The aim of this project was to enhance the awareness of asthma and its understanding through a new informative and engaging educational resource.

Methods We designed an educational booklet aimed at children aged 4–10 called Asthmanauts. The child becomes the hero defeating space villains (triggers including pollen and cigarettes) with their inhaler rockets named Captain Blue and Commander Brown. The booklet was trialled at two hospital outpatient departments. A questionnaire given before and after reading the booklet was used to assess its impact on patients' awareness and perceived understanding of key terms. In addition parents were asked if the booklet could enhance their child's knowledge and whether they would recommend it.

Results 87.5% of parents (n=16 mean age of child=6 years; 11 males and 5 females) found the booklet enjoyable to read with their children; over 90% found it useful in furthering their child's understanding and would recommend it to others. The correct understanding among children of key terms including triggers, reliever and preventer showed a 14% improvement with the greatest impact on trigger understanding. Doctors and nurses also complimented the booklet requesting copies to use.

Conclusion The initial results of this pilot study are very promising and have shown that Asthmanauts has a potential role in asthma education for children. It provided effective information and was recognised and commended by health professionals and patients while being engaging and enjoyable. We now hope to distribute the booklet and determine its role in longer term management.

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G463(P)

FIRST CASE OF CONSTITUTIONAL MISMATCH REPAIR DEFICIENCY SYNDROME ASSOCIATED WITH BRONCHIECTASIS

¹C Swinburne, ²A Barclay, ³PE Beattie, ⁴R Davidson, ⁵D Murphy, ⁶R O'Kane, ⁷S Zuberi, ¹A Devenny. ¹Department of Respiratory Medicine, Royal Hospital for Children, Glasgow, UK; ²Department of Gastroenterology, Royal Hospital for Children, Glasgow, UK; ³Department of Dermatology, Royal Hospital for Children, Glasgow, UK; ⁴Department of Clinical Genetics, Queen Elizabeth University Hospital, Glasgow, UK; ⁵Department of Oncology, Royal Hospital for Children, Glasgow, UK; ⁶Department of Neurosurgery, Royal Hospital for Children, Glasgow, UK; ⁷Department of Neurology, Royal Hospital for Children, