

About Food Protein-Induced Enterocolitis Syndrome

Written in collaboration by: The FPIES Foundation Board of Directors and Medical Advisory Board

Food Protein-Induced Enterocolitis Syndrome (FPIES) is a type of food allergy affecting the gastrointestinal (GI) tract. Classic symptoms of FPIES include profound vomiting, diarrhea, and dehydration. These symptoms can lead to severe lethargy, change in body temperature and blood pressure. Unlike typical food allergies, symptoms may not be immediate and do not show up on standard allergy tests. Furthermore, the negative allergy evaluation may delay the diagnosis and take the focus off the causative food. Nonetheless, FPIES can present with severe symptoms following ingestion of a food trigger.

FPIES Common Symptoms:

There are two ways that infants or children with FPIES might come to medical attention.

1. The classic pattern of an FPIES reaction is when a healthy infant or child develops symptoms shortly after eating a food. There is a characteristic delay of 2-3 hours before onset of severe and repetitive vomiting and eventually diarrhea. The child may appear very ill and sleepy (lethargic), and may become pale or blue. When evaluated by a doctor, he/she may be found to have low blood pressure, seem dehydrated, and have blood tests that mimic infection (sepsis); which in some cases can lead to sepsis-like shock. Many infants who are eventually diagnosed with FPIES are initially suspected to have a severe infection or sepsis.
2. The second common pattern of FPIES reaction symptoms occurs when infants who are ingesting a problem food (usually milk or soy-based formula or proteins in breast milk) as a consistent part of their diet might experience increasingly severe vomiting, diarrhea, and poor growth, possibly progressing to an illness mimicking a severe total-body infection. Please note that each child is unique and your child may experience their own range and intensity of these symptoms.

FPIES Common Triggers:

In the first months of life, FPIES reactions are most often caused by cow's milk protein formula, and sometimes by soy. Proteins in breast milk may also cause symptoms in some infants.

For infants experiencing FPIES with solid foods, rice and oats are the most common triggers. Current research reports other common triggers that include, but are not limited to, milk, soy, barley, poultry, peas, green beans, sweet potatoes, and squash. However, any food protein can be a trigger and some infants may be sensitive to other foods as well. In addition, some children may react to one or two foods whereas others may experience reactions to multiple foods.

FPIES Diagnosis and Testing:

FPIES is a non-IgE food allergy, which unlike classic food allergy, cannot be diagnosed with readily available food allergy tests such as skin prick test (SPT) or blood test that measure food IgE antibodies (RAST). These tests are helpful to identify triggers for typical food allergies that result in immediate hives, wheezing, and swelling and are characteristically *negative* in FPIES. An FPIES diagnosis is usually made by considering the history of the characteristic symptoms and exclusion of various alternative illnesses. A medical doctor, often an allergist and/or gastroenterologist, should be involved in making the diagnosis. Although Atopy Patch testing (APT) is often used for FPIES patients, it is not considered a validated test for FPIES diagnosis. Blood tests performed during a reaction may be helpful since the results often mimic the body's response to infection. The most definitive test is a medically supervised oral food challenge (OFC)-where the suspect food is given to the child in a controlled clinical environment. An oral food challenge, however, is not often needed initially if the doctor has excluded alternative illnesses and the medical history is consistent with FPIES.

FPIES Treatment and Course:

FPIES reactions can be severe. It is important to get to prompt medical attention where treatment, such as fluids given into the vein to help stabilize blood pressure and treat dehydration, can be given in order to avoid sepsis-like shock. Although some doctors prescribe epinephrine to stabilize blood pressure before medical treatment, the main therapy is to get intravenous fluids; also steroids can be used to quell the immune reaction. Preparing a letter for potential trips to the ER, containing both FPIES information and a list of your child's triggers, may be helpful.

Unfortunately, there are currently no simple tests for FPIES. The primary test, as mentioned above, is a medically supervised oral food challenge with the trigger food. The good news is that FPIES usually resolves with time. Your child will need to be closely followed by his/her doctor to discuss what foods are safe to eat and when it may be time to determine if FPIES has resolved. With proper medical attention and a personalized dietary plan to ensure proper nutrition, children with FPIES can grow and thrive.

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Additional Useful Resources:

1. Caubet, J.; Nowak-Wegrzyn, A. Current Understanding of the Immune Mechanisms of FPIES. Expert Review. *Clinical Immunology*. 2011; 7(3), 317-327
2. Jyonouchi, H. Non-IgE Food Allergy. Inflammation & Allergy. *Drug Target*. 2008; 7(3): 1-7.
3. Nowak-Wegrzyn, A.; Sampson, H.A.; Wood, R.A.; Sicherer, S.H. Food Protein-Induced Enterocolitis Syndrome Caused by Solid Food Proteins. *PEDIATRICS April 2003; 111 (4): 829-835*.
4. Sampson HA, Anderson JA. Summary and recommendations: classification of gastrointestinal manifestations due to immunologic reactions to foods in infants and young children. *J. Pediatr. Gastroenterol. Nutr.* 30(Suppl.), S87-S94 (2000).
5. Giovanna M., et al. Food Protein-Induced Enterocolitis Syndrome by cow's milk proteins passed through breast milk. *The Journal of Allergy and Clinical Immunology*. March 2011; 127 (3): 679-80.