consisting of QT prolongation, congenital heart defects, syndactyly, facial dysmorphism, and developmental delay and autistic spectrum disorder.

Timothy syndrome (TS) is a rare genetic disorder characterized by an abnormally prolonged cardiac repolarization time (long QT interval). This predisposes individuals to arrhythmias, cardiac arrest and sudden death.

Objectives We want to report a case of Timothy syndrome, incidentally detected during induction for general anaesthesia.

Methods Electronic records were used to collect data

Results An 8 yr old boy was admitted to hospital for elective orchidopexy, during induction he was developed 2-degree AV block with T alternans, maintaining reasonable cardiac output throughout. Past medical history of one admission with possible seizure. Of note he is currently being evaluated for autism.

He was born at 38 weeks. labour was induced due to poor growth.

His physical exam was normal He subsequently had a 12-lead ECG which showed a prolonged QTc 0.504s. His genetic testing shows a pathogenic gene mutation in CACNA1C. Parents have been counselled for the need for implantable defibrillator. He has been given an external automated defibrillator in the meantime.

He is currently on nadolol 40 mg OD. His parents are waiting gene testing.

Discussion Classic Timothy syndrome (TS) is a rare genetic disorder with dysfunction in multiple organ systems, clinically characterized by long QT syndrome and syndactyly. Timothy syndrome was first described in 1992 as sporadic cases of long QT syndrome, congenital heart disease and syndactyly. Since then rare cases have been reported in the literature. Classic TS is caused by a single missense mutation G406R of exon 8A of the Cav1.2 L-type calcium channel gene (CACNA1C) and is inherited in an autosomal dominant fashion, although it usually is the result of a de novo mutation. Patients with TS are prone to life threatening ventricular arrhythmias as a consequence of a prolonged QT interval. Other cardiac manifestations include septal defects, patent ductus arteriosus, cardiomyopathy and Tetralogy of Fallot.

Since the affected gene is widely expressed in multiple adult and foetal tissues including gastrointestinal system, brain, lungs, immune system and testes, extracardiac manifestations are common in patients with TS. Many present with developmental delay, cognitive abnormalities and autism.

These patients are at high risk for sudden death due to life threatening ventricular tachyarrhythmia. Implantation of an ICD at a very young age may be the best means to prevent sudden death.

Conclusions Timothy syndrome is a rare congenital arrhythmia disorder with dysfunction in multiple organ systems

The risk for life-threatening ventricular tachyarrhythmia is the limiting factor of TS. Since ventricular tachyarrhythmia is the leading cause of death in patients with TS, effective anti-arrhythmic medication and an implantable cardioverter defibrillator are the mainstay of therapy

Paediatricians with Expertise in Cardiology Special Interest Group

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EFFICACY AND ADVERSE EFFECTS OF IBUPROFEN IN THE TREATMENT OF HAEMODYNAMICALLY SIGNIFICANT PATENT DUCTUS ARTERIOSUS (PDA) IN PRETERM INFANTS

¹Catherine Graham, ²Yogen Singh. ¹University of Cambridge Clinical School; ²Cambridge University Hospitals NHS Trust

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Background Ibuprofen is used as first line therapy to close haemodynamically significant patent ductus arteriosus (PDA) but is associated with severe gastrointestinal and renal effects.

Objectives The aims of this study were to determine the efficacy and safety of ibuprofen for pharmacological closure of PDA in preterm infants ≤ 29 weeks gestation.

Methods Retrospective study between 2015–2019 at a tertiary centre. 82 infants ≤ 29 weeks gestation with a haemodynamically significant large PDA on echocardiogram were enrolled. Data was collected using an electronic patient record system.

Results As shown in table 1, cohort had a median gestational age of 25+3 (range: 23+2 to 29+1), a median birth weight of 0.79kg (range: 0.39–1.25kg) and a median PDA diameter of 2.7mm (range 1.6–4.0mm). 70/82 infants completed their first course of ibuprofen, achieving full PDA closure in 25 patients (30.5%), partial closure in 29 (35.4%), and no closure in 24 (29.3%). 13/82 patients (15.9%) required a second course of ibuprofen, achieving full PDA closure in 3 patients (23.1%), partial closure in 1 (7.7%), and no closure in 9 (69.2%).

Associated adverse effects were reported in 13/82 patients (15.9%). These were mainly gastrointestinal and included necrotising enterocolitis in 5/82, bowel perforation in 5/82, and a further 1/82 had an upper gastrointestinal bleed. 9 patients subsequently underwent surgical ligation of their PDA, and an additional 5 patients died prior to PDA ligation due to non-cardiac complications.

Conclusions In our cohort ibuprofen was moderately efficacious in closure of PDA. Ibuprofen was generally well tolerated, although 13/82 infants (15.9%) were noted to have adverse effects. Whether these gastrointestinal adverse effects

Abstract 597 Table 1 Baseline characteristics of cohort

Baseline characteristics	Cohort no. = 82
Gestational age (weeks+days)	25 + 3 (range: 23+2 - 29+1)
Birth weight (kg)	0.79 (range: 0.39 - 1.25)
PDA diameter (mm)	2.7 (range: 1.6 - 4)
LA:Ao ratio	1.8 (range: 1.2 - 2.7)
Left to right shunt (n)	79 (96.3%)
Pulsatile flow pattern (n)	53 (64.6%)
Diastolic steal documented (n)	34 (41.4%)

Abstract 597 Table 2 Extent of PDA closure and adverse effects following one and two standard courses of ibuprofen. Frequency of individual adverse effects are given as a cumulative% across both ibuprofen courses

