

Munchausen By Proxy and the Intestinal Failure Patient



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Disclosures

The following individuals have a relevant financial relationship with a commercial interest(s):

Name	Proprietary Entity	Nature of Financial Relationship
Alex Flores, M.D.	Ingenix, MOOG	Consultant

Individual faculty will disclose any discussion of off-label or unapproved uses.

The following individuals have no relevant financial relationship to report in the last 12 months with a commercial interest:

OUTLINE

1. BACKGROUND/DEFINITIONS

- **INTESTINAL FAILURE**
- **MUNCHAUSEN BY PROXY**

2. CASE SCENARIOS

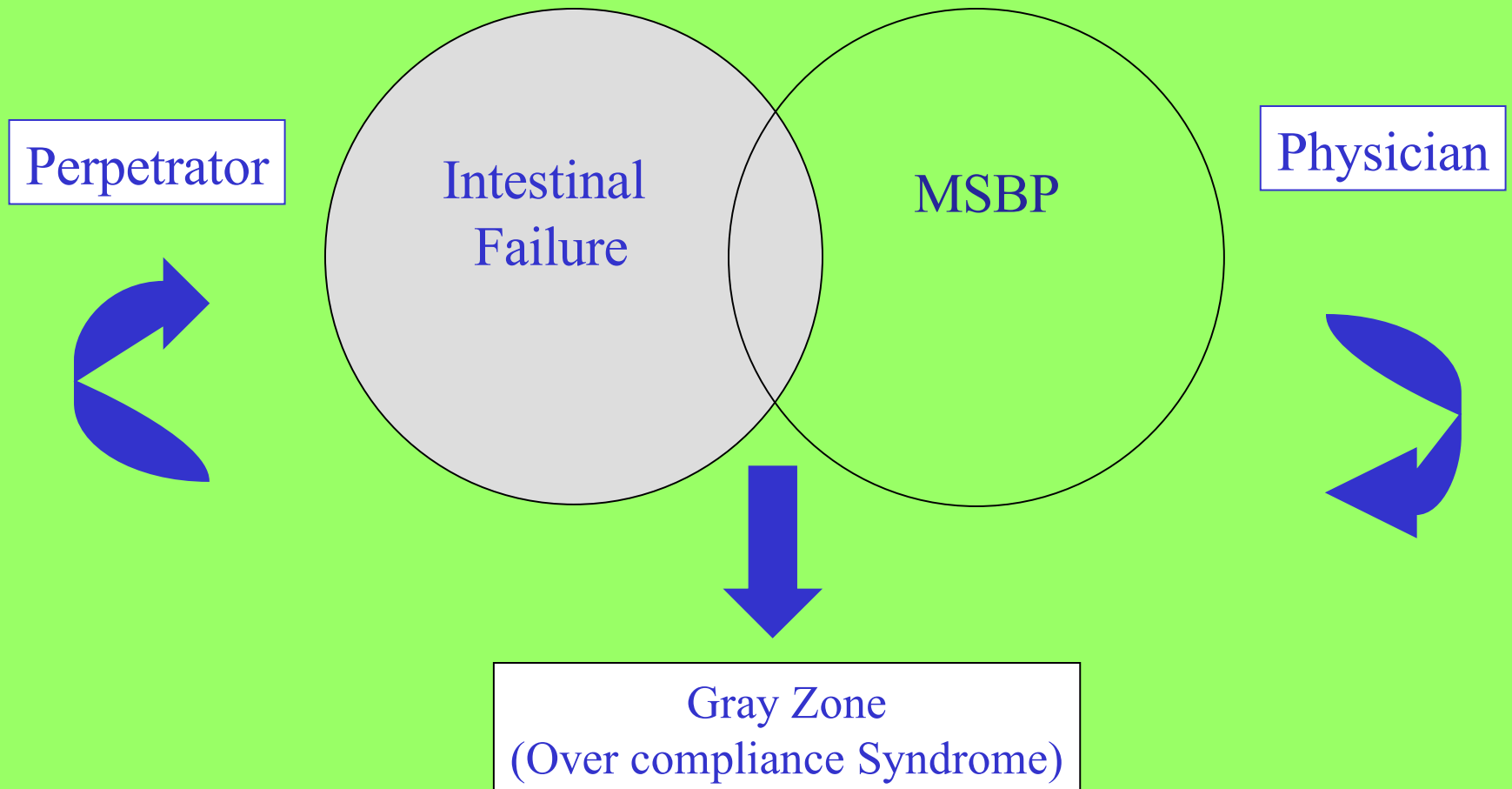
- **THE ONE WHO WAS**
- **THE ONE WHO WASN'T**

