COVID-19 exposure. Most patients were hypotensive at referral, and 67% needed transfer to PICU for inotropic support.

British Association of General Paediatrics

1095 POPULATION BASED SCREENING METHODS IN BILIARY ATRESIA – A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background Biliary Atresia (BA) is the leading cause of new-born cholestasis and the foremost reason for liver cirrhosis and liver transplantation in the paediatric population. The clinical course of BA can be improved with a Kasai portoenterostomy; however, this is time sensitive and delays in BA detection and treatment, with poor native liver survival, have been reported across paediatric hepatology centres worldwide.

The nature and clinical course of BA indicates a need for effective new-born screening. There is a clear definition of the illness, improved clinical outcomes from early recognition and potential cost savings. However, there is no consensus on the most effective method of population-based screening for the condition.

Objectives We aimed to systematically review and meta-analyse the methods of population-based screening for BA.

Methods We searched 11 databases between January 1 1975 and January 4 2021, identifying 5377 relevant titles. Studies exploring the use of a population screening tool to identify BA were included. Outcomes included sensitivity and specificity in screening for BA, age and time to Kasai, associated morbidity and mortality, and overall cost-effectiveness of screening. All studies underwent independent review by 2 trained reviewers, who extracted study data and assessed the risk of bias using the Newcastle Ottawa tool.

Results Twenty-four studies were identified, that included 2697 BA infants. Five methods of population-based screening for BA were present (number of papers): stool colour charts (SCC) (12), conjugated bilirubin measurements (4), assessments of stool light saturation (2), measurements of urinary sulphated bile acids (2) and assessment of bile acids in blood spots (4).

In a meta-analysis, conjugated bilirubin measurements were the most sensitive and specific in detecting BA, with an average sensitivity and specificity of 100.0% and 99.2% respectively. This was followed by urinary sulphated bile acid measurements (100.0%, 99.5%), SCC (88.7%, 99.9%), stool colour saturations (100.0%, 90.1%) and bile acid blood spot measurements (80.3%, 83.7%). Across 5 studies, the use of SCC was observed to reduce the age of subsequent Kasai to approximately 60 days, compared to 36 days for conjugated bilirubin measurements. The use of SCC and conjugated bilirubin was associated with improved overall and transplant free survival. Finally, the use of SCC was considerably more cost-effective than conjugated bilirubin measurements.

Conclusions Both SCC and conjugated bilirubin measurements are the most researched methods of population-based BA screening. Conjugated bilirubin measurements have improved sensitivity and specificity in detecting BA. However, its use is expensive and considered invasive. SCC appear to not provide acceptable improvements in the age of Kasai. Further research into the practicality of conjugated bilirubin measurements, as well as alternative methods of population-based screening for BA, are required.

British Academy of Childhood Disability

1096 MODERN ILLNESS OR A THING OF THE PAST? SURVEILLANCE STUDY OF CHILDHOOD/ADOLESCENT SYDENHAM’S CHOREA IN THE UK AND THE REPUBLIC OF IRELAND

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Background Sydenham’s chorea (SC) is a neuropsychiatric condition largely affecting children and adolescents, associated with prior group A streptococcal infection. SC is characterised by purposeless, involuntary, non-stereotypical movements of the trunk or extremities (chorea), often associated with muscle weakness and emotional and behavioural symptoms. Symptoms may range from mild to severe and last for two years or more. Although SC is considered a ‘rare disease’, such paediatric conditions may in fact have greater impact on families, who may become more isolated, lack information and experience more diagnostic delays. To date there have been no prior UK prospective surveillance studies to capture current incidence or to study presentation, management or outcomes. Working with partners including the Sydenham’s Chorea Association, we designed a surveillance study to be carried out through the British Paediatric Surveillance Unit (BPSU).

Objectives Our main objective was to conduct the first prospective surveillance study of SC in the UK and ROI, and describe the current paediatric service-related incidence, presentation and management of SC in children and young people aged 0–16.

Methods Using standard BPSU surveillance methodology, clinicians notified the BPSU when they saw a case meeting our inclusion criteria (new case of suspected or clinically confirmed SC). Clinicians were then contacted by the research team to complete a questionnaire on clinical presentation, investigation, management, and functional impairment. The case-reporting period lasted for 24 months from December 2018 to December 2020.

Results Over a 24 month period, 72 reports were made via BPSU, of which 40 were eligible cases of suspected or confirmed SC. The remainder were ineligible, duplicates, or did not have returned questionnaires from clinicians. The mean age of cases was 9 years, and 60% were female. The majority (65%) presented with ‘moderate’ severity of chorea. The most common neurological presenting features (apart from chorea) were loss of fine motor skills, gait disturbance, and dysarthria. Over 75% also presented with emotional and/or behavioural symptoms. Almost all cases had evidence of prior