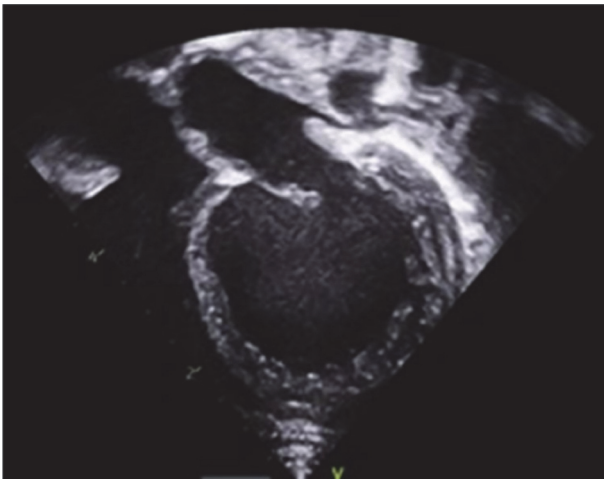


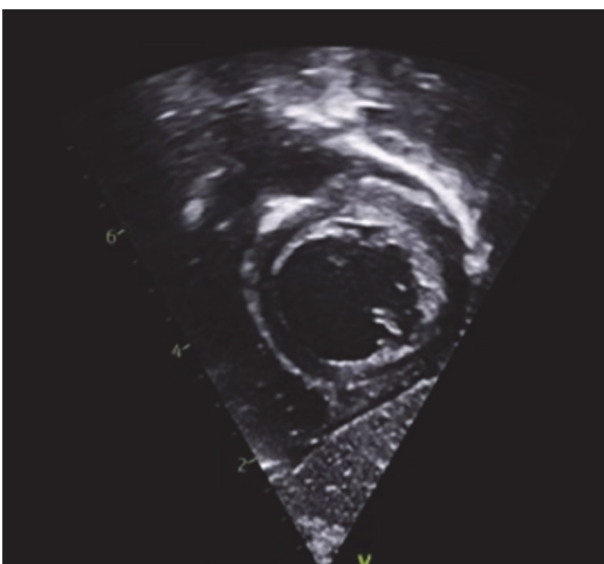
**Background** Patau syndrome is considered the third most common trisomy with an estimated prevalence of 1:20,000-29,000. However, the prevalence in the UK is documented as 2 in every 10,000 births. The overall 5-year survival rate for a child with trisomy 13 is 9.7%. Central apnoea and multi-organ failure are the most common causes of early death in this cohort of patients where cardiac lesions do not commonly lead to premature deaths.

Common cardiac lesions found in patients with Trisomy 13 include ventricular septal defects (VSD), Tetralogy of Fallot (TOF), Patent Ductus Arteriosus (PDA), VSD + coarctation/arch hypoplasia, Atrial Septal Defect (ASD), coarctation of the aorta, single ventricle, complete atrioventricular septal defect, and Pulmonary atresia VSD. Very rarely, left ventricular non-compaction associated with progressive heart failure had been documented in 2 patients with trisomy 13 in the literature.

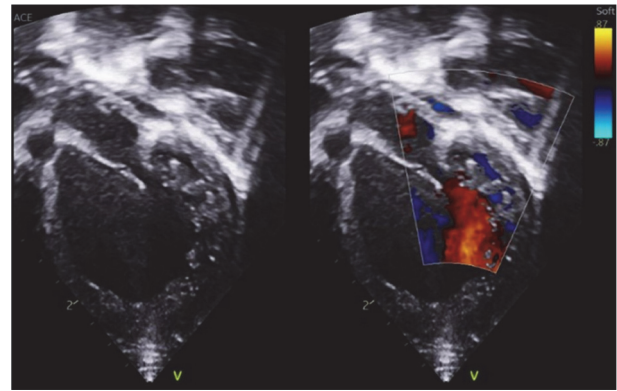
**Case Presentation** A baby girl was born at 40+2 weeks gestation by emergency C-section due to lack of progression. She had an uncomplicated birth. However, her genetic studies