	Commenced first course (n=82)	Commenced second course (n=13)
Completed course	70 (85.4%)	11 (84.6%)
Full closure	25 (30.5%)	3 (23.1%)
Partial closure	29 (35.4%)	1 (7.7%)
No response	24 (29.3%)	9 (69.2%)
Response not documented	4 (4.9%)	0
Adverse effects	11 (13.4%)	2 (15.4%)
Adverse effects	Cohort n = 82	
Bowel perforation	5 (6.1%)	
Necrotising enterocolitis	5 (6.1%)	
Intraventricular haemorrhage	2 (2.4%)	
Renal failure	1 (1.2%)	
Thrombocytopenia	1 (1.2%)	

were the consequence of a persistent haemodynamically significant large PDA or the adverse effects of ibuprofen, or a combination of the two, is hard to say from our small cohort. A future prospective study should be planned to address this.

Association of Paediatric Emergency Medicine

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CHANGING PATTERNS OF EMERGENCY PAEDIATRIC PRESENTATIONS DURING THE FIRST WAVE OF COVID-19: LEARNING FOR THE SECOND WAVE FROM A UK TERTIARY EMERGENCY DEPARTMENT

¹Shaarna Shanmugavadivel, ¹Jo-Fen Liu, ²Colin Gilhooly, ³Loai Elsaadany, ³Damian Wood. ¹University of Nottingham; ²Children's Emergency Department; ³Nottingham Children's Hospital

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Background The SARS-CoV2 pandemic and initial public health response led to significant changes in health service delivery, access and utilisation. The SARS-CoV2 illness burden in children and young people (CYP) is significantly lower than in adults. To inform effective child public health interventions we aimed to compare patterns of paediatric emergency department presentations.

Objectives Describe the ED attendance pattern of CYP during COVID-19 compared to the same period in 2019.

Methods Retrospective review of all attendances (0–18yrs) over the initial pandemic (02/03/20–03/05/20) compared to 2019. Outcome measures included demographics, number of attendances, source of referral, presenting complaint, discharge diagnosis and disposal. Descriptive statistics, with subgroup analysis by age/sex/ethnicity and pandemic time-periods (pre-lockdown, lockdown weeks 1–3 and lockdown weeks 4–6) were performed.

Results 4417 attendances (57% illness; 43% injuries) occurred between 02/03/20–03/05/20, compared to 8813 (57% illness; 43% injuries), a reduction of 50%, maximal in lockdown week 2 (-73%).

Presenting complaints

2529 (57%) attendances were illnesses and 1868 (43%) injuries, compared to 5005 (57%) and 3764 (43%) respectively in 2019. The top five illness complaints were breathing difficulty, fever, abdominal pain, diarrhoea & vomiting and rash, unchanged from 2019. Ranking of top 3 illness presentations changed across the pandemic weeks. Breathing difficulty dropped from 1st (300;25%) to 2nd (117;21%) to 3rd (59;11%); ($p<0.001$). Abdominal pain rose from 3rd pre-lockdown (87;7%) and lockdown weeks 1–3 (37;7%) to 2nd in weeks 4–6 (62;12%; $p=0.004$). Fever ranked 2nd (235;19%) pre-lockdown, 1st (134;24%) in week 1–3 and week 4–6 (94;18%; $p=0.035$).

Triage category

88 (2%) attendances were triaged as a category 1 (CAT1) and 544 (12%) attendances as a category 2 (CAT2); this is compared to 146 (2%) and 1134 (13%) in 2019. There was no variation in CAT1 across pre-lockdown (2%), lockdown weeks 1–3 (2%) and lockdown weeks 4–6 (2%) and minimal variation in CAT2 (12%, 13% and 11% respectively across pandemic weeks), indicating no change in severity of presentations.

Source of referral (SOR)

3065/4417 attendances (69%) were by self-referral, proportionally the same as in 2019 (6149/8813; 70%). Emergency services/urgent care were the second most common SOR for 487 (11%) attendances, similar to 2019 (762/8813; 9%). NHS111 referral ranked third (416/4417; 8%) and GP ranked 4th (315/4417;7%). This was different to 2019, where GP referral ranked 3rd (755/8813; 9%) and NHS111 ranked 4th (734/8813;8%).

Disposal

708 (16%) attendances resulted in admission, 3647 (83%) were discharged, 61 (1%) did not wait (DNW) and 1 patient died during the pandemic period. This was compared to 1675 (19%) admissions, 6755 (77%) discharges, 383 (4%) who DNW and no deaths in 2019.

Conclusions Despite a 50% reduction in attendances there was no significant change in proportions of illness/injury, acuity of illness, source of referral or disposal. Rank of illness presentations changed, with abdominal pain ranking second, and fever first, an important change which should prompt further research into causes. Clear public health messaging for CYP and collaborative guidance for primary care is required in this second wave to ensure CYP have access to appropriate emergency, routine and mental health services.

British Association for Community Child Health

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ADHD MONITORING: AUDIT OF NICE GUIDELINES

¹Anna-louise Power, ²Elham Abbas. ¹NHS; ²Wigan Warrington and Leigh NHS Trust

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Background ADHD is a complex, diverse and common neuro-developmental disorder, associated with many comorbidities. NICE guidelines set out the monitoring requirements for children being treated for ADHD. Wigan has more than 1000 children on its database with ADHD on medication, mostly on stimulant medication. Monitoring of the effectiveness of