services, demonstrating that fundamental nursing skills such as compassion, communication, flexibility and kindness are transferable, and the contributions of Children's Nurses in these and other fields were important and valued by others.

63 DOCUMENTATION OF CO-MORBIDITIES IN PATIENTS HAVING A SURGICAL PROCEDURE

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10.1136/archdischild-2020-gosh.63

Children with multiple comorbidities are typically referred to GOSH as GOSH is a tertiary centre for management of children with complex medical and surgical conditions.

Often these children require multiple admissions. This requires several patient encounters, which raises opportunity for missing important clinical information. The team re-audited how well, patient co-morbidities were documented on EPIC (electronic patient record). This is not only useful for the clinical team for accuracy of information but also has financial implications for the hospital.

A total of 71 children underwent a general surgical procedure over a 4-week period. Their inpatient notes were analysed, specifically Pre-op clerking (Admission note), Operation note & Discharge summary. Of these, only 16 patients did not have any associated comorbidity. The findings show 92.9% (Pre-op clerking), 35.2% (Op-note) & 57.7% (Discharge Summary) had accurate documentation of comorbidities. Only 1.8% of patients had missed comorbidities post EPIC compared to 29% pre-EPIC. This echoes that EPIC has made it effortless for coding team to record clinical information accurately & has important financial implications for the trust.

64 WALKER WARBURG SYNDROME (WWS) WITH ISPD GENETIC MUTATION- A CASE REPORT

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10.1136/archdischild-2020-gosh.64

Background Walker Warburg dystroglycanopathy is the most severe subtype of congenital muscular dystrophies. It is characterised by four consistent features -cobble stone lissencephaly, cerebellar malformation, retinal abnormalities and muscular dystrophy. The incidence is estimated at 1.2 per 100,000 live births and the life-span in these children is around 3 years. There are more than fifteen causative genes and transmission is autosomal recessive.

Case We describe a boy, born in Kuwait to parents of a consanguineous marriage, premature at 34 weeks gestation with hypotonia, antenatal hydrocephalus and severe eye abnormalities. An MRI brain done at the referring hospital showed cobblestone lissencephaly and severe ventricular dilatation. He was transferred to our centre at corrected age 38 weeks for neurosurgical and medical management. Clinical examination showed central hypotonia with a lack of purposeful movements, macrocephaly from hydrocephalus, left eye buphthalmos and congenital glaucoma, a blind

microphthalmic right eye, hearing impairment and bilateral medullary nephrocalcinosis. The creatinine kinase was elevated at 3996U/l. A genetic panel for muscular dystrophy done here detected a homozygous ISPD c.1186G>T p. (Gly396Ter) pathogenic sequence variant, confirming ISPD-related congenital muscular dystrophy. Complications included cerebrospinal fluid leak post ventriculoperitoneal (VP) shunt insertion, recurrent aspiration pneumonia and urinary tract infections. Of note, he developed hypothermia as a side effect from Timolol eye drops as part of his treatment for glaucoma. He underwent a fundoplication gastrostomy; slowly tolerating feeds with anti-reflux medications, breathing comfortably in air and cooing in response to parents' handling. Within a comprehensive multidisciplinary team effort, lung protective strategies, regular occupational and physiotherapy, he made a good recovery. He was successfully repatriated home with good parental care at corrected age 7 months.

Conclusion Prognosis may be poor in WWS, however it is most ethical to provide holistic care, optimise the quality of life and direct genetic counselling.

65 MANAGEMENT OF EARLY-ONSET SCOLIOSIS IN PATIENTS WITH PRADER WILLI SYNDROME WITH MAGNETIC GROWTH RODS – A CASE SERIES REVIEW

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10.1136/archdischild-2020-gosh.65

Background Prader Willi syndrome (PWS) is a rare genetic disorder characterised by developmental delay and hyperphagia. It is strongly associated with scoliosis, largely attributed to hypotonia and ligamentous laxity. Management of scoliosis in these patients can be challenging due to early onset of disease, higher BMI, poor bone quality and poor brace compliance. Skin picking behaviours may also have a significant impact on poor healing. Magnetic Expansion Control growing rods (MAGEC) have become the surgical management of choice in some patients with progressive early-onset scoliosis, however there have been no published reports on their use in the PWS population. Here we report our experience of treating children with PWS with MAGEC rods.

Methods A retrospective review of children with PWS who underwent MAGEC rod insertion between March 2010 to March 2020 at a quaternary paediatric hospital. Spinal column distraction was measured between the proximal and distal vertebrae in which fixations were inserted.

Results 4 patients with PWS had MAGEC rod inserted during the study period. All had undergone MRI imaging prior to surgery. Median age at primary operation was 5.5 years. All patients were overweight at time of primary operation (median BMI 94.5th centile).

A total of 11 MAGEC rods were used of which 5 had reported instrumentation failures (45%). These included 2 pin fractures and 3 proximal construct pull-outs. 2 of the patients developed post-operative infection requiring surgical intervention with 1 undergoing removal of metalwork.

Overall a median of 4.3 mm (IQR=1.1–25.3) of spinal distraction was achieved either prior to definitive spinal fusion or at last follow-up with median distraction per year of MAGEC of 1.1 mm (IQR=0.5–5.1).

