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

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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 the millennium.

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. The recognised association with impaired immune functioning in Trisomy 21, coupled with underlying cardio-respiratory co-morbidities, may heighten susceptibility to mortality in sepsis.

To exacerbate this, communication and behavioural difficulties can make assessment more challenging and may mask or impede recognition of the severity of illness. Therefore, it is important for clinicians to be mindful of sepsis and have a low threshold for initiating early and aggressive management including timely administration of antibiotics.

IMMUNODEFICIENCY IN CHILDREN WITH DOWN SYNDROME

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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.
Case Report

G90(P) SINUSITIS: JUST ANOTHER CAUSE OF HEADACHE OR AN INNOCUOUS KILLER?

A 13-year-old boy presented to A and E with left sided weakness, altered consciousness and right gaze deviation. His parents found him in on the morning of admission confused and aphasic. Over the preceding ten days, he had complained only of a headache with pain over the right eye but no other neurological symptoms. He had presented to the GP once prior to admission and was diagnosed with possible migraine. On examination, he was aphasic, confused with a GCS 11/15 and had left sided weakness. On initial presentation, he was afibrile with otherwise normal observations.

Investigations He had an urgent CT, which was reported as normal followed by an urgent MRI. MRI revealed right fronto-temporal subdural empyema with pneumatocele and evidence of maxillary sinusitis. Blood investigations revealed a raised white cell count (20) with a neutrophilia (18) and raised CRP (217).

Treatment/outcome The patient received IV antibiotics, 3% saline and dexamethasone. Shortly after this, he was transferred to a neurosurgical unit and had an emergency right frontal craniotomy and drainage. Subsequently he required regular maxillary sinus washouts for four days. Bacterial culture revealed *Streptococcus milleri*. He received 6 weeks of IV antibiotics and has made a full neurological recovery. Repeat MRI reveals resolution of empyema and he has been discharged from secondary care.

Conclusions This case was chosen to highlight to paediatrician and primary care givers the potential serious sequelae of untreated sinusitis. Systematic review reports the morbidity rate as 27% and mortality rate was 3.3%.1 Early aggressive medical, ENT and neurosurgical intervention has proven to improve neurological outcome.2 Patients should also be evaluated from an immunological perspective, as serious sinogenic infections can be a sign of immunodeficiency.

REFERENCES


G90(P) I’M SORRY, BUT WE HAVE JUST GIVEN YOUR CHILD THE WRONG MEDICINE

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Aims To evaluate families views on how they would like to be informed about medication errors.

Methods We devised a parental questionnaire asking if their child used medication and if they had experienced an error. Three scenarios for medication errors with; no/low/significant harm were presented along with 4 options as to how to be told:

1. Tell them over the phone immediately
2. If we can’t contact them, leave an answer phone message that there is a problem
3. Tell them face to face when they return
4. Wouldn’t want to be informed

Their preference for who would inform them was sought. Parents of any children attending/resident in hospital were eligible.

Results We asked 100 parents: 85 mothers and 15 fathers; 52 inpatients and 48 outpatients. 53% of the children were taking regular medications and 7% had experienced a medication error.

<table>
<thead>
<tr>
<th>Scenario</th>
<th>No harm</th>
<th>Low harm</th>
<th>Significant harm</th>
</tr>
</thead>
<tbody>
<tr>
<td>Phone</td>
<td>39%</td>
<td>69%</td>
<td>82%</td>
</tr>
<tr>
<td>Leave answer-phone message</td>
<td>6%</td>
<td>6%</td>
<td>8%</td>
</tr>
<tr>
<td>Face to face</td>
<td>49%</td>
<td>25%</td>
<td>10%</td>
</tr>
<tr>
<td>Not informed</td>
<td>6%</td>
<td>0</td>
<td>0</td>
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Parents preferred to be told by: a doctor 54%, nurse 10%, either doctor or nurse 32% and 4% did not mind. Several specified that this would depend on the severity of the error and who was most available.

Conclusions Most parents would like to be informed about a medication error by a doctor as soon as it is identified. If they are not present they would prefer to be contacted by telephone, although not via an answering machine message. Where no harm is expected, waiting until parents are present was generally acceptable to them (table 1). Trust between families and healthcare professionals is encouraged by honesty and openness. Best practice should be to provide information as soon as possible; however this may present logistical challenges to meet these expectations.

G91(P) PRESCRIBING IN OVERWEIGHT AND OBESE CHILDREN

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Aims Childhood obesity is a growing global health burden. Prescribing medications for this cohort has until now relied on empirical experience with no set guidelines to inform drug dose calculations. Standard body weight calculated dosages have led to supratherapeutic doses commonly being prescribed. Recent research has highlighted the need for a new body weight adjusted approach to prescribing. This project aimed to evaluate whether these recent developments are being