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**Food protein-induced enterocolitis
syndrome (FPIES) acuta da
alimenti solidi multipli**

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ELYAS, 6 mesi



- Nato da I gravidanza gemellare
- Diabete Gestazionale materno in terapia dietetica
- EG 35 sett. + 3 gg
- PN 2.300 gr; LN 49 cm
- Accrescimento staturo-ponderale regolare



Allattamento esclusivo con Latte di tipo 1 (Formulat 1)



Da 2 giorni allattamento con Latte tipo 2 (Formulat 2)



Da 15 giorni assume omogenizzato di frutta misto (agrumi, banana, mela e pera)

6 mesi, Elyas inizia lo svezzamento:



- Latte scremato in polvere
- Frumento
- Orzo
- Avena
- Biscotto
- Burro
- Farina di semi di carruba
- Cacao magro
- Tracce di soia e lupino

Dopo 2 ore dal pasto

- Episodi ripetuti di vomito
- Iporeattività
- Pallore cutaneo





- GB 7.590/mm³
- GR 4.770.000/mm³, Hb 12.3 g/dl
- PLT 400.000/mm³
- PCR < limite
- Creatinina 0,28 mg/dl
- Elettroliti (mEq/L) Na 138, Cl 101, Ca 10.6

- Iporeattivo
- Cute pallida
- FA depressa
- Sudorazione algida
- Ripetuti episodi di vomito

- EGA: pH 7.25, pCO₂ 52 mmHg, pO₂ 30 mmHg, HCO₃⁻ 22.4 mmol/L, Lac 3.1 mmol/l, BE - 5.2

- Eco FAST: anse del tenue modicamente distese da contenuto liquido corpuscolato con peristalsi accentuata e lieve ispessimento parietale; piccola falda fluida peritoneale tra esse

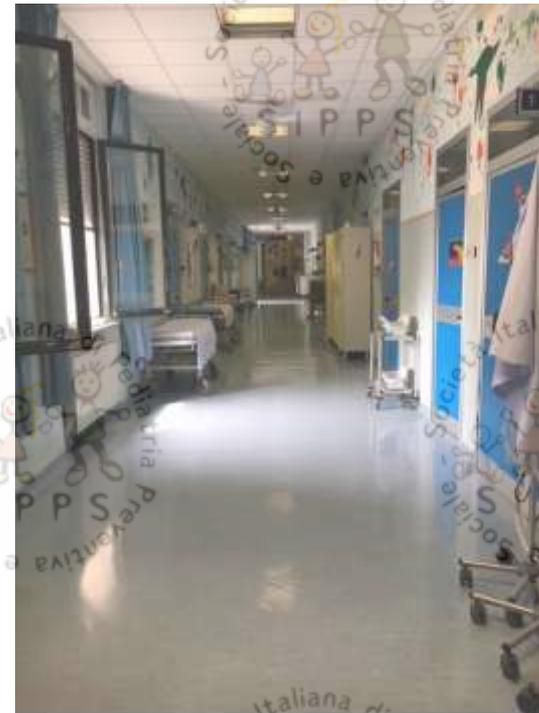


- Bolo di Soluzione Fisologica
- Ranitidina ev
- Metilprednisolone ev
- Ondansetron ev

Numerosi episodi di diarrea verdastra (7 ore dall'assunzione dell'alimento trigger) con risoluzione dopo 48 ore.

Dopo 24-48 ore il bambino riprende ad alimentarsi regolarmente solo con latte Formulaf 2 con ottima tolleranza.

Coprocoltura: negativa
IgE totali: 5 U/ml
IgE specifiche: albume, latte, grano, segale, orzo, avena, granoturco, riso integrale: negative



FPIES

Test di provocazione orale:

1. 1 cucchiaio di farina di Mais e Tapioca in 180 ml di latte Formulac 2
2. 5 ml di una preparazione ottenuta diluendo 1 cucchiaino da caffè di crema di riso in 120 ml di acqua

...dopo circa 3 ore..