Conclusion Management of early onset scoliosis in patients with PWS using MAGEC rods is challenging and can be associated with a high complication rate. Limited spinal growth was observed amongst this cohort.

66 INCREASING RECRUITMENT TO GOSH SAMPLE BANK

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10.1136/archdischild-2020-gosh.66

Background GOSH Sample Bank is a key Research Hospital initiative, enabling patients' leftover samples to be stored and potentially used for child health research. It was launched to staff, patients and families in 2019, following extensive piloting to determine acceptability, feasibility and resource requirements. Initial recruitment was slow, with 83 patients recruited between September 2017 and April 2020. As all patients should have the opportunity to participate, we investigated different methods to reach more patients and increase recruitment.

Aim To increase inpatient recruitment to Sample Bank, raise awareness of the initiative amongst ward staff, deliver training to staff and develop a model for sustainability to help achieve the Research Hospital vision for this initiative.

Method To drive engagement, especially at an extremely busy time clinically, Research and Innovation (R&I) staff worked with nursing staff to identify wards to visit to seek consent. The training needs of R&I staff, who are familiar with seeking research consent and are GCP compliant, were determined and a training plan was implemented to ensure familiarity with the initiative and competency in obtaining informed consent.

Results Over a 5 month period from April 2020, 308 patients were consented to Sample Bank, across 4 wards (Sky, Koala, Pelican and Panther). Of all patients identified or approached, 65% consented, 13% declined, 6% were ineligible, 13% were discharged before consent could be taken and 3% were eligible but not approached. In total, 24 clinical and 18 non-clinical staff were trained to obtain consent.

Conclusion Although resource intensive, this method of obtaining consent proved highly effective at increasing recruitment. We will now investigate opportunities for embedding obtaining consent for Sample Bank into routine clinical practice, including providing training for clinical staff and amending the clinical consent process, providing all GOSH patients with the opportunity to participate.

67 ADVANCED NURSE PRACTITIONERS IN CLINICAL RESEARCH: AN INNOVATIVE ROLE AT GREAT ORMOND STREET HOSPITAL

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10.1136/archdischild-2020-gosh.67

Advance Nurse Practitioner (ANP) roles in clinical research provide significant scope to utilise research resources more efficiently while offering a high level of specialised holistic care. However, within Clinical Research Facilities (CRF) throughout the United Kingdom (UK), there are very few ANP roles.

Great Ormond Street Hospital (GOSH) now have two ANP roles and one trainee ANP role within the Research and Innovation department. This is a role currently only present in three other trusts across the UK with the first such role commencing in Manchester in 2012. The first paediatric position was established in Birmingham in 2015, and the first trainee position at GOSH in 2017. Since 2019 there have been two fully qualified ANPs in research at GOSH with another trainee post developed in 2019.

A national forum for ANPs in research was formed in 2018. This has been pertinent in tackling the historic and evolving challenges collectively, collaborating on initiatives to raise the awareness of these unique roles within the NHS and across key stakeholders, as well as supporting clinical research teams who are keen to establish the roles within their departments.

There are many challenges involved in any new role and there have been common challenges recognised across the national forum. These vary from local issues of recognition of the role within the trust to wider issues related to Good Clinical Practice guidelines. Many of these challenges have been overcome and there have already been many successes within this role.

It is hoped in the future these roles will grow into a pivotal part of clinical research networks with the profile being raised nationally and internationally.

68 PREVALENCE AND TREATMENT OF VITAMIN K DEFICIENCY IN PAEDIATRIC PATIENTS WITH RECESSIVE DYSTROPHIC EPIDERMOLYSIS BULLOSA- SEVERE SUBTYPE (RDEB-S)

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10.1136/archdischild-2020-gosh.68

Introduction Patients with RDEB-S are at risk of vitamin K deficiency, potentially causing abnormal clotting, excessive bleeding, poor bone metabolism and abnormal vascular calcification.

This study quantifies vitamin K deficiency prevalence in this cohort and identifies potential risk-factors to prevent deficiency.

Methods RDEB-S patients who attended the EB service between 2014–2020 were included. Serum vitamin K and PIV-KAII were measured within the nutritional blood screen. Dietetic and medical notes were reviewed to establish: antibiotic use, enteral feed intake and micronutrient supplementation.

Results 16/25 64% (10/16 female), 22–180 months, had serum Vitamin K and PIVKAII analysed. 6/16 (37.5%) patients had vitamin K deficiency requiring supplementation.

2/6 (33.3%) normalised serum vitamin K after 12 weeks supplementation with oral Menadiol Diphosphate. 4/6 (66.6%) await re-testing following supplementation.

6/6 (100%) patients with vitamin K deficiency were not consuming a gastrostomy/sip feed. 9/10 (90%) patients with sufficient vitamin K levels were consuming either; minimum 200 mls prescribed sip feed or 400–800 mls gastrostomy feed daily (containing 5.9µg–11µg/100 mls vitamin K). One patient with normal vitamin K and no enteral feeds had raised PIVKAII.

16/16 (100%) received antibiotics (range 1–4 courses/year). Patients with the most frequent antibiotics (n=4) had normal