

FPIES – CASE SERIES

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WHY THIS PRESENTATION ? CHRONIC DIARRHEA – KNOWN FACTS

Stool volume of more than 10g/kg/day that lasts for more than 14 days.

In India, major causes of chronic diarrhea are attributed to infections, which are designated as protracted/ persistent diarrhea.

Causes < 2 years

- Immune/ inflammatory
- Sepsis
- Structural defects – tufting enteropathy etc.,
- Malabsorption – pancreatic disorders
- Syndromic

USUAL WORKUP

Infections
Immune

- **CBC / CRP / Urine Routine / Blood – urine cultures**
- **Ig Profile, HIV, IgE levels, IgE cows milk & Soy milk antibody**

Malabsorption

- **Fecal chymotrypsin assays/ Enzyme assays**
- **Celiac screening**

- **Upper GI & proctoscopy, Biopsy, Electron microscopy**
- **X-ray abdomen / USG**

CASE HISTORY

- 2 ½ months, female infant
- Loose stools & vomiting for 4 weeks, watery associated with blood.
- Vomiting +
- Low grade fever +
- Weight loss & poor weight gain (Birth weight – 2.8kg, weight at admission – 2.9kg)

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- Exclusively breast fed for 10 days
 - Started on lactogen based feeds – developed loose stools. Seen outside and was started on ORS, vitamin supplements. Child was started on lactose free feeds. Child continued to have loose stools and was hence brought here.

ON EXAMINATION

- Marasmic
- Dehydrated
- No perianal rash

LABS

- CBC – 18000/8.7/3.4 L
- CRP – 76 mg/dl
- RFT – acidosis
- Albumin – 2.6 gm/dl

- Stool/urine - N
- Reducing substance +

INITIAL MANAGEMENT

- Fluid therapy as WHO SAM protocol
- Broad spectrum antibiotics pending cultures
- NPO & later Soy protein based feeds initiated
- Worsening diarrhea, recurrence of dehydration
- ? Nosocomial sepsis. Repeat counts & culture.

WORKUP OF THIS INFANT

Infections
Immune

- **Leucocytosis, positive CRP, negative cultures**
- **Normal Ig Profile, HIV - neg, IgE level - 64, IgE cows milk & Soy milk antibody – negative**

Malabsorption

- **Fecal chymotrypsin assays/ Enzyme assays – not done**
- **Celiac screening – not done**

- **Upper GI & proctoscopy, Biopsy – non specific duodenitis, proctitis, few eosinophils seen**

Leucocytosis
Positive CRP
Cultures sterile
Hypoalbuminemia

Not tolerating animal
protein & soy protein
based diet

-Young infant
initially breast fed.
started on artificial
feeds.
-CHRONIC
DIARRHEA
-FTT

IgE antibodies
negative

WHAT NEXT?

DOES THIS INFANT HAVE
"FPIES" ?



Food Protein Induced Enterocolitis Syndrome

- Non-IgE-mediated food allergic disorder
- Accounts for nearly 40% of milk protein allergy in infants and young children
- Under recognized

Food Protein Induced Enterocolitis Syndrome

- Usually manifests as profuse repetitive emesis & diarrhea that may be accompanied by lethargy
- Respiratory and skin manifestations are absent
- Usual triggers are cows milk protein and soy protein.
- Symptoms may start in the newborn period or up to one year of age. Later onset usually results from delayed introduction of triggers.

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- Family history of atopy is positive in 40% – 80 %.
 - Family history is positive for food allergy - 20%.
 - Approximately 30% of infants with FPIES develop atopic diseases such as eczema , asthma or rhinitis or drug hypersensitivity later in life.
 - Family history of FPIES is reported in 6%.

DIAGNOSIS

- Clinical features, exclusion of other etiologies
- OFC (oral food challenge test) – gold standard
- Atopy test – not recommended
- Hypoalbuminemia and weight gain less than 10g/day - independent predictors of FPIES severity.

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- Stool examination – nonspecific
 - Total IgE may be elevated.
 - Methemoglobinemia – can occur
 - Endoscopy : rectal ulceration and bleeding with friable mucosa

	INFANT 1	INFANT 2	INFANT 3
Age at presentation	3 months	2 months	2 ½ months
Nutritional h/o	Lactogen feeds	Buffalo milk/lactogen/soy milk diet	Buffalo milk/lactogen
Loose stools - duration	> 4 weeks	3 – 4 weeks	4 weeks
Freq per day	5- 10	5 – 10	> 10
Consistency	Watery ass with mucus	Mucoid and bloody	Watery
Emesis	+	–	+
Fever	Yes	Yes	No
FTT	+	+	+

	INFANT 1	INFANT 2	INFANT 3
CBC	24000/11.3/6.1L	19800/ 11/4.5 L	22000/9.3/3L
CRP	+	-	+
Hypoalbuminemia	+	-	+
RFT	Met acidosis	Met acidosis-	Met acidosis
Blood culture	No growth	No growth	No growth
Stool / urine routine	Few leukocytes	Normal	Normal
X ray	Prominent small bowel loops	Normal	Prominent -small bowel loops
USG	Normal	Normal	Normal

	INFANT 1	INFANT 2	INFANT 3
Immunoglobulin profile	Normal	Normal	Normal
Total IgE	46	38	32
IgE for cow's milk protein & soy protein	Negative	Negative	Negative
UGI scopy	-	Normal	Normal
LGI scopy	Patchy proctitis	Chronic deudenitis/ mod villus atrophy/ non specific proctitis eosinophils seen	Mild non – specific erosions Eosinophils seen Moderate villous blunting

MANAGEMENT OF OUR INFANTS

- Started on elemental aminoacid based formula
- Remarkable improvement
- Hypoallergenic formula was continued for few months (average of 4 – 6 months) and gradually changed over to soy protein based formula and later to normal diet.

HYPOALLERGENIC FORMULAS

PARTIALLY HYDROLYSED

COMPLETELY HYDROLYSED

ELEMENTAL AMINOACID BASED

AMINOACID BASED FORMULA

- Also known as “elemental” formulas.
- Used in children who do not tolerate extensively hydrolyzed formula

LESSONS LEARNT

- The features of FPIES mimics “SEPSIS” & can be misdiagnosed
- Good response to elemental aminoacid based formula. Though expensive, it has a major role in the management of such infants
- Chronic diarrhea, vomiting, failure to thrive in an infant – Allergic etiology should be considered than sepsis.

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- Proctocolitis
 - Food protein induced enteropathy
 - Eosinophilic gastroenteritis

 - **FPIES**



THANK YOU