

Methods Two cohorts of SCD children and age and ethnic matched controls were recruited. Cohort one (47 SCD children and 26 controls) had a median age of 8.8 years at recruitment and were followed for two years. Cohort two (45 SCD children and 26 controls) were recruited at an older age (median age 10.2 years) than cohort one ($p = 0.007$) and were followed for ten years. Forced expiratory volume in one second (FEV_1), vital capacity (VC), forced expiratory flow between twenty-five and seventy-five% of VC (FEF_{25-75}), total lung capacity (TLC) and residual volume (RV) were measured at recruitment and at the end of follow-up.

Results In both groups of SCD children, but in neither control group, lung function declined significantly. The rate of decline was greater in cohort one than cohort two for FEV_1 ($p = 0.008$), VC ($p = 0.001$), FEF_{25-75} ($p = 0.030$), TLC ($p = 0.004$), and RV ($p = 0.043$). During follow-up, ACS episodes were more common in cohort one than cohort two (one episode per 1.93 patient/years versus one episode per 12.6 patient/years) $p < 0.0001$. ACS episodes were the only independent predictor of a greater decline in lung volumes.

Conclusions Lung function deteriorated in SCD children compared to similar aged and ethnic matched controls. The most rapid period of deterioration took place during early childhood when ACS episodes were more common. Our results suggest that treatment strategies to prevent ACS episodes need to be started in young SCD children if they are to be most effective in preventing the decline in lung function.

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THE IMPACT OF A STANDARDISED TRANSCRANIAL DOPPLER TRAINING PROGRAMME IN SCREENING CHILDREN WITH SICKLE CELL DISEASE: A EUROPEAN MULTI-CENTRE PERSPECTIVE

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10.1136/archdischild-2015-308599.356

Background Routine use of Transcranial Doppler (TCD) screening is standard management for the prevention of Stroke in children with Sickle Cell Disease (SCD). However, due to a number of factors including the lack of adequately trained TCD operators, less than 50% of children receive this service. The study objectives were to determine the effectiveness of modular TCD training, to improve the quality and standardisation of TCD assessment and thereby facilitate an increase in the number of children screened.

Methods The modular training programme comprised of a two-day course, covering theory and practical aspects of TCD and incorporating significant hands-on instruction. This was followed by local scanning with continuous monitoring and feedback from the training centre in the United Kingdom (UK). Competency evaluations were undertaken at the end of the instructional course and 6–12 months later when a log book of at least 50 scans was completed. Data were compared with that acquired from the same patients in the year prior to the training programme using imaging and/or non-imaging TCD. Statistical

analysis was performed using Pearson Chi-Square controlling for possible treatment bias.

Results Data were obtained from 326 patients (male 168 (51.5%); female 158 (48.5%); mean age 7.6 ± 3.5 , range 1–17) in the UK, Ireland and Italy. Genotypes were; HbSS 79%, HbSC 19%, HbSbetathalassaemia^o 1%, HbSbetathalassaemia⁺ 1%. 462 pre-training scans (imaging and/or non-imaging TCD); 134 from the UK, 193 from Ireland and 135 from Italy, and 377 post-training scans were available; 114 from the UK, 167 from Ireland and 43 from Italy. Statistical analysis revealed a significant difference in the STOP distribution between the three centres ($C^2 = 53$, $p < 0.001$) prior to training, with no treatment bias (no treatment $C^2 = 47$, $p < 0.001$; treatment $n = 82$, $C^2 = 23$, $p < 0.001$). Anomalous technique between centres pre-training included the erroneous use of Doppler angle correction, poor vessel/Doppler angle optimisation and inconsistent STOP velocity thresholds for imaging and non-imaging studies. After training the STOP distribution was similar in the three centres ($C^2 = 7.1$, $p = 0.311$; no treatment $C^2 = 11$, $p = 0.074$; treatment $n = 81$, $C^2 = 7.8$, $p = 0.252$). The consistent STOP distribution post-training, achieved using either imaging or non-imaging TCD,

Conclusion This is the first modular TCD training programme that has demonstrated efficacy when delivered in different European countries. TCD was either imaging or non-imaging techniques and should facilitate the more widespread.

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ASSESSING THE VALUE OF BONE MARROW ASPIRATE AND TREPHINE IN IDENTIFYING METASTATIC INVOLVEMENT IN CHILDREN WITH EWING'S SARCOMA: A RETROSPECTIVE SINGLE CENTRE EXPERIENCE

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10.1136/archdischild-2015-308599.357

Background Bilateral Bone marrow aspirates and trephines are part of the initial staging evaluation of patients with Ewing's sarcoma. However, the utility of performing this invasive investigation in addition to imaging with MRI and Technetium 99 bone scan has not been assessed.

Aim To assess the value of performing bone marrow aspirates and trephines in identifying metastases when compared to imaging, particularly Technetium 99 bone scan.

Methods Retrospective review of 48 children aged 16 and under with Ewing's sarcoma treated in our institution over a 14 year period (August 2000–September 2014).

Results The demographic details of our patients were as follows—we treated 25 males and 23 females (M:F = 1.08:1). 54% of patients were over 10 years old while 12.5% of patients were under 5 years old; the remaining 33.3% were aged between 5 and 10 years. Using imaging alone, 69% had localised disease while 31% had metastatic disease. 81% of patients ($n = 39$) had bone marrow aspirates and trephines performed, of which 3 were positive for disease; one of these patients had a pelvic primary and the marrow was positive on the left side which was the location of the primary site. 43 patients (90%) had a bone scan, of which 10 were positive for bony metastases. All three patients who had bone marrow positivity also had metastatic