3. DIAGNOSIS/MANAGEMENT

4. CONCLUSIONS



The Spectrum of Disease



INTESTINAL FAILURE (IF) DEFINITION

Condition characterized by the inability to maintain protein, energy, fluid, electrolyte or micronutrient balance owing to gastrointestinal disease when on a normal diet. IF ultimately leads to malnutrition and even death if NOT treated by total parenteral nutrition or intestinal transplantation.

Jeejeebhoy in
Gastroenterology 2008; 135: 303-305

FACTS ABOUT IF

- 5 year survival rate with or without liver transplant is 54 – 58 %
 - Deaths due to sepsis, rejection or lymphoma
- 5 year survival on TPN varies according to diagnosis
 - 82% in Crohn's Disease
 - 35 – 40 % in ischemic bowel, radiation enteritis and CIPO

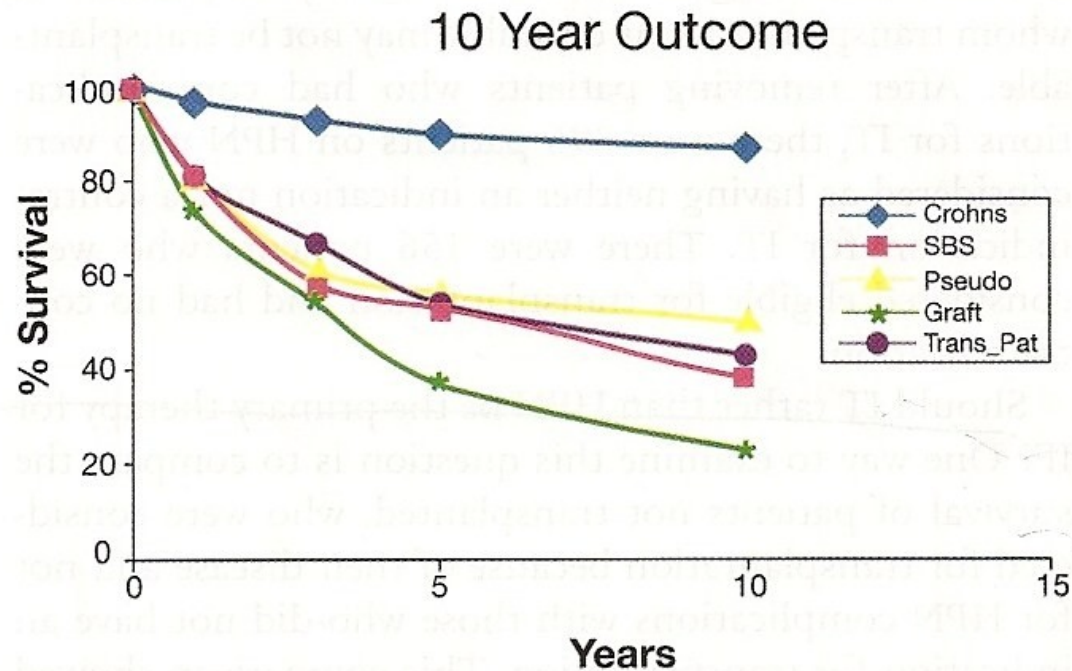


Figure 1. Ten-year survival of HPN, graft, and transplant patients. CROHNS, patients with Crohn's disease on HPN; SBS, patients with short bowel syndrome on HPN; Pseudo, patients with pseudoobstruction on HPN; GRAFT, graft survival in patients receiving intestinal transplant; TRANS_PAT, Patient survival in patients who received intestinal transplant. HPN data from Jeejeebhoy et al.¹⁴ Transplant based on data from Freeman et al.¹³