- Ripetuti episodi di vomito
- Sopore
- Sudorazione

FPIES DA ALIMENTI SOLIDI MULTIPLI

Ambulatorio di Allergologia Pediatrica

OFCs: zucchine, carne di manzo, carote, patate e quinoa.





International consensus guidelines for the diagnosis and management of food protein-induced enterocolitis syndrome: Executive summary—Workgroup Report of the Adverse Reactions to Foods Committee, American Academy of Allergy, Asthma & Immunology

Definizione: Forma di allergia alimentare non IgE mediata

FPIES ACUTA

SINTOMI:

Dopo 1-4 ore dall'ingestione dell'alimento trigger (di solito 2 ore)

- Episodi ripetuti di vomito
- Pallore cutaneo
- Letargia

Dopo 5-10 ore dall'ingestione dell'alimento trigger

- Diarrea acquosa (raramente con muco e sangue)

FPIES CRONICA

SINTOMI:

- Vomito ricorrente
- Diarrea acquosa cronica
- Mancata crescita

Le forme più severe possono evolvere in:
Disidratazione

- Shock ipovolemico

L'eliminazione dell'alimento trigger determina miglioramento delle condizioni generali in 3-10 giorni. La reintroduzione può determinare una FPIES acuta

ALIMENTI TRIGGER

60-70% reagiscono a un singolo alimento (principalmente latte vaccino).

20-30% reagiscono a 2-3 alimenti.

10% reagiscono a più di 4 alimenti.

Latte vaccino/proteine della soia del latte formulato

- 40% reagisce a entrambi
- Insorgenza primi gg-12 mesi (3-5 mesi)

Cibi solidi

- Cereali (riso, avena, grano, orzo)
- Uova
- Frutta (mela, banana, pera, pesca)
- Verdure (patate, zucca)
- Carne (maiale, manzo, pollame)
- Legumi (soia, arachide, piselli)
- Pesce (crostacei, molluschi)
- Noccioline
- Insorgenza 4-7 mesi

	US	AU	UK	Israel	S. Korea	Spain	Italy
CM	Red	Red	Red	Red	Red	Red	Red
Soy	Red	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow
Rice	Red	Orange	Orange	Orange	Orange	Orange	Orange
Fish	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow	Yellow

Figure. Geographic differences in FPIES triggers. Red, most common trigger foods; orange, less common trigger foods; yellow: the least common trigger foods. FPIES indicates food protein-induced enterocolitis syndrome. AU, Australia.

FPIES è un disturbo autolimitantesi con una **storia naturale** favorevole. La tolleranza spontanea viene raggiunta più precocemente nei pazienti con FPIES per latte vaccino/soia rispetto ai paziente che reagiscono ai cibi solidi

Nowak-Wegrzyn A et al. *Food Protein-Induced Enterocolitis Syndrome. J Investig Allergol Clin Immunol* **2017**; Vol. 27(1): 1-18.

DIAGNOSI

TABLE IV. Diagnostic criteria for patients presenting with possible FPIES

Acute FPIES

Major criterion:

Vomiting in the 1- to 4-h period after ingestion of the suspect food and absence of classic IgE-mediated allergic skin or respiratory symptoms

Minor criteria:

1. A second (or more) episode of repetitive vomiting after eating the same suspect food
2. Repetitive vomiting episode 1-4 h after eating a different food
3. Extreme lethargy with any suspected reaction
4. Marked pallor with any suspected reaction
5. Need for emergency department visit with any suspected reaction
6. Need for intravenous fluid support with any suspected reaction
7. Diarrhea in 24 h (usually 5-10 h)
8. Hypotension
9. Hypothermia

The diagnosis of FPIES requires that a patient meets the major criterion and ≥ 3 minor criteria. If only a single episode has occurred, a diagnostic OFC should

CRITERIO MAGGIORE + 3 CRITERI MINORI

several-day time course of gastroenteritis. The patient should be asymptomatic and growing normally when the offending food is eliminated from the diet.

