**G56** Novel Primary Immunodeficiency?: Cases of Interferon-alpha/beta Receptor 2 Deficiency

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10.1136/archdischild-2018-rcpch.54

**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 and laboratory data were reviewed.

**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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**G58(P)** 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 2016/17 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

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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.

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**Abstracts**

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