G369(P) THE DIAGNOSTIC UTILITY OF BONE MARROW ASPIRATE IN PAEDIATRIC LIVER DISEASE

¹R Akhtar, ²E Fitzpatrick, ²D Hadic, ³S Chakravorty. ¹Department of Haematology, Imperial College London, London, UK; ²Department of Paediatric Hepatology, King's College Hospital, London, UK; ³Department of Paediatric Haematology, King's College Hospital, London, UK

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Aim To identify the usefulness of bone marrow aspirate (BMA) in the diagnosis of paediatric liver disease in order to prioritise its use in the diagnostic algorithm.

Methods A single-centre 17 year retrospective analysis of BMA procedures in paediatric liver patients was undertaken. Clinical and laboratory data were obtained from the Hospital Electronic Patient Records. Sensitivity and specificity of BMA as a diagnostic tool for Lysosomal storage disorders (LSD) and Haemophagocytic Lymphohistiocytosis (HLH) were computed.

Results A total of 184 paediatric liver patients undergoing 218 BMAs were identified. Of these, BMAs in 137 patients (74.5%) did not have any morphological features suggestive of a specific diagnosis, such as LSD or other haematological conditions (i.e. non-diagnostic). In 84% of children with a final diagnosis of idiopathic liver failure and in 87% with metabolic or developmental liver disease, no specific BMA features were identified. In 116 (63%) patients BMA was specifically undertaken to exclude LSD. A final diagnosis of LSD was made in 14 patients. Thirteen (92.9%) of these patients had morphological features on BMA supporting a diagnosis of LSD. In this case, BMA had an overall sensitivity of 75% (95% CI 42.8%–94.51%) and specificity of 92.55% (95% CI 85.26%–96.95%) in diagnosing LSD. A further nine patients received a diagnosis of HLH. Three (33.3%) of the HLH patients had a diagnostic marrow with a sensitivity of 37.50% (95% CI 8.52%–75.51%) but a specificity of 88.24% (95% CI 63.56%–98.54%).

Conclusion BMA is a safe, quick and effective test to undertake in critically ill children with liver disease, where urgent decisions regarding liver transplantation (LT) need to be taken. It is very useful in the diagnostic algorithm of suspected lysosomal storage disorders, particularly in Niemann-Pick C, where nearly all cases have specific morphological findings and where it requires a significantly longer time to obtain definitive molecular diagnoses. Thus, a morphologically non-diagnostic BMA in children with idiopathic liver failure, metabolic or developmental liver disease allows the clinical team to urgently proceed to LT. Diagnostic yield of BMA in HLH was poor highlighting the diagnostic challenges faced by clinicians in this condition.

G370(P) RISK OF INVASIVE PNEUMOCOCCAL DISEASE IN CHILDREN WITH SICKLE CELL DISEASE IN THE ERA OF CONJUGATE VACCINES: A SYSTEMATIC REVIEW OF THE LITERATURE

¹G Oligbu, ²L Pay, ²M Fallaha, ³O Oligbu, ⁴U Nwaomu. ¹Paediatric Infectious Diseases Research Group, St George's University of London, UK; ²Department of Paediatrics, Imperial College London, UK; ³Accident and Emergency, Queen Elizabeth Hospital, Woolwich, London, UK; ⁴Department of Paediatrics, Derby Royal Hospital, Derby, UK

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Introduction Invasive pneumococcal disease (IPD) is the leading cause of morbidity and mortality in children with sickle

cell disease (SCD). 7-valent pneumococcal conjugate vaccine (PCV7) was first introduced in 2000 in US and at various times in other countries. It was replaced by 13-valent pneumococcal conjugate vaccines (PCV13) in 2010. This PCV is highly effective in preventing IPD in children with SCD. The risk of IPD has not been systematically assessed in children with SCD since the introduction of PCV.

Methods We undertook a systematic review of the English literature published from 2000 to October 2017 to evaluate the risk factors, serotype distribution, clinical presentation and outcomes of IPD in children with sickle cell disease. Data sources included MEDLINE, EMBASE Cochrane library, and references within identified articles.

Results We identified 475 potential studies, of which 66 were duplicates. A further 299 were excluded on the basis of title and abstracts and another 94 studies did not meet eligibility criteria on full-text screening. We included 16 publications involving 11 383 children less than 21 years-olds with SCD. A total of 161 (1.4%) IPD was identified: 147 homozygote for haemoglobin S (HbSS), 10 double heterozygote for haemoglobin S and C (HbSC) and 4 others. Among the nine studies reporting clinical presentation, septicaemia was the commonest (n=69/108; 63.8%) followed by pneumonia (n=20/108; 18.5%). The serotypes associated with IPD in SCD were mainly non-PCV13 (n=117/133; 88%), of which more than a quarter was due to serogroup 15A/B/C (n=34/117; 29%). Serotype 23 F (n=6/16, 37.5%) and 7 F (n=4/16; 25%) were the main vaccine serotypes. Majority were sensitive to penicillin and ceftriaxone. The crude case fatality rate was 11.2% (n=18/161).

Conclusions and clinical implications This report demonstrates the effectiveness of conjugate vaccines to reduce the rate of IPD in children with SCD. However, these children remain at increased risk and are also more likely to die of their infection compared to their peers without SCD. Most IPD cases are now due to serotypes not covered by PCV13. Therefore better prevention strategies are needed to reduce the overall burden IPD in children with SCD.

Quality Improvement

G371 UPLOAD AT HOME: A STEP TOWARDS AUTONOMY FOR PATIENTS WITH DIABETES

S Goyal, C Bidder. Department of Paediatrics, Abertawe Bro Morgannwg University Health Board, Swansea, UK

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Background Analysis of patient data on glucometers and pumps is required to make clinical decisions. These devices require to be uploaded onto a computer so that data can be reviewed in a systematic way. We observed difficulties in uploading glucometers and pumps in outpatient settings. We identified contributing barriers related to the technology, staff and parents/patients. Uploading devices occupied valuable time of clinicians and specialist nurses, leaving less time to spend with patients and families.

Aim To encourage uploading at home prior to attending the outpatient appointment, which will lead to-

- Improved waiting time in OPD.
- Allow more time with the clinical team.