

~50% of epilepsies have some genetic basis^{1,2,3} Find the underlying cause of the seizure



Real patient with CLN2 disease

- **Some rapidly progressive neurodegenerative disorders present with subtle and non-specific symptoms**, mimicking more common paediatric epilepsies.³
- **CLN2 disease (TPP1 deficiency) is one devastating genetic condition** that often presents with new-onset unprovoked seizures between 2-4 years of age and/or language delay and/or motor disturbance/ataxia.^{4,5}
- **Genetic testing can help identify the aetiology** of 100+ paediatric epilepsies with one test*²
- **Many genes are actionable:** A prompt diagnosis can be essential to adopt precise disease management strategies.^{3,2}
- If you have patients with seizures of unknown cause, **no-cost genetic testing may be available** in your country.³

Visit paediatricseizures.com to find a no-cost test sponsored by BioMarin near you.

The list of laboratories provided does not include all laboratories that perform CLN2 genetic and enzymatic diagnostic testing. Laboratories providing the no-cost tests are supported by BioMarin and participate in commercially-sponsored diagnostic testing programs. There may or may not be a sponsored program or test in your area. The sponsors receive de-identified patient data, but no patient identifiable information. Participants of this program have no obligation to recommend, purchase, order, prescribe, administer, use or support any product or service of the sponsors.

* Number of genes tested will vary in each country, ranging from a single gene to over 400+ genes

~ The genetic basis of epilepsy has a range according to country and publication from 14% -24.4%,² to 50%¹, to 70-80%³

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for with 49 children with spinal muscular atrophy, patients at various stages of the disease (in terms of clinical subtype and degree of disease progression). For nine patients, drug therapy was never begun or was suspended after a careful evaluation of the actual benefits and the burden of the treatment.

Today, we are increasingly faced with cogent clinical and bio-ethical dilemmas that impose a profound reflection on the appropriateness of medical interventions that should respect the best interest and the quality of life of the child living with serious illness.³

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Dear Editor,

ARE PACKAGING ERRORS THE REAL CAUSE FOR FOOD RECALLS AND ALLERGIC REACTIONS IN AUSTRALIA?

Reports suggest that unintended food allergen presence in processed foods caused by allergen cross-contact (i.e. food comes into contact with another food) may be the leading cause of contamination resulting in adverse reactions.

The aim of this study was to examine factors which contribute to undeclared allergens being present in processed foods in Australia, by examining food recalls.

Food recall data compiled by Food Standards Australia New Zealand (FSANZ) between 1 January 2009 and 31 December 2018 were collected and analysed. Poisson regression was used to examine trends in food recalls over the study period. Consent was not required for this type of study.

Over the period 2009–2018, FSANZ conducted 675 food recalls. Undeclared allergens ($n = 266$, 39%), microbial contamination

The root cause of all undeclared allergens food recalls from 2016 to 2018 $n=92^{\dagger}$

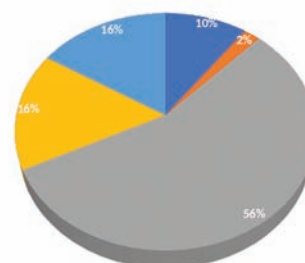


Fig. 1 The cause for the recall from 2016 to 2018. (■), Accidental cross contact ($n = 9$); (■), lack of skills and knowledge ($n = 2$); (■), packaging errors ($n = 51$); (■), supply verification issues ($n = 15$); (■), unknown ($n = 9$) (Due to food business not being able to determine the root cause or because FSANZ did not receive any information from the business following the recall). [†]In mid-2016, FSANZ added another question to determine the leading cause of undeclared allergen recalls, since this was added later in the year, a full year of data is unavailable.

($n = 176$, 26%) and foreign matter ($n = 108$, 16%) accounted for the majority of recalls. In mid-2016, FSANZ added another question to determine the cause of undeclared allergen recalls. For the period 2016–2018, packaging errors where the food product was either packed into incorrect packaging or was incorrectly labelled ($n = 51$, 56%), supplier verification issues ($n = 15$, 16%) and cross-contact ($n = 9$, 10%) were the most commonly identified factors leading to recalls due to undeclared allergen¹ (Fig. 1).