Chronic FPIES

Severe presentation: When the offending food is ingested on a regular basis (eg, infant formula); intermittent but progressive vomiting and diarrhea (occasionally with blood) develop, sometimes with dehydration and metabolic acidosis.

Milder presentation: Lower doses of the problem food (eg, solid foods or food allergens in breast milk) lead to intermittent vomiting and/or diarrhea, usually with poor weight gain/FTT but without dehydration or metabolic acidosis.

The most important criterion for chronic FPIES diagnosis is resolution of the symptoms within days after elimination of the offending food(s) and acute recurrence of symptoms when the food is reintroduced, onset of vomiting in 1-4 h, diarrhea in 24 h (usually 5-10 h). Without confirmatory challenge, the diagnosis of chronic FPIES remains presumptive.

Nowak-Wegrzyn et al. *International consensus guidelines for the diagnosis and management of food protein-induced enterocolitis syndrome: Executive summary—Workgroup Report of the Adverse Reactions to Foods Committee, American Academy of Allergy, Asthma & Immunology.* J Allergy Clin Immunol **2017**;139:1111-26.

TEST DI PROVOCAZIONE ORALE

1. Posizionare accesso venoso
2. Prelievo di base con emocromo
3. Somministrare 0.06-0.6 g (0.3 gr/Kg) in 3 dosi da somministrare ogni 30 minuti (la dose non deve essere superiore a 3 gr di proteine o 10 gr di cibo totale solido, 100 ml di liquido)
4. Osservare il paziente per 4-6 ore
5. Comparsa dei sintomi entro 1-4 ore, la diarrea entro 5-10 ore.
6. GB > 1500/mL dopo 4-6 ore
7. Esame chimico fisico delle feci: RSO, GB, GR

Infusione boli Soluzione fisiologica 20 mL/Kg
Ondansetron (0.1-0.15 mg/Kg max 16 mg)
Metilprednisolone 1 mg/Kg

TABLE V. Diagnostic criteria for the interpretation of OFCs in patients with a history of possible or confirmed FPIES

Major criterion	Minor criteria
Vomiting in the 1- to 4-h period after ingestion of the suspect food and the absence of classic IgE-mediated allergic skin or respiratory symptoms	<ol style="list-style-type: none">1. Lethargy2. Pallor3. Diarrhea 5-10 h after food ingestion4. Hypotension5. Hypothermia6. Increased neutrophil count of ≥ 1500 neutrophils above the baseline count

Criterio maggiore +
almeno 2 criteri minori

GESTIONE IN ACUTO

Infusione boli Soluzione fisiologica 10-20 mL/Kg
 Ondansetron 0.1-0.15 mg/Kg max 16 mg
 Metilprednisolone 1 mg/Kg (MAX 60-80 mg)

Medical Facility

Mild

Moderate

Severe

Symptoms

1-2 episodes of emesis
 No lethargy

>3 episodes of emesis and mild lethargy

>3 episodes of emesis, with severe lethargy,
 hypotonia, ashen or cyanotic appearance

Management

1. Attempt oral re hydration (eg, breastfeeding or clear fluids)
2. If age 6 months and older: Consider intramuscular ondansetron* (0.15 mg/kg/dose, maximum 16 mg/dose)
3. Monitor for resolution about 4-6 hours after the onset of a reaction

1. If age older than 6 months: administer intramuscular ondansetron* (0.15 mg/kg/dose, maximum 16 mg/dose)
2. Consider placing a peripheral intravenous line for normal saline bolus 20 mL/kg, repeat as needed
3. Transfer the patient to the emergency department or intensive care unit in case of persistent or severe hypotension, shock, extreme lethargy, or respiratory distress
4. Monitor vital signs
5. Monitor for resolution at least 4-6 hours after the onset of a reaction
6. Discharge home if patient is able to tolerate clear liquids