**Abstract 86 Figure 1** Apical four chamber view showing dilated left ventricle with evidence of noncompaction



**Abstract 86 Figure 2** Subcostal short axis view of LV showing thin LV muscle mass



**Abstract 86 Figure 3** Apical four-chamber view showing dilated left ventricle with evidence of non-compaction

performed due to the antenatal suspicion, confirmed Trisomy 13 (47, XX,+13). Her echocardiogram at 5 months of age revealed a small stretched PFO with a dilated, thin-walled and noncompaction left ventricle associated with mild-moderate left ventricular systolic dysfunction. She had no post-tricuspid shunts and had normal coronary origins. Her left ventricular outflow tract and aortic arch were patent with otherwise normal intracardiac anatomy.

Her ECG revealed sinus rhythm without ventricular ectopics or repolarisation changes.

She was commenced on diuretic therapy.

**Conclusion** This case report demonstrates the rare association of left ventricular noncompaction cardiomyopathy in a patient with Patau syndrome which could be the life-limiting factor in patients with Trisomy 13. The case highlights the importance of detailed cardiac evaluation and follow-up in this cohort of patients

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#### DELIVERING QUALITY HEALTHCARE FOR ALL THROUGH EQUITABLE ACCESS, EXCELLENT EXPERIENCE AND OPTIMAL OUTCOMES: HOW GREAT ORMOND STREET HOSPITAL IS TACKLING HEALTH INEQUALITIES

Pippa Sipanoun, Darren Darby, on behalf of the Great Ormond Street Hospital Health Inequalities Steering Committee. <sup>1</sup>Great Ormond Street Hospital for Children NHS Foundation Trust, UK, UCL Great Ormond Street Institute of Child Health, UK and University of Surrey; <sup>2</sup>Great Ormond Street Hospital for Children NHS Foundation Trust, UK

10.1136/bmjpo-2023-GOSH.62

**Background** Health inequalities are the avoidable, unfair and systematic differences in health outcomes between different groups of babies, children and young people. We know that over half of the families at Great Ormond Street Hospital for Children are disproportionately at risk of or are vulnerable to experiencing avoidable health inequalities.

**Our Commitment** Great Ormond Street Hospital is committed to reducing health inequalities affecting our patients and families, with the aim of bridging the gap for patients with complex health needs who are impacted by health inequalities.

**Strategies in place to tackle health inequalities**

- Bi-monthly Health Inequalities Steering Committee meetings
- The formation of five working groups to examine and implement initiatives related to: access, patient experience, outcomes, digital exclusion, and awareness; reporting to the Health Inequalities Steering Committee

- Learning from the findings of the Going Digital Study, which highlighted key health inequalities that may lead to inequities
- The collection and analysis of accurate, up-to-date data to determine the hospital's current health inequality priorities, for example, 'Was not brought' and digital exclusion data, and the development of a health inequalities data dashboard
- Collaboration across clinical services to understand and implement changes to reduce inequities in access, experience and outcomes for defined cohorts of patients/families most at risk of experiencing health inequalities
- Collaboration with Integrated Care Boards, the Children's Hospital Alliance, partner organisations, and policy makers to address health inequalities, and to implement changes to improve outcomes in population health and healthcare
- Developing awareness, accountability, and insight on the impact of health inequalities as an organisation through the delivery of health inequalities workforce education
- Advocating for change and improvement on a national scale.

**Conclusion** The impact of health inequalities on families is high on the Trust's agenda. Through a strategic, collaborative approach, steps are being taken to address these inequalities and resulting inequities.

