Food protein-induced enterocolitis syndrome (FPIES)

Food protein-induced enterocolitis syndrome (FPIES) is an adverse food reaction involving the immune system. It is uncommon and seen almost exclusively in infants and young children. It is caused by an allergic reaction to one or more ingested foods which results in inflammation of the small and large intestine. Symptoms of profuse vomiting (and sometimes diarrhoea) most commonly occur 2-4 hours after eating a food which has been recently introduced into the diet. Some children may become pale, floppy, have reduced body temperature and/or blood pressure during a reaction. Avoidance of the trigger food protein/s is currently the only effective treatment option, however most children will outgrow their FPIES in the preschool years.

FPIES is different to many common food allergies because:

- It is usually a delayed reaction, and is not caused by Immunoglobulin E (IgE) antibodies.
- Reactions only involve the gastrointestinal system (no hives, welts or swelling are seen on the face or body).
- It is not associated with anaphylaxis.
- Adrenaline autoinjectors are NOT used to treat the reaction.

It is possible for a child with FPIES to also have IgE mediated allergies to other foods and to have other allergic diseases such as eczema and asthma.

Which foods can trigger FPIES?
The most common FPIES triggers are rice, cow's milk (dairy) and soy. However, almost any food can cause an FPIES reaction, including cereals such as rice, oats, eggs, legumes and meats such as chicken and seafood. FPIES rarely occurs in exclusively breastfed infants.

Is it possible to have FPIES to more than one food?
Some children have FPIES to more than one food protein. For example, some children reactive to cow's milk have been noted to react to soy and some reactive to rice have also reacted to oats. Children with FPIES to chicken may react to other poultry such as turkey.

Symptoms of FPIES
A typical FPIES reaction begins with profuse vomiting around 2 to 4 hours after ingesting the trigger food/s, often followed by diarrhoea which can last for several days. Occasionally a shorter time frame may be seen. In the most severe FPIES reactions, vomiting and diarrhoea can cause serious dehydration. Children with FPIES can have poor growth if they continue to ingest trigger food/s.

How is FPIES Diagnosed?
There are no laboratory or skin tests which can confirm a diagnosis of FPIES. This can make FPIES difficult to diagnose.

- Some children during an FPIES reaction have an elevated white cell and platelet count, and may be mistaken for having an infection.
- Allergy skin prick tests or allergy blood tests to the food protein/s are not helpful in making the diagnosis.
- Medically supervised oral food challenges can be useful when the history is not clear or when other foods from similar food groups are being introduced into the diet for the first time. They are also useful to establish when the child has outgrown FPIES.

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How is FPIES treated?
Currently the only specific treatment option for FPIES is avoidance of the trigger food. Infants who have reacted to cow’s milk and soy formulas will usually be trialled on extensively hydrolysed formula (eHF) or amino acid based formula (AAF) if eHF is not tolerated.

Treatment during an FPIES reaction may include intravenous (IV) fluids, because of the risk of dehydration. Children experiencing more severe symptoms may also require corticosteroids and in-hospital monitoring. Most families of children with FPIES will be given a letter to present to emergency departments explaining their child’s condition and the appropriate treatment.

There is no role for the use of adrenaline autoinjectors in the management of FPIES.

Does FPIES resolve?
Most children outgrow FPIES by about three to four years of age. However, this varies between individuals and foods. However, only 40-80% of those with FPIES to rice, and 60% to dairy tolerated these foods by three to four. The best way to determine whether a child has outgrown their FPIES is a medically supervised food challenge.

References

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