep [precribing information]. Irvine, CA: Allergan USA, Inc; 2017.

PERT: pancreatic enzyme replacement therapy

PERT – Effectiveness in Pancreatic Insufficiency

Taylor CJ, et al 2016	
Design	Multicenter, randomized, double-blind, active-controlled, crossover, non-inferiority (n=96)
Population	CF (Male, mean age 19.2 years) with PI on PERT
Intervention	Zenpep® vs Creon® x 28 days (MAX 10,000 lipase units/kg/day OR 4,000 units/g fat/day)



PERT: pancreatic enzyme replacement therapy
CF: cystic fibrosis

J Cyst Fibros 2016;15:675-80.

PERT – Effectiveness in Pancreatic Insufficiency

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Population	CF (Male, mean age 19.2 years) with PI on PERT
Intervention	Zenpep® vs Creon® x 28 days (MAX 10,000 lipase units/kg/day OR 4,000 units/g fat/day)

Results	• CFA-72h: Zenpep® (84.1%) vs Creon® (85.3%) – non-inferior
	• Signs/Symptoms of PI
	  • Average number stools/day: Zenpep® (1.5) vs Creon® (1.5)
	  • No difference in number of patient-days with bloating
	• Similar in overall health, well-being, and CF symptoms (except lung function)

CFA-72h: {[dietary fat intake (grams) − stool fat excretion (grams)] / dietary fat intake (grams)} × 100

J Cyst Fibros 2016;15:675-80.

PERT: pancreatic enzyme replacement therapy
CFA-72h: coefficient of fat absorption over 72 hours
CF: cystic fibrosis
PI: pancreatic insufficiency

Audience Participation Question

The 2017 Pancreatic Insufficiency in Cystic Fibrosis Consensus Statement recommends which of the following for the management of pancreatic insufficiency?