Current Recommendations for Management of IF

- Initial therapy should be TPN
- Intestinal transplantation is recommended when:

4. Failure of TPN

- Impending or overt liver failure
- Thrombosis of ≥ 2 central veins
- 2 or more episodes of sepsis/year
- Frequent episodes of dehydration

2. High risk of death

- ## **3. Severe short bowel** (G & J tube residual small bowel < 10 cms in infants and < 20 cms in adults

- ## **4. Frequent hospitalizations, narcotic dependency, or CIPO**

- ## **5. Patient unwillingness to accept long term TPN**

American Society of Transplantation
Medicare/Medicaid

Table 6.1 Patient population with intestinal failure. (Reproduced from Pironi et al. [27], with permission from Blackwell Publishing.)

	Adults (N = 688)	Pediatrics (N = 166)
Gender (No.)		
Male/female	293/395	87/79
Age (yr)	52.9 ± 15.2 (18.5–88.0)	6.1 ± 5.1 (0.2–18.0)
Cause of intestinal failure (No. (%))		
Short bowel syndrome	514 (74.7)	87 (52.4)
Motility disorder	124 (18.0)	38 (22.9)
Extensive Enteropathy	35 (5.1)	41 (24.7)
Primary disease in the pediatric population		
Congenital short bowel		42 (25.3)
Congenital mucosal disease		29 (17.5)
Chronic intestinal pseudo-obstruction		29 (17.4)
Volvulus		13 (16.5)
Necrotizing enterocolitis		13 (7.8)
Hirschsprung's disease		9 (5.4)
Crohn's disease		6 (3.6)
Primary disease in the adult population		
Mesenteric ischemia	185 (26.9)	
Crohn's disease	159 (23.1)	
Radiation enteritis	73 (10.6)	
Chronic intestinal pseudo-obstruction	72 (10.5)	
Surgical complications	55 (8.0)	
Familial polyposis	21 (3.0)	
Cancer	17 (2.5)	

Table 7.1 Etiology of short bowel syndrome in children.

	USA (1)	CANADA (2)	France (3)	International (4)
• Atresia	30%	30%	39%	23%
• Volvulus	10%	10%	24%	24%
• Gastroschisis	17%	12.5%	14%	14%
• NEC	43%	35%	14%	27%

(Reproduced from Groulet and Sauvat. *Curr Opin Clin Nutr Metab Care* 2006;304–13, with permission from Lippincott Williams and Wilkins.)

GASTROSCHISIS



COURTESY DR. HENRIK
EHRÉN

“ ALWAYS LISTEN TO EXPERTS
THEY’LL TELL YOU WHAT
CAN’T BE DONE AND WHY,
THEN THEY DO IT.”



ROBERT HEINLEIN

Baron Karl Fredrick Von Munchausen



- German mercenary who entertained guests with apocalyptic and fantastic stories of his adventures
- Kept the Royal College of Physicians in London suspended in the air for 3 months!!!
- “It is a well known fact that during the three months the college was suspended in the air, and therefore incapable of attending their patients, No deaths happened, except a few...If the apothecaries had not been very active during the above time, half the undertakers in all probability, would have been bankrupt.”

Raspe
Grosset & Dunlap 1936

Munchausen By Proxy

“It is a form of child maltreatment and a malignant disorder of parenting in which an adult falsifies signs or symptoms in a victim, causing that victim to be regarded as ill or impaired.”



