G56

NOVEL PRIMARY IMMUNODEFICIENCY?: CASES OF INTERFERON-ALPHA/BETA RECEPTOR 2 DEFICIENCY

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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.

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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.

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 2016/7 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

surveyed was 8.3 years (range 0–3 months to 18 years). Numbers of at-risk patients by speciality were as follows: respiratory 33, oncology 28, neurology 34, rheumatology 12, diabetes 24, nephrology 19 and immunology 3. 83 (54.2%) of at-risk children had received influenza vaccination in the past and 62 children (40.5%) had received this season's influenza vaccine. 88.7% of these children had received their influenza vaccination in general practice, 9.7% in school, and 1.6% at Alder Hey. Out of the 62 at-risk children vaccinated against influenza this season, 20 (32.3%) lived in a household in which all family members had been vaccinated. The most common reason given for children not receiving influenza vaccination was the lack of discussion over the vaccine with parents by health care professionals, in both primary and secondary care settings.

Conclusions The poor uptake of influenza vaccine for at-risk children attending Alder Hey reflects the global issue of inadequate influenza vaccination for such at-risk children. This brief survey provides further evidence that the importance of seasonal influenza vaccination is not being conveyed to the parents of at-risk children. Hospital specialists need to do more to assess the influenza vaccination status of their at-risk children. To facilitate this service improvement, the implementation of a hospital-based 'Immunisation Co-ordinator' role, working across both primary and secondary care providers, should be evaluated.

G59(P) AN ABSCESS ON THE CHEST WALL – DON'T FORGET THE GREAT MASQUERADER

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Introduction Musculoskeletal tuberculosis (TB) occurs in 1%–3% of patients with the disease. However cold abscess of the chest wall is rare and constitutes 1% to 5% of all cases of musculoskeletal TB. Moreover it is only described in cases of severe or disseminated form of the disease and not as the first presenting feature. Patients are often young and have no comorbidity. Diagnosis is often delayed because of the atypical presentation and lack of awareness. We describe a case of anterior chest wall mass as the first presenting feature of tuberculosis.

Case history A 15 year old Bangladeshi girl presented to us with a 2 month history of progressively increasing mass on the left anterior chest wall. She had a history of cough and intermittent fever over the preceding 2 weeks. There were no other systemic symptoms. Her immunisations were up-to-date and had received BCG at birth.

On examination she had a 6 cm × 4 cm mass overlying the left 8th and 9th ribs, associated with an enlarged left supraclavicular lymph node. A chest x-ray showed the mass eroding into the underlying 8th rib and a left hilar nodal shadow with a small left pleural effusion.

The patient underwent a diagnostic biopsy to exclude malignancy. This revealed a fluid filled centre and a significant amount of pus was drained. A post-biopsy chest X-ray showed a markedly enlarged pleural effusion. The patient's QuantiFERON test was positive (16.05 iu/ml). The fluid was sent for culture and sensitivity and this confirmed drug sensitive *Mycobacterium tuberculosis*. The patient was treated with

standard Anti-Tubercular Therapy and had an uneventful recovery.

Conclusion Tuberculosis is a great masquerader and atypical presentations are known to occur. TB should be considered in children presenting with a chest wall mass even if they have been vaccinated with BCG. Other differential diagnoses include benign and malignant tumours. Prompt diagnosis and treatment of chest wall TB results in complete recovery and helps in preventing serious complications.

G60(P) SMALL GROUP PAEDIATRIC DIETETIC SESSIONS FOR COW'S MILK PROTEIN ALLERGY (CMPA): ONE YEAR FOLLOW UP SURVEY

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Background/aim Small group dietetic sessions were held for parents and carers of patients with non-IgE mediated Cows Milk Protein Allergy (CMPA) to meet increasing demands. Initial feedback had been extremely positive with high participation and satisfaction rates. Participants found the group sessions informative, useful, and felt more confident in managing CMPA. We contacted the participants to obtain follow up feedback a year from the initial session, to evaluate the sustained effectiveness and confidence in parents managing their child's CMPA.

Method Parents and carers who attended the group dietetic sessions held between November 2015 and July 2016 were included. Feedback was obtained via a self-designed questionnaire using a Likert-type scale, rating several questions from 1 (least satisfied) to 5 (most satisfied), similar to the design of the initial questionnaire. We contacted the participants initially via telephone, and subsequently postal questionnaire with pre-paid and labelled envelopes.

Results Of the 40 participants, we successfully obtained feedback from 13 participants. Majority of the participants agreed that the group sessions have been informative (mean score 4.8), felt confident in weaning their child on milk-free diet (mean score 4.8), and also reintroduction of cow's milk in diet (mean score 4.3).

30% (n=4) preferred a one-to-one session. 38% (n=5) have contacted the dietitians via telephone after the initial session, and of these, 2 had requested further individual consultations. 69% (n=9) have attempted reintroduction of cow's milk in their child's diet using our local milk reintroduction guide. The mean age at first challenge was 14 months (age range 6 to 26 months), with average of two attempts. 31% (n=4) of the children have been successfully challenged and are managing well on a normal diet.

Conclusion We recognised the limitation in obtaining feedback via telephone and postal questionnaire, which resulted in the poor response rate. Overall, responses were positive and sustained a year on. Parents felt they had enough information and is confident in managing their child's CMPA. We have suggested emphasis on availability of follow up telephone or face-to-face consultations with the dietitians to allay parental anxiety. We will also pilot 'milk reintroduction' sessions to support and improve parental confidence in challenging their child with cow's milk at an appropriate age.