A. Evaluation and repletion of water-soluble vitamins

B. Increase fat in diet to promote pancreatic activity

C. PERT (i.e. Creon®) dosed 500 – 2,500 units amylase/kg/meal + none with snacks

D. PERT (i.e. Zenpep®) dosed 500 – 2,500 units lipase/kg/meal + half content with snacks

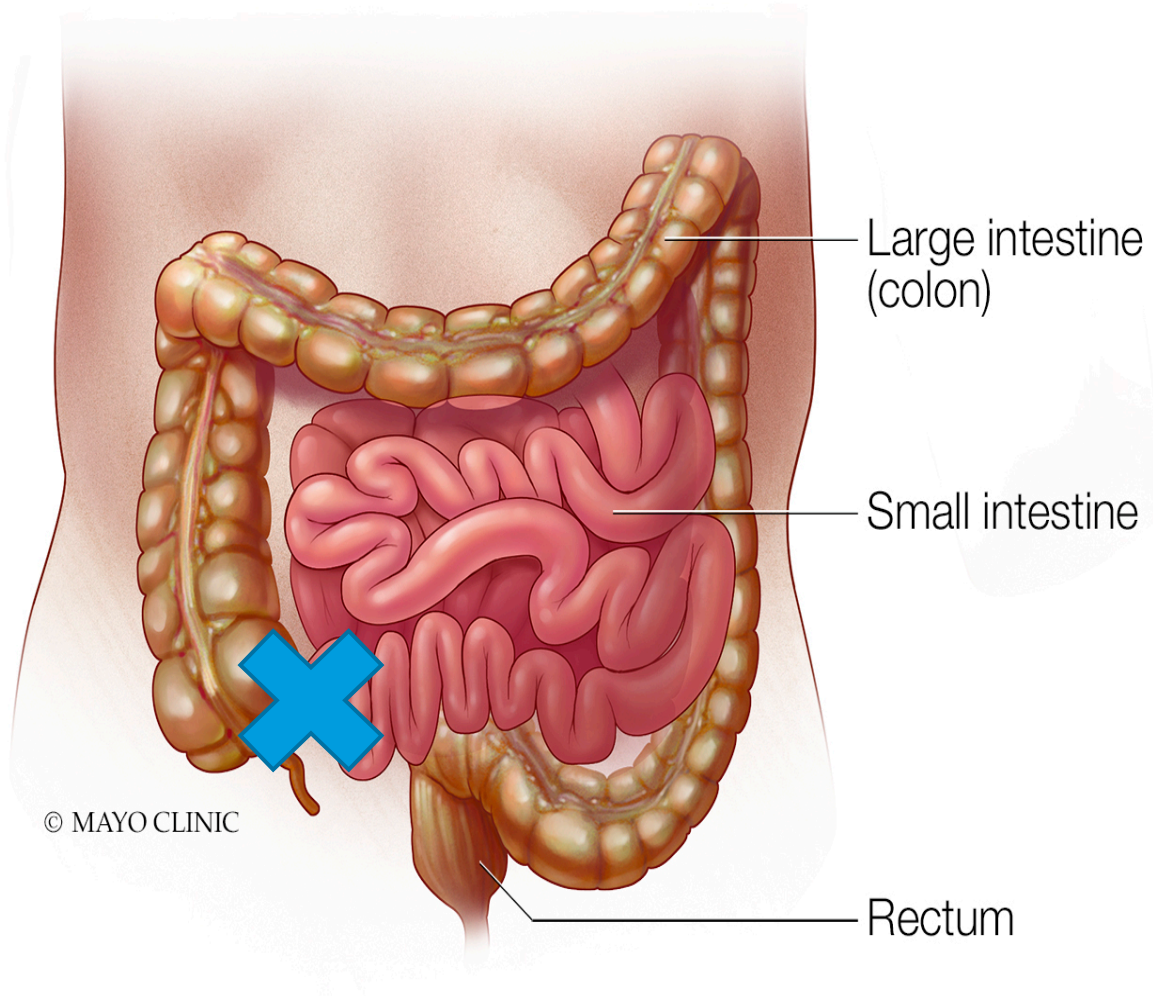
Distal Intestinal Obstruction Syndrome (DIOS)

Distal Intestinal Obstruction Syndrome (DIOS)