- Meadow R: Munchausen by Proxy:
the hinterland of child abuse
Lancet 1977; 2: 343 – 345
- Asher: Munchausen Syndrome
Lancet 1951; 1: 339 – 341

Components

1. Victimization of a child
2. Psychopathology of the abuse

Other Terms:

3. Pediatric condition/illness falsification
 - * Exaggeration
 - * Simulation
 - * Fabrication
 - * Induction
4. Factitious Disorder by Proxy

REMEMBER...

Most frequent complaints reported by caregivers who falsify ARE GI !!!

Hyman, et al
Child Maltreatment
2002; 7 : 132 – 137

Signs of Pediatric Illness Falsification

- Recurrent illness that appears unusual
- Unexpected symptom occurrence
- Lack of continuity of care and multiple serial providers
- Inconsistencies (false reports, record anomalies)

Manifestations of Munchausen by Proxy in Pediatric GI

- **Chronic Diarrhea**
 - **Failure to Thrive**
 - **Vomiting**
 - **Abdominal Pain**
 - **Hematemesis**
 - **Gastric Erosions**
 - **Mallory-Weis Tear**
1. **Colitis**
 2. **Hematochezia**
 3. **Constipation**
 4. **Cystic Fibrosis**
 5. **CVL Complications**
 6. **Dysmotility / CIPO**
 7. **Mitochondrial disorder**

Ridder L.
J. Pediatric Gastroenterol Nut
2000; 31: 208 – 211

Munchausen by Proxy Clinical Cases (1980 – 2008)

- 28 years in GI practice
- Over 1500 patients evaluated for motility disorders
- Patients:
 4. Dysmotility with multiple line septic episodes – 5 episodes in 12 months. (Pseudomonas, Candida, Enterobacter, Klebsiella, Enterococcus)
 2. Feeding intolerance with CVL, Gastrostomy and Jejunostomy
 3. s/p Fundoplication, gastrostomy, severe retching episodes with pseudo-seizures
 4. Ipecac poisoning

TABLE 1: Features That Differentiate Pediatric Condition Falsification (PCF) From Chronic Intestinal Pseudo-Obstruction (CIP)

<i>Feature</i>	<i>PCF (N = 8)</i>		<i>CIP (N = 14)</i>		<i>p</i>
	<i>n</i>	<i>%</i>	<i>n</i>	<i>%</i>	
Daily abdominal pain	8	100	0	0	< .001
Multisystem disease (> two organ systems ^a)	8	100	0	0	< .001
Accelerating trajectory	8	100	0	0	< .001
Reported preterm birth	5	71.4	1	7.1	< .001
Dilated bowel (x-ray)	0	0	14	100	< .001
Abnormal manometry	2	28.6	14	100	< .001
Genitourinary neuromuscular disease	0	0	8	57	< .01

a. Not including the gastrointestinal system.

MSBP/Falsification and CIPO Dysmotility

WARNING SIGNS !!!

- Symptoms occurring in caregivers presence only
 - Caregiver medically knowledgeable
 - Multiple consultations to experts in same specialty (NOT just a 2nd opinion)
4. Excellent socialization with medical/nursing staff
 2. Team and doctor splitting
 3. Absentee father
 4. Multiple hospitalizations in multiple institutions
 5. Opposition to de-escalate medical care

MSBP: Pattern of Presentation to Pediatric Surgeons

BELIEVE IT: SURGEONS ARE AWARE

- North Carolina Children's Hospital
- Over 5 years ➡ 10 children
- 7 years to 14 years old
- Diagnosis's: apnea, seizures, FTT, GER
- Surgery: G-tube with Nissen fundoplication
- Diagnosis: Video telemetry, toxic screening, separation
- Outcome: 4 children still at home / 6 in foster care

Lacey, et al
J. of Ped Surgery
28: 827 – 831, 1993

Over Interpretation of Gastroduodenal Motility Studies: Two Cases Involving Munchausen Syndrome by Proxy

Patient # 1:

- ❖ 2 ½ yo, male with history of CVL/gastrostomy and feeding intolerance.

