Abstracts

A RETROSPECTIVE REVIEW OF DEATHS IN TRISOMY 21: A TERTIARY CENTRE’S EXPERIENCE SINCE THE MILLENNIUM

P Shires, E Marder, H Vyas. Nottingham Children’s Hospital, Nottingham University Hospitals NHS Trust, Nottingham, UK

Aims Children with Trisomy 21 often have complex health needs and are at increased risk of mortality than age-matched peers. A retrospective review was undertaken of children with Trisomy 21 who had died at our tertiary centre since the millennium. We audited demographics, cause of death and preceding events to identify any themes.

Methods A retrospective review of electronic, paper and archived microfilm patient records was undertaken in those with a diagnosis of Trisomy 21 who died in our trust after a diagnosis of Trisomy 21 who died at our tertiary centre since the millennium. Aims were to establish any recurrent or unusual causes of death, and to assess whether these deaths could have been prevented.

Results 16 cases were identified; the mean age at death was 34 months (ranging 2 days – 15 years). 50% of deaths occurred within the first year of life. Of the 13 cases where a cause of death was identified, cardiac pathology was attributed in 2 of 13 cases. Infection was implicated in 9 out of 13 cases, with 7 cases of primarily respiratory illness and 2 cases of line sepsis. Underlying respiratory disease was a significant contributing factor in 4 out of 13 cases. There was 1 case of Trisomy 21 with co-existent lethal skeletal dysplasia and 1 death related to congenital airway abnormality. In the 6 cases where immune function was tested, only one had completely normal function.

Conclusions The burden of cardio-respiratory disease in Trisomy 21 is well recognised. In our experience, sepsis, particularly with respiratory focus, was responsible for a high proportion of deaths. It is important that health care professionals have an awareness of the increased susceptibility and risk of mortality related to sepsis in children with Trisomy 21.

D Huggard, M Mahon, F McGrane, N Lagan, C Purcell, J Balle, E Roche, E Molloy, Academic Paediatrics, Trinity College Dublin, National Children’s Hospital, Tallaght, Dublin, Ireland

IMMUNODEFICIENCY IN CHILDREN WITH DOWN SYNDROME

Aims Down Syndrome (DS) is the most common genetic syndrome associated with abnormal immune function and immune defects. There is an increased susceptibility to both bacterial and viral infections. We aimed to examine the degree of immunodeficiencies in children with DS.

Methods Children who attended the specialist multidisciplinary DS clinic in Tallaght were included, and medical details collected especially in relation to infections, recurrent respiratory tract infections (RTIs), hospital admissions and vaccinations. Results of Full blood counts, T and B cell subsets and immunoglobulins were analysed and compared to age specific reference ranges.

Results Twenty-eight children (age range 1–12 years) were included and 16/28 (57%) had recurrent RTIs. Hospitalisation at least once was necessary in 15/28 (54%) patients, and 6/28 (21%) required multiple admissions. All but one patient’s routine immunisations were up to date (96%). Although 22 children had a normal white cell count (WCC), Neutrophil and lymphocyte levels, T and B cell subsets (n=13) revealed decreased CD3+, Helper T, Cytotoxic T and CD19 +B cells, with the latter being significantly reduced. IgA and IgG levels were normal or high in all cases, and levels were either normal or low for IgM.

Conclusion We found that children with DS were at increased risk of infections, especially recurrent RTIs, with a significant hospitalisation rate. Vaccination compliance was very high, however the CD19 +B cells were found to be low, which may point to a poor memory B cell response. Further research to evaluate individualised vaccination and prophylactic programmes would be valuable in this cohort.