Loss of CFTR function → decreased chloride and fluid secretion

Dehydrated mucosa, altered mucin properties → mucus plugging, viscid stool

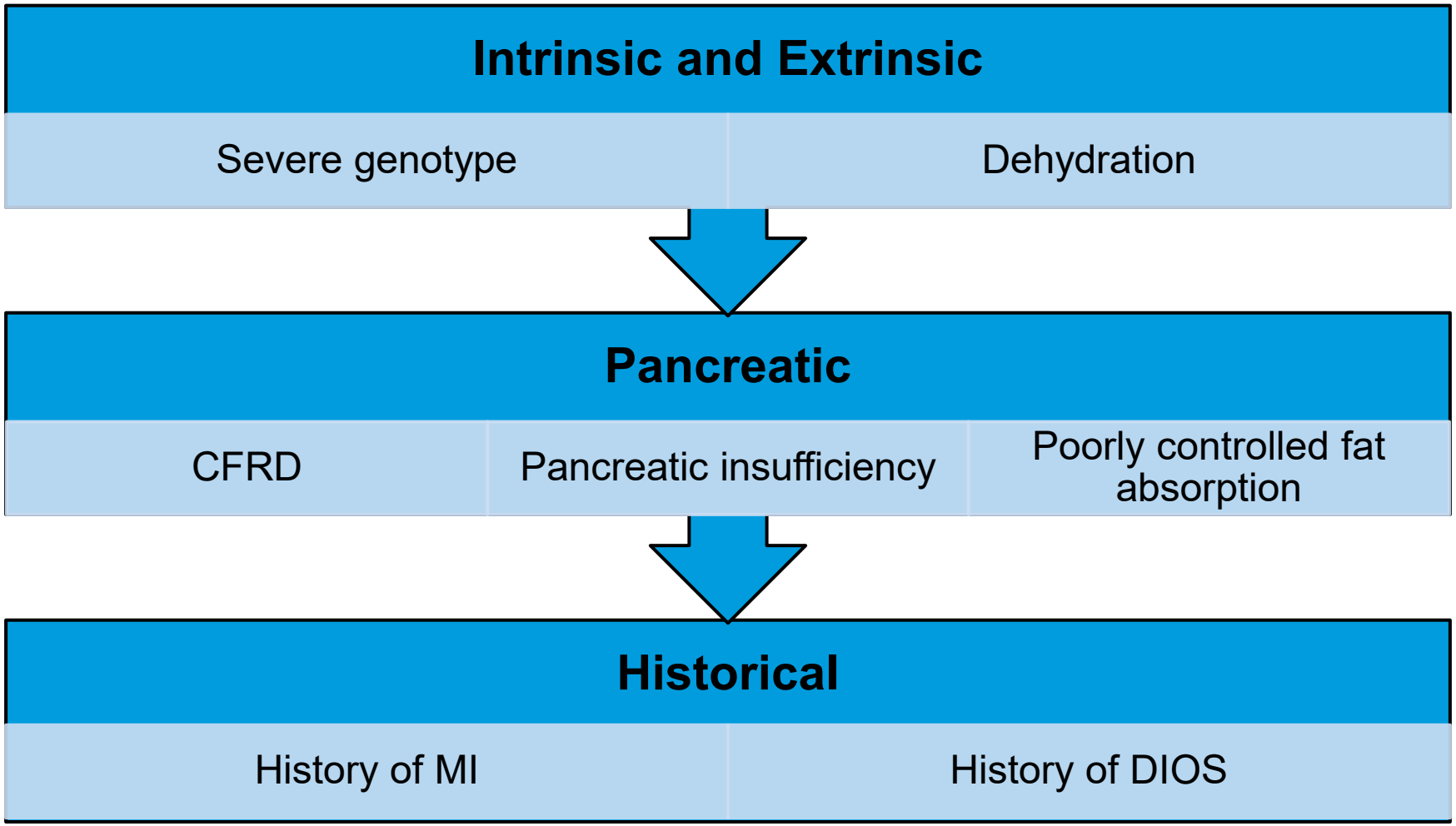
Prolongation of gut transit and accumulation in distal ileum and proximal colon → obstruction



CFTR: cystic fibrosis transmembrane conductance regulator

J Cyst Fibros 2017;16:S40-9.

DIOS Risk Factors



CFRD: cystic fibrosis-related diabetes
MI: meconium ileus
DIOS: distal intestinal obstruction syndrome

J Cyst Fibros 2017;16:S40-9. | *JPGN* 2010;50:38-42.

DIOS Classification

CF Related Constipation

- Abdominal pain and/or distension
- Reduced frequency of BMs in the last few weeks/months
- Increased stool consistency in the last few weeks/months
- Symptoms relieved using laxatives

Incomplete or Impending DIOS

- Fecal mass in the ileo-cecum
- Abdominal pain and/or distension

Complete DIOS

- Fecal mass in the ileo-cecum
- Abdominal pain and/or distension
- Complete intestinal obstruction: vomiting bilious material and/or fluid levels in intestine on abdominal imaging

J Cyst Fibros 2017;16:S40-9. | *JPGN* 2010;50:38-42.