#### 101 ANGIOGRAM AND RENAL ANGIOPLASTY IN A PATIENT WITH WILLIAMS SYNDROME

David Cunningham, Maryam Zaky. *Great Ormond Street Hospital for Children NHS Foundation Trust, UK*

10.1136/bmjpo-2023-GOSH.63

**Background** Williams Syndrome is a genetic condition occurring in approximately 1 in 10,000 live births. The deletion of genes in chromosome 7 affects tropoelastin, a protein involved in vascular wall formation. This results in a multisystem disorder predominantly affecting connective tissue, the central nervous system, and the cardiovascular system. Some surgical interventions can exacerbate haemodynamic changes and pose challenges for the anaesthetist when a patient with Williams Syndrome requires a general anaesthetic.

**Case Report** We present the case of a 7-year-old girl with Williams Syndrome undergoing angiography and angioplasty to her only kidney and explore the perioperative challenges for the anaesthetist. Prior to her transfer to Great Ormond Street Hospital, she had a prolonged hospital admission for control of hypertension and underwent neurosurgical intervention following an intracranial haemorrhage. Blood pressure was being controlled with seven antihypertensive medications. Echocardiogram showed left ventricular hypertrophy, severe left atrial dilatation, mitral stenosis and pulmonary hypertension. A cardio-stable induction was achieved with ketamine and total intravenous anaesthesia with propofol. An arterial line was sited in the right radial artery to monitor beat-to-beat changes in systemic blood pressure and non-invasive blood pressure monitoring sited on the leg. Blood pressure was optimised throughout the procedure using phenylephrine. Following a short recovery period, she was transferred back to the ward and was discharged two days after the procedure.

**Conclusion** Patients with Williams Syndrome undergoing general anaesthesia offer challenges to the anaesthetist. Careful consideration should be given to the cardiovascular implications of such cases and the approach to minimising changes in blood pressure.

#### 103 ATTACHMENT AND TRAUMA: A CASE STUDY ON THE CHALLENGES OF IMPLEMENTING FAMILY CENTRED AND TRAUMA INFORMED CARE ON A PAEDIATRIC PATIENT

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10.1136/bmjpo-2023-GOSH.64

**Background** 'Mother is the whole world' was an article written in 1952 by attachment theory pioneer, John Bowlby. How are we, as healthcare providers in 2023, influenced by the theory of attachment? How do we support one another and families in providing care that supports the emotional needs of the child, in particular children who have experienced trauma? Such questions are pertinent to the care of our Paediatric population, their families and our colleagues, post-pandemic.

**Methods** A qualitative case study of a patient exhibiting high levels of psychological distress within the hospital environment during the pre-operative phase was compiled. The case-specific factors analysed were: patient background and attachment style, their response to the hospital environment, concerns voiced by parents, supports utilised by the family, outstanding supports available and staff approach to care. The themes included in the research process were: attachment, family centred care, trauma informed care, principles and practical approaches, challenges in practice and staff engagement. This cross-sectional analysis was then summarised into a presentation, aiming to deepen the understanding of the practical approaches and challenges faced across a multidisciplinary team.

**Results** This study showed that family centred care is considered a practical and preferred approach to promoting bonding within the contemporary family unit. Furthermore, the study revealed an array of challenges to providing trauma informed care such as: a lack of education concerning attachment and trauma, breakdowns within the multidisciplinary team's approach to care which aggravate these problems, an increase in adult professional anxiety and reduced resources.

**Conclusion** In conclusion, to rise to the challenges of implementing family centred and trauma informed care there is a need for targeted improvements in education, systemic approaches and the availability of key resources in order to adequately support our paediatric population, their families and our colleagues, post-pandemic.

**Acknowledgements for funding or support** I would like to express my appreciation and gratitude to Kevin Viviers-Hogreve, whose support and guidance aided me in completing this work.

#### 104 HOW THE CRF SUPPORTED SATELLITE RESEARCH TEAMS TO DRIVE OUR RESEARCH HOSPITAL VISION

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10.1136/bmjpo-2023-GOSH.65

GOSH hospital has a strategic vision to become a research hospital and build on our position as a key Paediatric partner for research industry by fully integrating research into clinical services. Our vision is in line with the UK 10-year vision for