G450(P) HYPERIMMUNOGLOBULINE E SYNDROME CASE PRESENTATION

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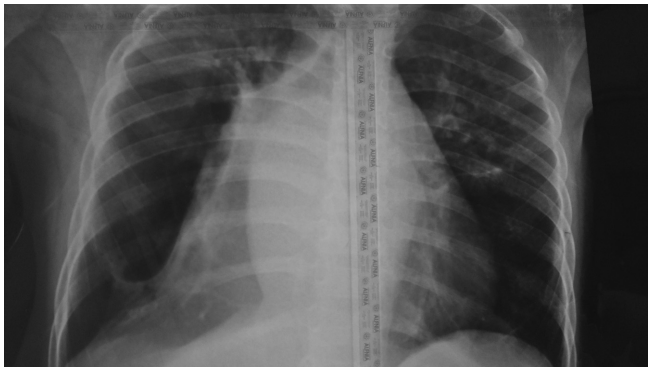
Background Hyperimmunoglobuline E syndrome (HIES), also known as Job Syndrome, is a rare primary immuneodeficiency disorder characterised by recurrent eczema, skin abscesses, lung infections, eosinophilia and high serum level of IgE. Two forms of HIES have been described, type 1(Autosomal Dominant) and type 2 (Autosomal Recessive) which is more severe. These two forms share overlapping clinical and laboratory features. However, they also exhibit distinct clinical manifestations, courses and outcomes. This paper will present our experience with this rare disease in our new Immunology unit.
Methods Six cases were diagnosed between end 2008 to early 2014, depending on clinical features, laboratory and radiological evaluation.
Results Two cases of HIES presented before their second birthday and the other four before their fifth. Two cases were brothers. Male to female ratio was 2:1.

Each case initially presented with different chronic infections. All have coarse features, eczema, high IgE (>2500 iu/ml), high eosinophil count and recurrent oral thrush. Five of the six cases have onychomycosis. Three cases had extensive varicella eruption. Infections were treated according to the international guidelines. Chest x–ray showed lung cysts in two cases, three cases had no lung changes and one patient refused x–ray and any follow up. Echocardiogram was normal in five cases. Serum Immunoglobulins, T and B lymphocyte subset were normal for four cases. The other two cases, one of them showed low T helper cell and low CD4 percentage with normal total count, and the other case showed low natural killer cell CD56 percentage and high count CD19, CD4, CD8, CD3 with normal percentage. All cases were put on oral prophylactic Trimethoprim and regular follow up except one who refused (Figure 1).
Conclusion The rising awareness among junior doctors and early referral of cases with symptoms to suggest primary immunodeficiency disorders help to diagnose and treat cases of HIES (Job Syndrome) which used to be missed and arrived to hospital in serious or terminal state. The incidence rate of Job syndrome internationally is rare. Further studies are needed to estimate the incidence rate and prevalence in different areas of our country.



Abstract G450(P) Figure 1

G451(P) IMPROVING SCHOOL ATTENDANCE AT A SPECIAL NEEDS SCHOOL – A SERVICE EVALUATION

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Introduction Pupils with learning disabilities find it more challenging to learn than their peers. Due to their complex needs, they also have a greater rate of school absenteeism (1). Attendance is vital for learning new skills (target attendance >95%) (2). We looked at medical reasons for school absenteeism at a special needs school in Camden.
Aims To identify medical reasons for school absenteeism during winter months, with a view to introducing targeted interventions to improve attendance.
Methods Retrospective data collection of absentees from school register between September 2013 to March 2014 and clinical case notes review.
Results Out of 229 primary and secondary pupils attending, we included the ones with attendance <85%. 17% (n = 38) of the pupils had an attendance of <85%. The main diagnoses were varying severity of learning disabilities (41% n = 14), epilepsy (26% n = 9), autism (21% n = 7) and cerebral palsy (12% n = 4). Adolescents (aged 14–19) formed the bulk of the group affected (35%). Illness (unknown 47%, sleep 15%, seizures 9%, others 29%) were the most prevalent reasons (67% n = 19) followed by social factors (21% n = 6) and medical appointments (12% n = 4). No major respiratory illnesses or safeguarding concerns were highlighted in any of the cases. All, except 4 children had named consultants. 73% had medical reviews in date and had their medical needs identified and treated.
Conclusion Illness (type uncertain) was the main reason for absenteeism. Sleep difficulties and poorly controlled seizures were noted frequently. Unclear coding made further evaluation difficult.
Recommendations School attendance should be made an integral part of school medical reviews.

Absenteeism data should be coded clearly to enable identification medical reasons for absenteeism. School should receive notification from local Accident and Emergency departments when a special needs pupil attends. Targeted interventions to improve attendance can then be implemented.

G452(P) NOT JUST A PAIN IN THE NECK? AN UNUSUAL CASE OF AN INFANT PRESENTING WITH A STIFF NECK, DIAGNOSED WITH SEPTIC ARTHRITIS OF THE ATLANTOAXIAL JOINT AND OSTEOMYELITIS OF THE ODONTOID PEG

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Primary acute osteomyelitis of the first and second vertebrae is extremely rare in the paediatric population, and there is no literature documenting septic arthritis of the atlantoaxial joint secondary to odontoid osteomyelitis. We present the case of a 10–month–old, well child presenting with a stiff neck, with no clinical features suggestive of infection.

Blood tests revealed a mildly elevated white cell count and CRP, with a normal ESR. Notably his alkaline phosphatase was