CF: cystic fibrosis
DIOS: distal intestinal obstruction syndrome

Distal Intestinal Obstruction Syndrome Management

Cystic Fibrosis & Disorders of the Large Intestine – DIOS Consensus Statement (2017)

Diagnosis

- Abdominal radiography
- Symptoms (see previous slide)

Nutrition

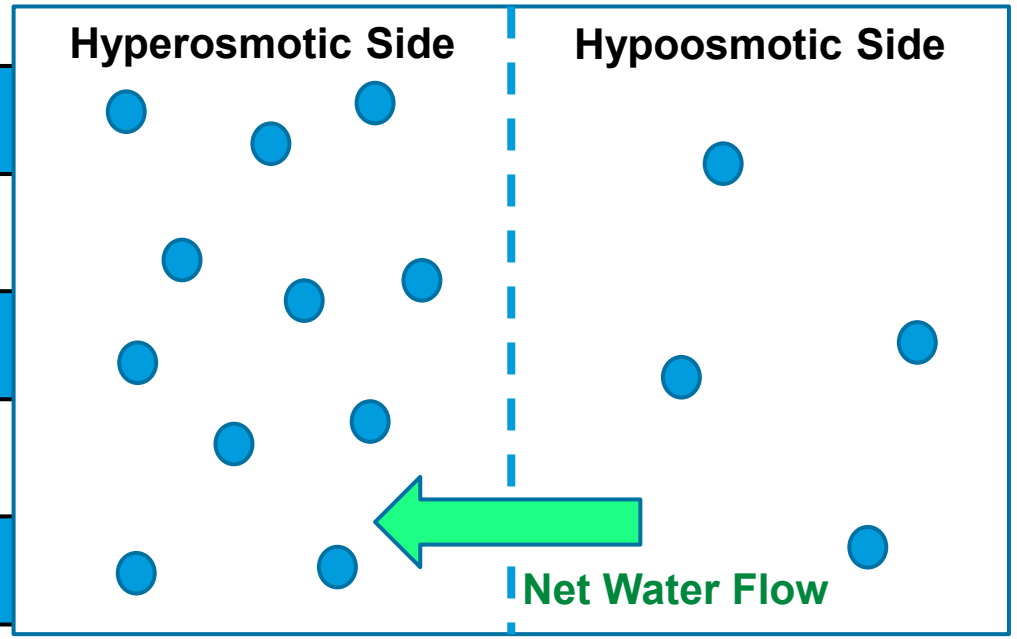
- Bowel rest
- Urgent nasogastric decompression (complete DIOS)

Management

- Osmotic Laxatives
- Sodium meglumine diatrizoate (Gastrografin®)
- N-acetyl cysteine

J Cyst Fibros 2017;16:S40-9. | *JPGN* 2010;50:38-42.

Diatrizoate meglumine and diatrizoate sodium (Gastrografin®)



Mechanism of Action

- Hypertonic radiopaque (iodine-containing) contrast agent
- Highly osmolar (osmolality 2150 mOsm/kg H_2O at 37°C)

Dosage Form

- Sodium diatrizoate 100 mg/mL
- Meglumine diatrizoate 660 mg/mL

Adverse Reactions (undefined frequency)

- GI (diarrhea, nausea, vomiting)
- Respiratory (dyspnea, hypoxia)
- Erythema, urticaria
- Hypotension secondary to fluid shifts

Precautions for Use

- CI with iodine hypersensitivity, pre-existing dehydration, prolonged obstruction

GI: gastrointestinal
CI: contraindicated

J Cyst Fibros 2017;16:S40-9. | *JPGN* 2010;50:38-42. | Diatrizoate meglumine and diatrizoate sodium [prescribing information]. Princeton, NJ: Bracco Diagnostics Inc.; 2013.

DIOS Treatments – Effectiveness

	Design	Population	Intervention	Results
O'Halloran S, et al. 1986[1]	Single center, case series (n=67, 37 episodes)	CF, DIOS episode • Female, 9.8 years (mean) • Follow-up: 12 months	Oral Gastrografin® (50mL or 100mL) solution followed by 4x oral volume repletion	• 30 episodes treated with single dose (81%) • 6 episodes required a second dose 24 hours later (16%) • 1 treatment failure requiring Gastrografin® enema (3%)

CF: cystic fibrosis DIOS: distal intestinal obstruction syndrome.

