Your quick guide to:

Food Protein Induced Enterocolitis Syndrome (FPIES) - Information for GP's

What is FPIES?

Food protein induced enterocolitis syndrome (FPIES, pronounced F-pies) is a severe form of non-IgE mediated food allergy. As a non-IgE mediated allergy, FPIES cannot be detected using standard allergy tests (skin and blood tests for food specific IgE) and can present diagnostic uncertainty.

How does FPIES present?

FPIES most often presents in infancy, with acute reactions occurring following the introduction of cow's milk formula or solid foods. Reactions have the following features:

- Delayed onset, typically occurring 1-4 hours after ingestion of a trigger food or cow's milk formula.
- Profuse, repetitive vomiting (often large volume and projectile, may become bilious).
- Vomiting may be accompanied by systemic features including marked lethargy, floppiness and pallor.
- Not associated with rashes, swelling or respiratory symptoms.
- Diarrhoea may occur, usually around 5-10 hours after ingestion of the trigger.
- Blood tests may show non-specific changes in keeping with an acute phase response.

Some infants/children will recover from an acute FPIES reaction with oral rehydration at home, but more severe reactions may progress to hypotension and shock.

Milder reactions may be mistaken for viral gastroenteritis, whereas more severe reactions can mimic sepsis or abdominal surgical pathology.

What food triggers FPIES?

Triggers for FPIES include formula (cow's milk) and a large range of solid foods.

In the UK, the commonest trigger foods for FPIES are cow's milk, hen's egg and fish. Other foods not usually associated with allergy, such as oats, rice, banana, avocado, sweet potato and chicken, are also common triggers.

The majority of affected children have only one or two trigger foods, but around 5% of children have multiple (3 or more) trigger foods. Reactions do not always occur on the first ingestion of a trigger food, and it may take several ingestions before a reaction occurs.

Who to refer

All children with suspected FPIES should be referred to an allergist or paediatrician with an interest in allergy. A tool for finding specialist allergy services can be found here.

https://www.bsaci.org/public-information/find-a-clinic/

Diagnosis of FPIES

Diagnosis of FPIES is usually made by a paediatric allergist, specialist dietitian, general paediatrician or gastroenterologist with knowledge of the condition.

There are no diagnostic tests specific for FPIES, and diagnosis is primarily based on a clinical history of characteristic signs and symptoms, with improvement after withdrawal of the suspected trigger food. Diagnostic criteria for FPIES are given in International Consensus Guidelines for FPIES, which can be found here:

https://www.jacionline.org/article/S0091-6749(17)30153-7/fulltext#tbl4

Oral food challenges may be carried out in hospital in cases of diagnostic uncertainty, but this is avoided where possible due to the severity of reactions.

Some children with FPIES may have other non-IgE- mediated allergies and/or IgE-mediated allergies, which can make diagnosis more complex.

Key facts:

FPIES most often presents in infancy, with acute reactions occurring following the **introduction of cow's milk formula or solid foods**.

All children with suspected FPIES should be **referred to an allergist or paediatrician** with an interest in allergy.

There is **no role for antihistamine**s or intramuscular adrenaline in the management of acute FPIES reactions.

Allergy UK Helpline

Mon-Fri, 9am-5pm:
Call: 01322 619 898
Email: info@allergyuk.org

Visit us at:

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If you have any comments about this Factsheet, contact Allergy UK on info@allergyuk.org. The guidance in this Factsheet is based on current best practice and may be subject to change in the light of new relevant information.

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Management of acute FPIES reactions

Once a diagnosis of FPIES is made, families are usually supplied with a letter to present at the Emergency Department should an acute reaction occur, explaining that the child has a diagnosis of FPIES and outlining appropriate treatment for suspected reactions.

Vomiting in acute reactions usually responds to treatment with ondansetron.

If the infant/child is deteriorating despite attempts to rehydrate them orally with clear fluids or breast milk, treatment with intravenous fluids and other supportive care may be required in hospital.

There is no role for antihistamines or intramuscular adrenaline in the management of acute FPIES reactions (although these may be prescribed if there are concurrent IgE-mediated allergies).

On-going management of FPIES

- Strict avoidance of known trigger foods is recommended, and should result in resolution of FPIES symptoms.
- Breastfed babies should continue to be breastfed. No
- maternal elimination diet should be recommended
- unless the symptoms are clearly related to exposure
- to breastmilk.
- Babies who present with FPIES on cow's milk formula,
- will need a specialist formula. In the majority of
- cases an extensively hydrolysed formula will be well
- tolerated, but some infants may continue to react and
- require an amino acid formula.
- Babies/children with FPIES are at increased risk of
- nutritional deficiencies and faltering growth, and
- should have access to a specialist dietitian in order to:
  - Create an individualised complementary feeding plan that meets the infant's nutritional needs for optimal growth and development whilst eliminating the offending allergen(s).
  - Ensure normal growth and development.
  - Advise/support in avoidance of known/suspected allergens.
- Feeding difficulties and food aversions are more
- common in babies/children with FPIES, particularly
- those with multiple (3 or more) trigger foods.
- Caring for a child with FPIES has been found to have
- a significant psychosocial impact on parents/carers,
- with increased anxiety and stress, and they may
- require psychological support.
- Babies/children with FPIES are more likely to have
- other atopic conditions, such as eczema, wheeze and
- IgE-mediated food allergies.
- Children with FPIES usually remain under the care of
- a paediatric allergy team until resolution.

Prognosis/Reintroduction of foods in FPIES

- The prognosis for children with FPIES is very good
- and the majority outgrow it by around 5 years of age.
- Reintroduction of trigger foods should be carried out
- under supervision of a specialist allergy team as an
- oral food challenge in hospital (rather than using a
- food ladder-based approach at home).
- The timing of reintroductions varies and is usually first
- considered around the age of 2-3 years old.

With thanks to FPIES UK for kindly donating the content and resources that contributed to this factsheet. Their support has helped us provide accurate information and guidance for those affected by FPIES.

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