

Food Protein Induced Enterocolitis Syndrome (FPIES)**Background**

FPIES is a paediatric non-immunoglobulin E mediated allergic disorder triggered by ingestion of certain food protein(s). The majority of children present with their first FPIES episode when less than 12 months of age. The diagnosis of FPIES remains a clinical one, with children presenting typically 2-4 hours after ingestion of a food protein recently introduced into the diet, with profuse vomiting and some subsequently develop diarrhea^{1,2}. Some children present in a moribund state, with pallor, floppiness, reduced body temperature, hypovolemic shock and/or metabolic acidosis¹⁻³.

Delays in diagnosis are frequent, since common FPIES food triggers such as rice are considered to be hypoallergenic, and presenting clinical features can mimic more common paediatric conditions, such as sepsis and surgical abdominal emergencies^{2,3}. Consequently children often have multiple episodes, additional investigations and prolonged hospital admissions before a diagnosis of FPIES is made²⁻⁴. The prognosis for FPIES is good and most attain tolerance to the food trigger by three years of age². Early and accurate diagnosis is important to enable appropriate dietary advice and prevention of recurrences, emergency management plans in case of future reactions and a food challenge to be arranged, typically at 3-4 years of age, to ascertain tolerance.

The prevalence of FPIES in Australian children is unclear. To date the majority of studies published have examined cases presenting to individual tertiary Paediatric hospitals or specialised Allergy outpatient clinics and have been retrospective in design²⁻⁵.

The prospective collection of information on FPIES reactions in the paediatric setting will enable the estimation of the incidence of FPIES, and facilitate a better understanding of the demographic and clinical features, causative foods and common associated food sensitivities, and current management practices of clinicians caring for children with FPIES. Such a study would also draw the attention of the paediatric and broader community to this allergic condition, enabling the development of educational material for clinicians and parents, and formulation of clinical practice guidelines.

Study Objectives

- Estimate the incidence of FPIES in Australian children
- Describe the demographic features of children with FPIES
- Determine the causative food trigger(s) and common co-sensitisations
- Describe clinical and laboratory features of FPIES reactions
- Describe the current diagnostic and management practices of children presenting with FPIES.

Case Definition and reporting instructions

Please report any child ≤ 24 months of age whom you have seen within the last month and have not previously reported to the APSU, meeting the following case definition criteria:

1. Repeated episodes of vomiting typically presenting within 4 hours after ingestion of a food protein that has been recently introduced into the diet without any other cause identified for the episode of vomiting **AND**
2. Absence of fever (>38°C), any cutaneous (urticaria, hives, and/or angioedema) and respiratory clinical features (difficulty in breathing/talking, swelling of tongue, tight throat/hoarse voice, wheeze and/or persistent cough) at the time of reaction **AND**
3. Removal of the offending food protein from the diet resulted in resolution of symptoms

Follow-up instructions

A questionnaire requesting de-identified details about the patient and aspects of the diagnosis will be sent to clinicians who notify a case of FPIES via email, post or may be downloaded from the APSU website (www.apsu.org.au)

Please return the completed questionnaire to the APSU by post or fax as instructed on the questionnaire.

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References

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