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Food Protein Induced Enterocolitis Syndrome Triggers at Children's Hospital

Rachel Nelson

Russell Hopp

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INTRODUCTION

Food protein-induced enterocolitis syndrome (FPIES) is a non-lgEmediated food disease characterized by repetitive vomiting 1-4 hours after ingesting a trigger food. The vomiting is often followed by watery and sometimes bloody diarrhea, which in severe cases can progress to lethargy, pallor, hypothermia, and hypotension.¹ Diagnosis is made by a history of delayed reaction to a specific food and clinical improvement after eliminating the trigger food from the diet.² Acute episodes are treated with antiemetics, intravenous hydration, and occasionally corticosteroids.¹

The clinical phenotype, including trigger food and disease course, varies by region.³ In a diverse cohort from the International FPIES Association representing patients from Australia, Austria, Brazil, Egypt, Germany, Italy, and the United States, cow's milk was the most commonly avoided food followed by soy, oats, rice, and egg.¹ Shellfish is a rare trigger in children, but accounts for 82% of adultonset FPIES.⁴ Rarely, FPIES patients are found to be IgE positive for their trigger food, a condition termed Atypical FPIES. The most common trigger food for Atypical FPIES is egg.⁴ It is also possible to have FPIES with a concomitant food allergy, with patients most often allergic to peanut and egg.⁴

We sought to determine survey the most common FPIES foods seen at Children's hospital in the past five years. A data search of the EPIC electronic medical record using FPIES codes ICD-10 (K52.21) and ICD-9 (558.3) as the primary diagnosis revealed 40 cases and 69 trigger foods (Figure 1).

We also present two unusual FPIES cases recently diagnosed at Children's Hospital. One case with two less common food triggers, and a case of FPIES with concomitant IgE positivity to the food (atypical FPIES).

Food Protein Induced Enterocolitis Syndrome Triggers at Children's Hospital Rachel Nelson; Russell Hopp, DO Creighton University School of Medicine, Omaha, NE;

Children's Hospital & Medical Center, Omaha, NE

CASE 1

A 4-year-old male with a history of chronic nasal congestion presented to clinic for evaluation of vomiting. He had three separate episodes of vomiting 2 hours after eating. Two episodes occurred after eating shrimp, and one occurred after eating salmon. Each time, the vomiting self-resolved, and was not associated with fatigue, rash, shortness of breath, diarrhea, or lethargy. He was previously able to eat shrimp and fish without issue. Exam was grossly normal. Allergy skin testing was negative for fish, shellfish, and environmental allergies. A sinus x-ray showed left maxillary sinusitis. He was prescribed Augmentin for the sinus infection and given a diagnosis of FPIES with instructions to avoid all fish and seafood and follow up in 2 years for oral food challenge.

CASE 2

A 20-month-old male with a history of cardiomyopathy awaiting heart transplant presented with 2 separate episodes of vomiting after eating eggs. Allergy blood test showed > 100 kUA/L of IgE to egg. Skin testing was done for eggs and peanuts as he does not like eating peanut butter at home. Skin prick testing was markedly positive to peanuts, but not tree nuts, and was not done for egg given the high blood IgE level. He was diagnosed with atypical FPIES to egg plus allergy to peanut and egg. Due to his heart condition and high titer IgE, if he does vomit, he should be given an EpiPen and taken to the emergency room. He will return in a year for a repeat egg allergy blood test.

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Insights from 441 Children with FPIES as Provided by Caregivers in the International

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In our Children's' hospital cohort from the last 5 years, the most common FPIES triggers were oat, rice, sweet potato, milk, and egg. Sweet potato FPIES is more common in Omaha than in previously studied populations.^{1,4} Fish FPIES appears to have a higher prevalence in Omaha than other regions,^{1,4} as 4 out of 40 FPIES in our review had a fish trigger.

FPIES can be a tricky diagnosis when patients present with an atypical trigger or reaction, but it is a condition that can significantly affect quality of life. FPIES can result in food aversion, poor body weight gain, and decreased school attendance.^{1,4} While many patients fully outgrow FPIES, fish and egg are the most common trigger foods for persistent FPIES.⁴ Understanding these complications and variations can help us advise our patients on their expected disease course.

This report serves to highlight the common FPIES triggers in our region and illustrate the variability in FPIES presentation and course of disease.



Figure 1. FPIES Triggers in the last 5 years at Omaha Children's Hospital. $N_{patients} = 40$; $N_{foods} = 69$.

DISCUSSION

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