encompassed psychiatric, psychological, psychosocial and behavioural consequences post-mTBI.

Results Electronic databases identified 577 journal articles, from which 11 studies were included in review. Based on literature, prevalence of inattention and conduct mean ratings increased in ages 10–13 with hospitalisation compared to those after outpatient clinic. Furthermore, 10% of children under 18 years with a history of multiple previous mTBIs, showed having significantly higher scores in emotionally reactive, withdrawn and aggressive problems compared to single or no mTBI history. Majority studies also found that that 35.7% of mild TBI group suffered new onset anxiety disorders compared to 11.4% in control group, particularly when patients had pre-existing psychiatric illness.

Conclusions Although the correlation between mTBIs and subsequent behavioural or psychological changes reached significance, there is still little evidence to support the true nature of this relationship. Ideally, future studies should identify the risk factors that may exacerbate post-head injury symptoms to ensure more holistic patient care. Moreover, detecting the persistence of psychiatric and/or behavioural changes long-term, rather than acute, would add greater significance to this correlation in children and adolescents following an mTBI.

Association of Paediatric Emergency Medicine

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IS THERE AN ASSOCIATION BETWEEN MILD TRAUMATIC BRAIN INJURY AND SUBSEQUENT BEHAVIOURAL AND/OR PSYCHOLOGICAL PROBLEMS IN CHILDREN UNDER 18 YEARS OF AGE?

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Background The incidence of paediatric mild traumatic brain injuries (mTBIs) diagnosed have increased exponentially within the past decade. Approximately 1.4 million people attend the emergency department (ED) due to mild head injury, within which 33–55% are children under 15 years. Once treated, the general practice is to acknowledge the emergence of medical post-concussive symptoms such as nausea or dizziness. Contrastingly, less so recognised are the behavioural changes that may arise, mainly due to lack of awareness regarding the myriad of possible outcomes. Thus, greater research regarding the association of behavioural and psychological changes following an mTBI are imperative to ensure follow-up after head injury is being conducted so holistic management can be achieved.

Objectives To conduct a literature review and investigate whether psychological or behavioural changes occur within children under 18 years proceeding mild traumatic brain injury.

Methods Six electronic databases searched for studies within the past 30 years for this literature review. Studies needed to meet the following criteria in order for selection: primary research studies, population age <18 years for both with mTBI and the control group (healthy children or those admitted for orthopaedic injury), 'mild' traumatic brain injury (loss of consciousness for <30 minutes and GCS 13–15) and

encompassed psychiatric, psychological, psychosocial and behavioural consequences.

Results Electronic databases identified 577 journal articles, from which 11 studies were included in review. Based on literature, prevalence of inattention and conduct mean ratings increased in ages 10–13 with hospitalisation compared to those after outpatient clinic. Furthermore, 10% of children under 18 years with a history of multiple previous mTBIs, showed having significantly higher scores in emotionally reactive, withdrawn and aggressive problems compared to single or no mTBI history. Majority studies also found that that 35.7% of mild TBI group suffered new onset anxiety disorders compared to 11.4% in control group, particularly when patients had pre-existing psychiatric illness.

Conclusions Although the correlation between mTBIs and subsequent behavioural or psychological changes reached significance, there is still little evidence to support the true nature of this relationship. Ideally, future studies should identify the risk factors that may exacerbate post-head injury symptoms for more holistic patient care. Moreover, detecting the persistence of psychiatric and/or behavioural changes long-term, rather than acute, would add greater significance to this correlation in children and adolescents following an mTBI.

British Inherited Metabolic Disease Group

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LESSONS FROM THE NEWBORN SCREENING FOR POMPE DISEASE: A SINGLE-CENTER UK EXPERIENCE BASED ON USA NEWBORN SCREENING PROGRAM

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Background Pompe disease is a rare genetic disorder caused by deficiency of alpha-glucosidase enzyme. The classical infantile form clinically presents in the first two months of life with myopathy and cardiomyopathy and is fatal in the first year of life if untreated. Enzyme replacement therapy (ERT) should be initiated as early as possible to improve outcomes through early identification of cases. Pompe disease can be identified by measuring alpha-glucosidase activity in newborn blood spot samples (NBS). Pompe disease is not included in the UK Newborn blood spot screening program, however babies resident on American military bases in the UK have their blood spot samples analysed in the USA by screening programmes that may include Pompe disease. Any abnormal screening results are followed up locally by UK paediatricians.

Objectives The aim of this study was to characterise the cohort of cases with increased risk of Pompe disease due to a positive initial newborn screening test.

Methods We have recorded the epidemiological, clinical, biochemical and genetic data of babies born at two local US Air Force bases and referred to CUH Metabolic outreach clinic in 2020 with increased risk of Pompe disease. NBS was undertaken by Maryland newborn screening laboratory.

