The FPIES Foundation
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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 individuals with FPIES might come to medical attention.

1. The classic pattern of an FPIES reaction is when a healthy individual 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. There are reports in the medical literature of reactions that occur at a delay of >3 hours post-ingestion as well. The individual 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 individuals 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 individuals who are ingesting a problem food as a consistent part of their diet (i.e. added milk to a daily morning coffee) might experience increasingly severe vomiting and diarrhea, possibly progressing to an illness mimicking a severe total-body infection. Please note that each individual is unique and 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 and children 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 individuals may be sensitive to other foods as well. In addition, some individuals may react to one or two foods whereas others may experience reactions to multiple foods.

For adults experiencing FPIES symptoms, the most common triggers reported in the current medical literature include types of shellfish. That said, it is possible for any food protein to cause an FPIES reaction in affected individuals. Reports of FPIES triggers for adults in the medical literature include fish, vegetables, and other foods. Though adults affected by FPIES typically react to only one food, it is reported that some individuals experience FPIES 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) may be 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 individual in a controlled clinical environment. An oral food challenge, however, is not often needed 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; steroids can also be used to quell the immune reaction. Preparing a letter for potential trips to the ER, containing both FPIES information and a list of the individual's triggers, may be helpful.

There is not much information available regarding adults affected by FPIES, and as a result, the medical team needs to be prepared to advocate for affected individuals. Additionally, instructing the patient in the use of effective self-advocacy skills is essential to disease management. FPIES is not a common diagnosis in adults; therefore, awareness among emergency medical professionals and general practitioners may be lacking.

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. With proper medical attention and a personalized dietary plan to ensure proper nutrition, individuals with FPIES can maintain a healthy quality of life and thrive.

This document describes FPIES in Adults. FPIES in infants and children is addressed in the original About FPIES. Modifications for information regarding adults affected by FPIES was made by FPIES Foundation leadership and advisors, 2021.

About FPIES ©2011-2021 is a written collaboration of The FPIES Foundation Board of Directors and The FPIES Foundation's Founding Medical Advisory Board: Sakina S. Bajowala, MD; J. Andrew Bird, MD; April Clark, RD/LD; John J. Lee, MD; Fred Leickly, MD, MPH; David R. Naimi, DO; Harumi Jyonouchi, MD; Scott H. Sicherer, MD; Anna Nowak-Wegrzyn, MD.

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