Motility Study: Post prandial duodeunal hypomotility. Pain in post prandial period.

Outcome: After separation from mother....OFF TPN and Gastrostomy

Patient # 2:

- ❖ 18 mo, male with hx of CVL, gastrostomy and fundoplication. Multiple line sepsis episodes

Motility Study: NL phase I, II, III (MMC) and disorganized fed pattern.

Outcome: After separation from mother....OFF TPN and Gastrostomy



CAREFUL WITH MOTILITY STUDIES!!

Concerns about Dx of MSBP

1. Illness fabrication
 - Poor history taken by physicians
 - Maternal anxiety
2. Repeated visits to doctors (doctor shopping)
 - Real illness
 - Physician ignorance
3. Perpetrator denies causing illness
 - Innocence of perpetrator
 - Medical blackmail
4. Illness clearance with separation
 - Natural history of disease
 - Mother anxiety affecting child



Conditions That are Pathological but Not MSBP

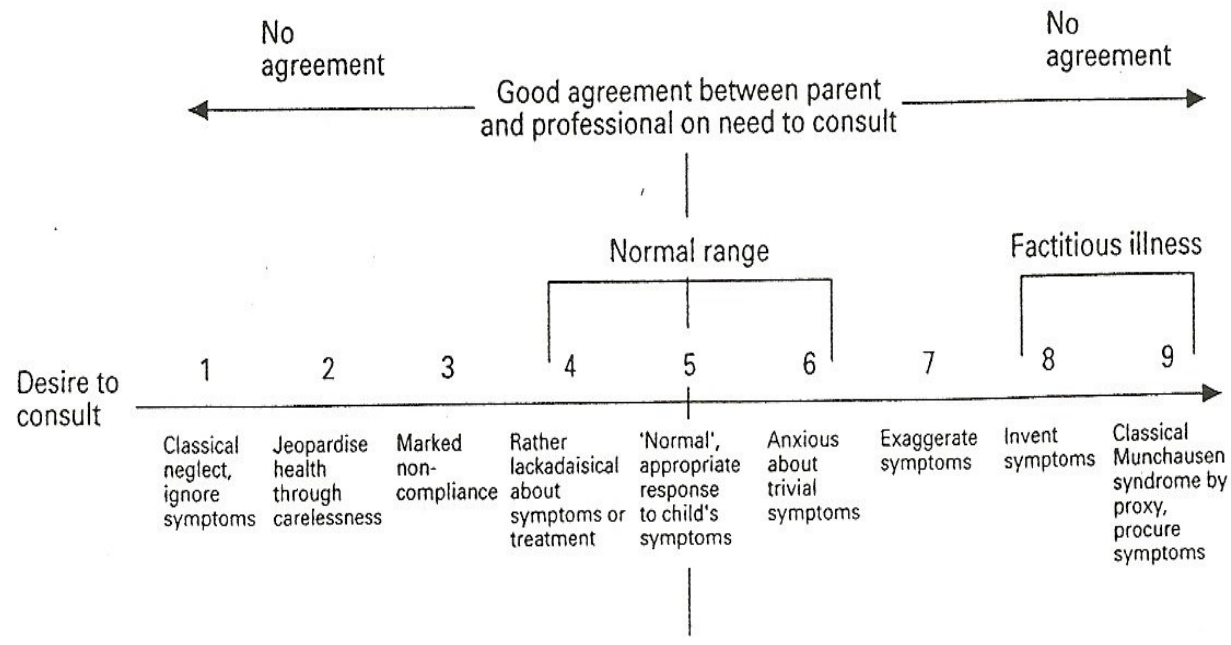
1. Unrecognized child abuse
2. Failure to thrive and/or neglect
3. Over anxious parents
4. Mothers with delusional disorders
5. Hysteria by proxy