Results Lymphocyte alpha-glucosidase activity was undertaken in four cases. Two cases had normal activity and no further investigations were undertaken. Two cases showed marginally

reduced alpha-glucosidase activity but had normal urine glucose tetrasaccharide excretion; one was compound heterozygote for variants of unknown significance (VUS) and the other had no clear pathogenic mutations. The remaining case had their blood spot alpha-glucosidase activity repeated which was towards the bottom of the reference interval, but urine tetrasaccharide excretion was normal. All 5 patients remain healthy on follow-up.

Case	Age at referral (months)	Bloodspot Enzyme Activity	Lymphocyte Enzyme Activity	Urine Glucose tetrasaccharide (HEX4)	GAA gene analysis
1	4.6	Normal	Marginally reduced	< 5 umol/L	2x VUS
2	3.5	Low end of normal range	Not done	< 5 umol/L	Not done
3	2.4	Not done	Marginally reduced	< 5 umol/L	No pathogenic mutations detected
4	2.7	Not done	Normal	Not done	Not done
5	8.0	Normal	Normal	Not done	Not done

VUS: variant of unknown significance

Conclusions Our experience of the follow-up of newborn screen-positive Pompe disease corroborates the high false positive reported by Pompe disease screening programs around the world. Since the UK does not screen for Pompe disease we developed a local confirmatory diagnostic pathway for the investigation of these asymptomatic infants. While we conclude that the results of NBS for Pompe disease should be interpreted cautiously, a clear algorithm is required for the rapid identification of true positives to expedite management decisions, particularly prompt treatment with ERT. A balanced approach is required when counselling families to reduce unnecessary parental stress caused by a false positive result.

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DOES THIS CHILD HAVE COVID-19 – A DESCRIPTIVE OBSERVATIONAL STUDY AND EDUCATIONAL STRATEGY FROM A PAEDIATRIC EMERGENCY DEPARTMENT

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Background When the WHO declared a global pandemic on 12 March 2020, as emergency paediatricians we prepared for an influx of cases similar to adult services. The picture that evolved was different with a dramatic reduction in paediatric emergency department (ED) attendances in the UK. While adults presented with severe respiratory illness children were mainly identified following universal screening of admitted patients and presented innocuously with a febrile illness, cough and tonsillitis or lethargy with poor feeding in

neonates. These children needed either symptomatic treatment or a short period of admission, usually less than 48 hours and had an uncomplicated clinical course. In April 2020 a new condition associated with COVID-19, PIMS-TS emerged with a clinical picture ranging from the benign to the life-threatening making identification challenging for Emergency and Paediatric staff.

Objectives The aim was to enable general and acute paediatricians in identification, initial investigation and management of children with potential SARS-CoV-2 infection and PIMS-TS.

Methods A descriptive analysis paper using local BHRUT data and incorporating national guidelines was produced for front-line ED clinicians and paediatricians for appropriate identification of children with PIMS-TS who presented to ED. Good clinical examination was emphasised, review of observations and treating unwell children according to National Institute for Health and Care Excellence and Advanced Paediatric Life Support guidelines with early escalation and discussion with tertiary centres.

Results On review of local data it emerged that the first four children with PIMS-TS had already presented to ED at BHRUT in April 2020 before the RCPCH defined the condition. Three children were admitted based on the clinical acumen of the receiving clinician. The children presented with fluid refractory shock or cardiac dysfunction or partial Kawasaki features of rash and conjunctivitis. A 15 year old appeared to be well whilst having abnormal observation and had a tonsillar focus. She was eating and drinking but hypotensive at presentation. She went on to develop fluid refractory shock within 6 hours and was transferred to PICU but ultimately had a good outcome. A fourth child was picked up on the third attendance to ED with fluid refractory shock. The timeline of presentation of both children was plotted on a template and used as an educational strategy to emphasise the varied and heterogeneous presentation of PIMS-TS.

Conclusions Subsequently 22 cases of PIMS-TS presented to BHRUT. Then intensive education and raised alerts resulted in appropriate identification of these children. For the emergency paediatrician, the mainstay of management remains considering the diagnosis and instigating supportive measures for children with PIMS-TS with early involvement of specialist teams. Observational studies using index cases ensures that there is a high index of suspicion to inform medical and nursing staff who will see these children in the early stage of disease when then can present non specifically mimicking an endemic viral illness. Emphasis was also laid on the fact that the proportion of febrile children without serious pathology presenting to ED would remain considerably higher than those with PIMS-TS to prevent potential over investigation.

Paediatricians with Expertise in Cardiology Special Interest Group

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A RARE CAUSE OF LONG QT- TIMOTHY SYNDROME

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Background TS is an extremely rare genetic disorder of the L-type cardiac channel Ca(V)1.2 encoded by *CACNA1C*. The syndrome is characterized by multi-system abnormalities