Our current results suggest that in Australia, packaging errors (i.e. product packed in the wrong packaging or incorrect labelling) are a leading cause of food recalls due to presence of undeclared food allergens, not allergen cross-contact. This finding may also explain the relatively high prevalence of consumer reported anaphylaxis to process food, as we have previously reported.^{2,3} Similarly in a 2002 study, Vierk *et al.* reported that over a single year in the USA, 34 consumers had experienced allergic reactions after consumption of recalled products.⁴

Undeclared allergens in packaged foods was the most common reason for food recalls in Australia, with packaging errors the most commonly identified factor contributing to undeclared allergen recall. It is likely that many other cases of mislabelling, unintended food allergen contamination occurred and were not detected or reported. If all packaged food products could have been assessed and analysed for labelling errors and food allergen cross-contact, it is probable the numbers would be much higher. There is a need for improved packaging practices to minimise foods with undeclared allergens and increase food safety for food allergic consumers.

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Conflict of interest: None declared.

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Dear Editor,

EARLY CHALLENGES IN ISOLATION AND DE-ISOLATION OF CHILDREN DURING THE SEVERE ACUTE RESPIRATORY SYNDROME CORONAVIRUS-2 PANDEMIC

Children frequently have viral acute respiratory illness (ARI) that is clinically indistinguishable from severe acute respiratory syndrome coronavirus-2 (SARS-CoV-2) infection. Appropriate investigation and disposition of children suspected of COVID-19 is important to utilise negative pressure rooms efficiently.¹ A de-isolation algorithm for suspect adult cases has been described.²

In our tertiary paediatric unit, from 22 January to 22 April 2020, we isolated 305 inpatients with ARI in a negative-pressure or neutral-pressure single-bed room depending on ARI severity, epidemiological or contact risk factors and aerosol-generating procedures. Two nasopharyngeal swabs ≥24 h apart were tested for SARS-CoV-2 polymerase chain reaction.

We summarise our isolation challenges early on in the COVID-19 pandemic.

- 1 Active modification of clinical admission workflows for an evolving national case definition, while maintaining consistency with national and institutional guidance, and recognising that children are not little adults.
- 2 Some patients downplayed their symptoms and travel/contact history at admission, which were revealed only later during observation. Subsequent isolation of the child generated staff and patient anxiety regarding potential exposure. Exploring

the patient's perception of COVID-19 helped us obtain an accurate history which could otherwise be concealed to avoid testing and isolation.

- 3 Children with underlying comorbidities or frequent hospital visits were concerned for their safety and requested use of an isolation room when not required. Clear accurate information was provided to allay anxiety, manage expectations and ensure compliance to infection control practices.
- 4 Asymptomatic children required isolation throughout their incubation period if they or their caregivers were under quarantine after close COVID-19 exposure, to avoid asymptomatic or pre-symptomatic transmission.
- 5 De-isolation of suspect cases with ongoing ARI symptoms was not straightforward. Polymerase chain reaction can be negative early in illness.² Difficulty in obtaining a nasopharyngeal swab in young children affects test sensitivity. Identification of another causative pathogen makes COVID-19 less likely but cannot exclude it due to possible co-infection; SARS-CoV-2 has been reported with dengue and influenza viruses.^{2,3} COVID-19 respiratory deterioration usually happens at about 1 week⁴; observation for symptom resolution before de-isolation may prolong hospital admission unnecessarily, as children may present early in illness, and rhinorrhoea and cough often persist for 2–3 weeks. Efficient inpatient management enabled quick, safe discharge to strict home isolation until recovery. Where bed occupancy allowed, we continued to isolate children with ongoing ARI needing unrelated inpatient care, under droplet instead of airborne precautions.

Optimal use of scarce isolation rooms will require rapid diagnosis and better understanding of SARS-CoV-2 transmission.

Conflict of interest

None declared.

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