Roy Meadow

Criteria for Diagnosis of MSBP AKA Factitious Disorder by Proxy (DSM IV)

- Intentional production or feigning of physical or psychological signs or symptoms in another person who is under the individuals care
- The motivation for the perpetrator's behavior is to assure the sick role by proxy
- External incentives for the behavior (Economic gains are absent)
- The behavior is NOT better accounted for by another mental disorder

Parents' Desire to Consult for Their Child's Symptoms



Parents' desire to consult for their child's symptoms. (Reproduced with permission from Eminson and Postlethwaite.³⁷)



“TRUST BUT
VERIFY”

Case Scenarios

The One Who Was (I)

10 year old female with CIPO evaluated for small bowel transplant

- Presented with abdominal distension
- Esophageal and Antro Duodenal Motility with Neuropathic CIPO
- Required TPN and narcotics for visceral pain
- Multiple surgeries: CVL's, Colectomy
- Eventually had small bowel transplant

The One Who Was (II)

- Initially did very well → recurrence of visceral pain
- Multiple hospital admissions → exploratory laparotomy/splenectomy per PH
- Suspected child abuse → de-escalation implemented → maternal separation → disappearance of symptoms
- Patient healthy on immunosuppression for transplant
- Mother refused psychiatric help

Kosmach, B et al
Transpl Proceeding
1996: 5: 2790 - 2791

Case Scenarios

The One Who Wasn't (I)

- 25 year old female with Hx of CIPO, on TPN/Gastrostomy & Jejunostomy
- Now a successful artist and graphic designer
- Presented at age 20 mo with albinism, abdominal distention, apnea/cyanotic spells, seizures and hypoglycemia
- GER/feeding intolerance/constipation
- Persistent and relapsing episodes of pseudoobstruction
- Gastrostomy/Jejunostomy placement



At age 2 years: Family investigated for MSBP
(see letter)



Mother - [REDACTED] Child - [REDACTED]

April 22, 1987

Alex Flores, M.D.
319 Longwood Avenue
Boston, MA 02115

Dear Dr. Flores,

Enclosed you will find some articles on Munchausen Syndrome by Proxy that I thought would be of interest to you. I faced [REDACTED] with the fact that [REDACTED] was being put in an abusive position of having medical procedures which follow from her [REDACTED] description of various symptoms. It was presented from a position of her being "hypervigilant" of [REDACTED] to make sure that she was ok -- thereby amplifying what she saw into a "problem" which had to be addressed -- and medical tests done. We talked of the destructive sequelae both from a psychological and physical perspective. We also discussed that if these symptoms persisted, only observed by her, [REDACTED] might have to be removed from her home for observation. We also talked of how some mothers have such a need for their child to be sick that they might even go as far as to poison him/her or "doctor" the symptoms seen, i.e.; add their (the mom's) blood to the urine or feces. (I also added that once factitious illness is suspected, it can be verified). [REDACTED] absorbed what I was saying and just replied that she would never poison [REDACTED]

As of now, except for [REDACTED]'s vision, the G.I. problems appear to be the one area of validity which need to be observed. It would be extremely important to document what is real and what is not -- and what mother could possibly be doing to create the symptoms. Again, mom took in what I said and appears to have accepted my explanation (at this time) of her hypervigilance causing the evolution of symptoms. We seem to have a good alliance and she will be continuing to see me. I do not think that involving legal authorities or filing a 51A is indicated at this time. Please keep me abreast of your interaction with her.

