We have found HLH in cases like enteric fever (3), other blood culture proved sepsis(2), pneumonia(1), primary immunodeficiency related(1), pyrexia of unknown origin(1), sJIA(2), acute pancreatitis(1), Infectious mononucleosis(1), congenital cytomegalovirus (CMV) infection associated(1).

Fever and hepatosplenomegaly were present in all the cases & other findings like rash (5), respiratory distress (4), lymphadenopathy (3), jaundice (1), bleeding (2) & seizures (2) were also present.

Laboratory features:

Anaemia (four needed blood transfusion) & thrombocytopenia and hyperferritinemia were present in all cases. Neutropenia present in 4 cases but others also showed gradual fall in total counts from leukocytosis towards normal range with clinical deterioration.

Other lab features like low fibrinogen level (10), high triglyceride (9), raised liver enzymes (12), hyponatremia (7) were also present. Fall in ESR present in 6 cases including the sJIA cases.

Bone marrow showed evidence of hemophagocytosis in seven cases.

Treatment:

All patients received supportive treatment including the treatment of cause.

Total 10 cases were infection related of which 4 were treated with IVIG and 3 were treated with steroid and rest 3 responded with treatment of the cause only.

One case was primary immunodeficiency (PID) related [leukocyte adhesion defect] which was treated with IVIG.

All of the sJIA (2) cases received steroid for macrophage activation syndrome and one was treated with cyclosporine also

Outcome:

Most of the cases (8) recovered with specific treatment (gamma/steroid) but three cases recovered with supportive treatment only. Unfortunately two children died - the case of immunodeficiency died of liver failure & the case of CMV infection died of profuse bleeding due to altered coagulation profile not responding to treatment with fresh frozen plasma and both of them died within one week of diagnosis.

Conclusions

- HLH not that uncommon in hospitalized children.
- A mind of suspicion for HLH - In a case of non remitting fever despite the appropriate treatment, Deterioration of clinical condition despite the appropriate treatment.
- Timely diagnosis & intervention can save lives.

Young People's Health Special Interest Group

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CHALLENGES TO THE TRANSITION OF YOUNG PEOPLE WITH COMPLEX HEALTH CONDITIONS TO ADULT SERVICES

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Background Transition to adult services is a critical time in a young person's life. NICE guidelines include five quality statements, which are; planning transition, an annual meeting, a named worker, introduction to adults' services and missed first appointments after transfer. Paediatrics as a speciality is becoming increasingly complex, with significant advances being made in medical care, such as gene therapy for spinal muscular atrophy, developments in neonatal resuscitation and non-invasive ventilation strategies being frequently used for respiratory failure. As a consequence, children are now surviving into adulthood with conditions that would have been previously fatal, many of which are rare and equally complex to manage, involving input from numerous members of the multi-disciplinary team (MDT). For this reason, the transition period for young people with complex conditions is challenging, not only for them but also for their families and needs to be appropriately managed at a time of vulnerability.

Objectives To compare the transition practice in a tertiary children's hospital using the NICE guideline for transition to adult services, NG43.

Methods Data was collected using a SurveyMonkey questionnaire, with twelve questions sent to all consultants in the Trust. Qualitative data was collected from four different specialities via focused interviews.

Results Of approximately 400 consultants, 68 responded over a wide range of specialities: 60.3% have a speciality specific transition pathway, 5.9% have a CNS designated to transition, 50% have a dedicated transition clinic (61.7% are held at the paediatric centre, 23% are held at both paediatric and adult centres and 14.7% are held at the adult centre). One service runs an out of hours transition clinic. 37% of transition discussions begin between the ages of 12 and 15 years. 26.5% start the process after 16 years. Barriers to the management of transition were clinic capacity and administration time, lack of a dedicated co-ordinator, and lack of an equivalent adult service to refer to. 38% of respondents reported negative consequences after transition, such as patient disengagement, anxiety and lack of input from other health professionals.

Interviews were carried out with 4 specialities: Cardiology, Epilepsy, Renal and ENT. Cardiology has the most clearly defined pathway, with a dedicated transition consultant and CNS. Starting at 12 years of age, there are 3 transition appointments with the majority of young people transferred to the designated the adult service by 16 years. The Renal and Epilepsy teams aim for a similar model, however begin transition discussions later and transfer by 18 years. ENT has a specific pathway only for young people with cochlear implants, but no transition CNS. The greatest challenge for all specialities was time available in the appointments and subsequent loss to follow up after transfer.

Conclusions There is considerable variation in the transition process across specialities within a tertiary children's hospital managing complex conditions in childhood. Variability in meeting NICE guidelines is evident. Key concerns are having an identifiable adult service to transfer to, patient disengagement and patient anxiety. A dedicated team for the speciality suggests improvement in successful transition.