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NOVEL PRIMARY IMMUNODEFICIENCY?: CASES OF INTERFERON-ALPHA/BETA RECEPTOR 2 DEFICIENCYKE Madueme, A Dumitrescu. *Paediatrics, Cork University Hospital, Cork, Ireland*

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Aims There is very limited understanding of the role of Type 1 Interferon (IFN) in human anti-viral immunity. However, IFNs are known to contribute to the activation of innate and adaptive immune responses. Human Interferon-alpha/beta receptor 2 (IFNAR2) deficiency causes fatal susceptibility to live viral vaccines, revealing a vital but narrow nonredundant role for IFN-alpha/beta in viral immunity.¹ We describe 2 cases of IFNAR2 deficiency. Sanger sequencing identified homozygous single nucleotide deletion c.A311del in IFNAR2 gene in both cases.

Methods The cases are siblings; the older one having died of fatal encephalitis before the younger one was born. Clinical and laboratory data were reviewed.

Results **Case 1:** A 14-month-old previously healthy boy presented acutely unwell with a preceding history of MMR vaccination. He deteriorated rapidly and subsequently died from haemophagocytic lymphohistiocytosis complicating vaccine-strain mumps and rubella encephalitis. Post-mortem systemic and brain samples demonstrated clear evidence of sustained replication of vaccine viruses and HHV-6. In retrospect, he had no problems with common viral infections during infancy. **Case 2:** Younger sister of the first case was diagnosed IFNAR2 deficient at about 6 weeks of age. She had uneventful neonatal and infancy periods. She has been on prophylactic aciclovir since post neonatal period. Subcutaneous immunoglobulin was commenced after her third childhood vaccine (while avoiding all live vaccines). She was admitted at 9 months of age for respiratory tract infection which completely resolved. Parents are heterozygous for this mutation. Wider family members were screened; all were homozygous wild-type at this position implying neither affected nor carriers of the disease.

Conclusion These are the first documented cases of IFNAR2 deficiency worldwide. Perhaps, this rather novel primary immunodeficiency will broaden the knowledge of the role of IFN-alpha/beta in human antiviral immunity.

REFERENCE

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G57(P)

EVALUATING A COW'S MILK PROTEIN ALLERGY MANAGEMENT PROTOCOL FOR CHILDREN PRESENTING TO THE EMERGENCY DEPARTMENT

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Introduction In spite of clear guidance for the diagnosis of cow's milk protein allergy (CMPA) there is often a significant delay in its recognition and management. This is on a background of an increasing number of emergency department (ED) visits for food induced allergy and anaphylaxis with one study reporting a more than doubling in the number of hospital visits between 2001–2006.

Given the potentially large number of infants and children presenting with undiagnosed CMPA we present the results of an evaluation of an emergency department CMPA management protocol.

Methods The study site was a large district general hospital with an audio-visually distinct children's ED. A diary system is used to allocate patient's follow-up ED 'hot clinic' appointments that are deemed necessary. The 2015 hot clinic diary was reviewed and children with a provisional diagnosis of CMPA following their acute ED attendance were identified for inclusion. The Emergency Department Information System was then interrogated and data collected in to a pre-designed study pro forma. The electronic clinical records for each patient were also consulted for the outcome of the hot clinic appointment. Data were analysed using Microsoft Excel for Mac 2011.

Results 99 patients were identified (1 January 2015 to 31 December 2015). 23 patients were excluded (11 did not have an original diagnosis of cow's milk protein allergy whilst 12 were return patients following their initial hot clinic appointment).

82% (62) had the initial diagnosis of CMPA from the acute ED presentation confirmed by a Consultant at hot clinic review. 10% (8) were found to have an alternative diagnosis following a Consultant review: secondary lactose intolerance, possetting, reflux, diarrhoea, and poor weight gain. 8% (6) failed to attend their planned follow-up of which 4 (5%) were subsequently diagnosed with CMPA.

Conclusions Our ED protocol for the management of infants with suspected CMPA is a simple algorithm that allows for the rapid management and follow-up of such children. It provides continuity of care without the need for referral to paediatric outpatient or allergy services that are already overburdened. We would advocate the initiation of such a protocol in other children's emergency department's in order to facilitate the smooth management of these children.

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ARE CHILDREN AT RISK OF SEVERE INFLUENZA RECEIVING SEASONAL INFLUENZA VACCINE? AN AUDIT OF OUTPATIENT CLINICS AT ALDER HEY CHILDREN'S HOSPITAL

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Aim To assess the uptake of seasonal influenza vaccine for at-risk children attending Alder Hey Children's Hospital Liverpool outpatient clinics, and their household family members, during the 2016/7 influenza season.

Methods An electronic questionnaire survey, for at-risk children and their household family members, was conducted in the waiting rooms of a variety of speciality outpatient clinics during a two-week winter period from 30th January 2017 until 10th February 2017. Clinics were chosen based on the Department of Health (England, UK) definition of patient at-risk groups. Patients were asked about their influenza vaccination status alongside the vaccination status of the household family members. Data was analysed on IBM SPSS Statistics Version 24.

Results 216 survey responses were analysed from which 153 at-risk patients were identified. Mean age of all children