



# INTERNATIONAL PROSPECTIVE STUDY ON DISTAL INTESTINAL OBSTRUCTION SYNDROME (DIOS). ASSOCIATED FACTORS and OUTCOMES..



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# NACFF Phoenix: Round-table

- 14 y.o. female, PI
- Compliant with enzymes /meds
- Central/lower abdo pain
- Getting worse
- Stool every day

# Case continued

- On exam, mild tenderness lower abdo
- Labs unremarkable
- Imaging not diagnostic
- Mega-work up including gynecology!!
- Diagnosis of DIOS was missed!!!!

# Abdominal pain (partial list)

- CF Related/associated
  - GERD
  - cholecystitis/cholangitis
  - Pancreatitis (PS)
  - Fibrosing colonopathy
  - intussusception
  - tumors
- CF not related
  - CMPH
  - IBD
  - PUD
  - celiac disease
  - Appendix
  - RAP

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- DIOS....constipation

# DIOS

- Chronic, recurrent form of partial intestinal obstruction - rarely complete
- More common in adults
- PI
- Unique to CF



# Proposed Precipitating factors

- Abnormal mucus
- Dehydration of luminal contents
- Poor compliance with PERT
- h/o meconium ileus ?
- Major surgery eg post transplant
- Low fibre ?
- Dysmotility ?
- drugs eg analgesics

# Symptoms

- Progressive, colicky abdominal pain
- Bloating
- Nausea
- Anorexia
- **Not necessarily reduced bowel movements**



# Examination

- Distended abdomen
- Mass in right lower quadrant
- Don't "forget" rectal exam!

# DIOS POST TRANSPLANTATION



*Nassenstein K et al Gut 2005;54:1662-3*

# DIOS in adults

*Clin Gastr Hep 2004;2:498-503*

- 27 patients reported 43 episodes (n=171)
- incidence of 15.8% (other studies 10-20%)
- No association with meconium ileus ( other studies disagree)

# Defining Distal Intestinal Obstruction Syndrome (DIOS) and Constipation in Cystic Fibrosis: A multicenter study on the incidence, characteristics and treatment of DIOS

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## ESPGHAN CF Working Group definition for DIOS in Cystic Fibrosis

#1	complete intestinal obstruction as evidenced by vomiting of bilious material and/or fluid levels in small intestine on an abdominal radiography
#2	fecal mass in ileo-cecal area
#3	abdominal pain and/or distension

*Complete DIOS: #1, #2 and #3*

*Incomplete/ Impending DIOS: #2 and #3, without #1*

**NOT NECESSARY TO HAVE CONSTIPATION!!!!!!**

# METHODS

- 28 Centers in 10 countries reported new cases of DIOS from 2009-2012 in a study organized by the CF ESPGHAN Working Group and funded by Vaincre La Mucoviscidose (Grant, N° 10RC1017)
- DIOS classification was based on the ESPGHAN CF Working Group Criteria (Houwen R JPGN 2010).
  1. complete intestinal obstruction: bilious vomiting or small intestine fluid levels on X-ray
  2. fecal mass in ileocaecum
  3. abdominal pain and/or distension

« Prospective multicentre study of distal intestinal obstructive syndrome (DIOS): treatment alternatives and concomitant risk factors »

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# Demographic characteristics

Sex M/F, N (%)	61/41 (60/40)	
Genotype , N ( %)		
Severe	89 (87)	
Mild	1 (1)	
Undetermined	12 (12)	
Meconium ileus (MI),N(%)	40 (40)	
Pancreatic insufficiency (PI), N (%)	94 (92)	
CF liver disease, N (%)	22 (22)	
CF related diabetes $\geq 18$ y	20/41 (49)	
Median age at entry in the study (y)	14.7 [6.3;23.8]	



## ***Proposed risk factors for DIOS***

	DIOS n=112
<u>Daytime temperature</u> $\leq 20^{\circ}\text{C}$ N,% $> 20^{\circ}\text{C}$ N,%	61 (54) 51 (46)
<u>Intensive sport</u> N,%	9 (8)
<u>Surgery anesthesia</u> ,N,%	4 (4)
<u>Dietetic intake</u> Fiber low, high Fat low, high Dietetic modification	41 (38); 2 (2) 15 (14); 10 (10) 16 (14)
<u>Insufficient hydration</u>	47 (50)
<u>Poor PERT compliance</u>	10 (10)

# Treatment management

N, (%)	C DIOS n=52	I DIOS n=60	p-value Chi <sup>2</sup> -Fisher
<b><u>Hospitalisation</u></b>			
Median duration stay,d	50 (96) 4 [3-7]	51 (85) 3 [1-4]	0.06 <b>0.004</b>
<b><u>Medical treatment</u></b>			
IV hydration	44 (85)	32 (56)	<b>0.002</b>
Fasting	47 (90)	34 (60)	<b>0.001</b>
Nasogastric aspiration	21 (40)	6 (11)	<b>0.004</b>
Pain relief non opioid	42 (81),	32 (53),	<b>0.003</b>
PEG lavage	24 (47)	28 (47)	0.97
Gastrografin enema	29 (58)	7 (12)	<b>&lt;0.001</b>
Gastrografin + PEG	10 (20)	1 (2)	<b>0.02</b>
<b><u>Colonoscopy</u></b>	2 (4)	0	-
<b><u>Surgery</u></b>	4 (8)	0	-
<b><u>Delay for DIOS resolution, d</u></b>	4 [3-7]	4 [3-6]	0.95
<b><u>Maintenance therapy</u></b>	45 (88)	50 (83)	0.46

# *Factors that impact the delay of resolution of DIOS (primary endpoint)*

- Time between symptoms and arrival at CF centre,  
d :  $p < 0.0001$*
- Weight loss compared to usual weight, % :  $p = 0.001$*

# AGE

## **Results**

### **➤ Incidence**

<18 years Incidence [95%], episodes/1000 patients-years: 8,20 [8.17-8.23]

≥ 18 years Incidence [95%], episodes/1000 patients-years: 8,57 [8.53-8.62]

## ***Conclusions of the first DIOS international prospective survey***

Incidence of DIOS was similar in children and adults

Characteristics of DIOS (complete/incomplete) had a significant impact on treatment management

Current medical practices across countries varied but were effective as surgery was rare

Delayed presentation and prior weight loss were factors that impact significantly delay of resolution

Maintenance long term therapy was prescribed

DIOS has a multifactorial aetiology: severe genotype, PI, MI do predispose; diabetes, low fibers

intake, insufficient hydration may be risk factors; climate and poor PERT compliance were not proven risk factors

Thank you!