Sincerely,

The One Who Wasn't (II)

Longitudinal follow-up

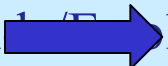
Age 10: Required TPN & Jejunal Feedings but poor enteral toleration

Developed: vasculitis, pancreatitis, sleep apnea, IgA nephropathy, cholelithiasis

Age 12: Abnl jejunal motility – non propagated MMC's after octreotide

Age 16: Abnl AD motility – No MMC's after EES

Age 16: Muscle biopsy for diagnosis of Mitochondrial Disorder (Complex III) Low decylubiquinol & succinate cytochrome c reductase

Patient has now been on TPN for 15 years → only two episodes of sepsis (Candida  li)

WE FINALLY HAD A DIAGNOSIS !!!

Date/Problems
(No. and Description)

FINDINGS (Subjective and Objective)

PLANS

20 mo

12/19/85

R#1 (Auto Abdominal distention)
→ Started last MARCH.

Hx:

- (A) F+up, BW: 7p⁵ (VERY distended
"mil c big belly" / C-Excess
6 1/2 p: 3 p: 0
X-rays in the nursery / ↑ clinical
New England Memorial

- (B) Episodes of Distention in the Nursery
6 days old → Very sleepy
Numbness // on breast feeding

- (C) 6 wks: Soft Mastitis (w/o exudate)

- (D) 4 mo: During Nursery / cyanotic
spell // seizure by A. Goldblatt
Unrelated to feeding / chronic atitis
strong family hx of sleep apnea

- (E) 9 1/2 mo: Seizures / sterility
spells (Molt - Neurology) EEG: C
seizure on phenobarbital 30mg/kg
Lp: ⊖ / CT SCAN: ⊖

- (F) 10 mo: Abdominal distention

13 mo =

OFF breast
milk

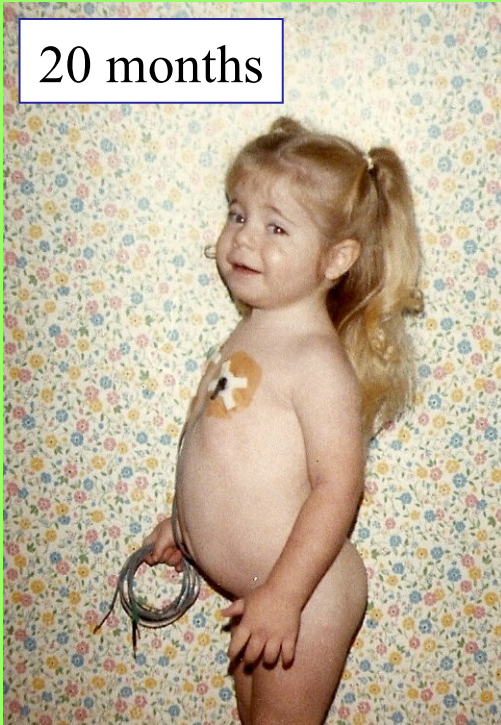
unable to wean.

DARK fatty stools (spray)

seizure in March 84 (convulsion / diminished)

- (G) Epilepsy c +4 Kellogg / 5 p: 5
after formula (low phenobarbital level)

20 months



Gotta love
school pictures



Look at my belly...



I'm ten years
old !!



18 years old



Management

Patients with presumptive diagnosis of Intestinal Failure and Question of MSBP →

2. Compulsive attention to minute detail verifying prior medical records, procedures and surgical interventions
3. Team approach including: Physicians, nursing, child protection team, social workers, psychiatry and psychology experts, nutritionist, physical therapy and legal team.
4. Hospital admission for evaluation
5. Separation test
6. Covert Video tape (?)

Conclusions

1. Intestinal Failure is relatively easy to diagnose and very difficult to treat.
2. When a patient has the presumptive Dx of CIPO/Dysmotility, we should be meticulous in documenting the manometric abnormalities and clinical course
3. Always listen to the parents and don't make a premature and irresponsible Dx of MSBP
4. Team Approach and experienced seasoned observers are essential in making Dx of MSBP in patients with this entity
5. Mandatory to have longitudinal follow-up in patients with MSBP/FF
6. Need of prospective and outcome studies in this extremely complicated group of patients

When we were young...



THE END