1. Place a peripheral intravenous line and administer normal saline bolus (20 mL/kg) rapidly, repeat as needed to correct hypotension
2. If age 6 months and older: administer intravenous ondansetron (0.15 mg/kg/dose, maximum 16 mg/dose)
3. If placement of intravenous line is delayed due to difficult access and age is 6 months or older administer ondansetron intramuscular 0.15 mg/kg/dose, maximum 16 mg/dose
4. Consider administering intravenous methylprednisolone 1 mg/kg, maximum 60 to 80 mg/dose
5. Monitor and correct acid base and electrolyte abnormalities
6. Correct methemoglobinemia if present
7. Monitor vital signs
8. Discharge after 4-6 hours from the onset of a reaction when the patient is back to baseline and is tolerating oral fluids
9. Transfer the patient to the emergency department or intensive care unit for further management in case of persistent or severe hypotension, shock, extreme lethargy, or respiratory distress

GESTIONE A LUNGO TERMINE

1. Dieta di evitamento

NB: Nei pazienti con **FPIES per latte vaccino/soia**:

- Prediligere allattamento al seno
- Utilizzare formule idrolisate
- Non utilizzare latte di capra e di pecora
- Si può utilizzare latte di asina e di cammello

2. Introduzione nuovi cibi: 20-30% reagisce a più cibi

E' consigliato eseguire l'introduzione di nuovi cibi in ambiente medico

Table 7. Common Food Co-allergies in Children With FPIES

FPIES to	Clinical Cross-reactivity/ Co-allergy	Observed Occurrence ^a
Cow milk	Soy	<30%-40%
	Any solid food	<16%
Soy	Cow milk	<30%-40%
	Any solid food	<16%
Solid food (any)	Another solid food	<44%
	Cow milk or soy	<25%
Legumes ^a	Soy	<80%
Grains (eg, rice, oats) ^a	Other grains (including rice)	About 50%
Poultry ^a	Other poultry	<40%

3. Challenge per valutare la

risoluzione Ovviamente sotto supervisione medica. Formale OFC. Viene di solito eseguito 12-24 mesi dopo la reazione più recente.

Svezzamento nel paziente con FPIES

Table 8. Empiric Guidelines for Selecting Weaning Foods in Infants With FPIES^a

Ages and Stages	Low-Risk Foods ^b	Moderate-Risk Foods ^b	High-Risk Foods ^b
<p>4-6 months (as per AAP CoN) If developmentally appropriate, safe and nutritious foods are available.</p> <p>Begin with smooth, thin purees and progress to thicker purees.</p> <ul style="list-style-type: none"> Choose foods that are high in iron. Add vegetables and fruits. 	<p>Broccoli, cauliflower, parsnip, turnip, pumpkin</p>	<p>Squash, carrot, white potato, green bean (legume)</p>	<p>Sweet potato, green pea (legume)</p>
	<p>Fruits</p> <p>Blueberries, strawberries, plum, watermelon, peach, avocado</p>	<p>Apple, pear, orange</p>	<p>Banana</p>
<p>6 months (as per WHO) Complementary feeding should begin no later than 6 months of age.</p> <ul style="list-style-type: none"> In the breastfed infant, high-iron foods or supplemental iron (1 mg/kg/d) is suggested by 6 months of age. Continue to expand variety of fruits, vegetables, legumes, grains, meats, and other foods as tolerated. 	<p>High-Iron Foods</p> <p>Lamb, fortified quinoa cereal, millet</p>	<p>Beef, fortified grits and corn cereal, wheat (whole wheat and fortified), fortified barley cereal</p>	<p>Fortified infant rice and oat cereals.</p>
<p>8 months of age or when developmentally appropriate Offer soft-cooked and bite-and-dissolve textures from around 8 months of age or as tolerated by the infant.</p> <p>12 months of age or when developmentally appropriate Offer modified tolerated foods from the family table—chopped meats, soft cooked vegetables, grains, and fruits.</p>	<p>Other</p> <p>Tree nuts and seed butters^b (sesame, sunflower, etc) Thinned with water or infant puree for appropriate texture and to prevent choking^b</p>	<p>Peanut, legumes other than green pea</p>	<p>Milk, soy, poultry, egg, fish</p>



Grazie per l'attenzione