1. *Archives of Disease in Childhood* 1986;61:1128-30.

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Koletzko S, et al. 1989[2]	Case series (n=22)	CF, R iliac fossa mass • Male, 21.8 years (mean) • Follow-up: 26 months	Iso-osmotic polyethylene glycol 5.6 +/- 1.9L oral or NG x 5.6 +/- 2.4 hours	• ↓ radiologic impaction signs ($p<0.001$) and hospital duration ($p<0.02$) • Mean 3 months symptom/pain free with minor adverse effects

CF: cystic fibrosis DIOS: distal intestinal obstruction syndrome R: right. NG: nasogastric

1. *Archives of Disease in Childhood* 1986;61:1128-30. | 2. *Pediatrics* 1989;83:727-33.

DIOS Treatments – Effectiveness

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Zahra M, et al. 2014[3]	Single center, case series (n=8, 12 episodes)	CF, DIOS • Male, 22.5 years (mean)	Gastrografin® enema • Mean dilution 1220 mL OR 31 mL/kg • Range 240 – 3500 mL OR 19 – 59 mL/kg	• Complete resolution (100%) • 3 patients required repeat enemas on consecutive days • 1 patients with 3 enemas on 3 consecutive days

CF: cystic fibrosis DIOS: distal intestinal obstruction syndrome R: right
NG: nasogastric

1. *Archives of Disease in Childhood* 1986;61:1128-30. | 2. *Pediatrics* 1989;83:727-33. | 3. *Journal of Pharmacy and Nutrition Sciences* 2014;4:76-80.

DIOS Treatments – Effectiveness

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Munck A, et al. 2016[4]	International, multicenter, prospective, observational (n=102 patients, 112 episodes)	CF, new DIOS • Male, 14.7 years (median) • Follow-up: 1.6 years (median)	Gastrografin® enema (n=36/108) Gastrografin® enema + PEG lavage (n=11/108) PEG lavage (n=52/110) Oral osmotic laxative (n=73/106) Colonoscopy or Surgery (n=6/112)	• Medical treatment was successful for all IDIOS episodes • Most patients with CDIOS were treated with Gastrografin® • Medical failure occurred in 6 patients (all CDIOS, required colonoscopy or surgery)

CF: cystic fibrosis DIOS: distal intestinal obstruction syndrome R: right
NG: nasogastric PEG: polyethylene glycol IDIOS: incomplete DIOS CDIOS: complete DIOS

1. *Archives of Disease in Childhood* 1986;61:1128-30. | 2. *Pediatrics* 1989;83:727-33. | 3. *Journal of Pharmacy and Nutrition Sciences* 2014;4:76-80. | 4. *J Cyst Fibros* 2016;15:531-9.

Incomplete DIOS – Recommendations

Cystic Fibrosis & Disorders of the Large Intestine – DIOS Consensus Statement (2017)

Oral rehydration +

Polyethylene glycol 2 g/kg/day (max 80 – 100 g/day)

OR

Iso-osmotic PEG (i.e. Golytely® 20 – 40 mL/kg/hour (max 1L/hour over 8 hours)

Diatrizoate meglumine and diatrizoate sodium (Gastrografin®) PO/NG 100 mL (diluted in 400 mL) on Day 1 + half dose on subsequent days if needed

NG: nasogastric

J Cyst Fibros 2017;16:S40-9. | *JPGN* 2010;50:38-42.

Complete DIOS – Recommendations

Cystic Fibrosis & Disorders of the Large Intestine – DIOS Consensus Statement (2017)

Moderate obstruction without vomiting

Oral rehydration +

Polyethylene glycol 2 g/kg/day (max 80 – 100 g/day)

OR

Iso-osmotic PEG (i.e. Golytely® 20 – 40 mL/kg/hour (max 1L/hour over 8 hours)

Failure

More severe obstruction with bilious vomiting

IV hydration +

NG decompression +

Diatrizoate meglumine and diatrizoate sodium (Gastrografin®) enema 100 mL diluted x 4 with water as retrograde lavage with hydrostatic pressure (IR procedure)

IV: intravenous
IR: interventional radiology

J Cyst Fibros 2017;16:S40-9. | *JPGN* 2010;50:38-42.

DIOS Prevention – Recommendations

Cystic Fibrosis & Disorders of the Large Intestine – DIOS Consensus Statement (2017)

Prophylaxis with history of DIOS

- Polyethylene glycol 0.5 – 1 g/kg/day (max 40 g/day) x 6-12 months
- Lactulose

→ Avoidance of dehydration

→ Reassess adequate PERT

J Cyst Fibros 2017;16:S40-9. | *JPGN* 2010;50:38-42.

PERT: pancreatic enzyme replacement therapy

Audience Participation Question

AB is a 27 yo female with a PMHx significant for CF with PI who was admitted from CF clinic for worsening abdominal pain and constipation. Her current symptoms include 10/10 cramping abdominal pain and nausea without vomiting. She states she has not had time to get to the grocery in the past week and has been eating more fast food with limited water intake. CT abdomen/pelvis reveals a large, obstructing mass in the right ileocecal region with inspissated bilious material.

Audience Participation Question

Given the current presentation, how would you classify AB's DIOS episode per the 2017 Cystic Fibrosis & Disorders of the Large Intestine – DIOS Concensus Statement?

A. CF-Related Constipation

B. Incomplete DIOS

C. Complete DIOS

D. CF-Related Diarrhea

Audience Participation Question

During rounds, AB has had multiple episodes of bilious vomiting without improvement in symptoms despite multiple treatments with polyethylene glycol.

Audience Participation Question

Which of the following would be an appropriate next treatment for AB's DIOS episode?

A. Oral lactulose titrated to bowel movements

B. IV rehydration + NG decompression + oral diatrizoate meglumine and diatrizoate sodium (Gastrografin®)

C. IV rehydration + NG decompression + diatrizoate meglumine and diatrizoate sodium (Gastrografin®) enema

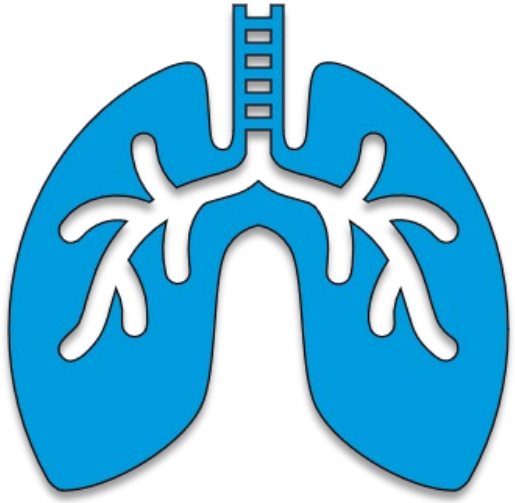
D. Re-trial oral polyethylene glycol + rectal lactulose titrated to bowel movements

Summary

- Cystic fibrosis is an autosomal recessive disease that carries a high risk of mortality and extrapulmonary complications
- Ursodiol may prevent the progression of cystic fibrosis – related liver disease, but larger trials with long-term data is needed
- Pancreatic insufficiency is managed with pancreatic enzyme replacement therapy that is individualized to the patient in order to prevent malabsorption
- Those patients with DIOS should receive aggressive bowel regimens, possibly including sodium meglumine diatrizoate, in order to prevent surgical interventions

DIOS: distal intestinal obstruction syndrome

MAYO CLINIC



It’s Not Just the Lungs?

Non-Pulmonary Complications of Cystic Fibrosis

Ashley Otto, PharmD, BCPS

Pharmacy Grand Rounds
January 5, 2021

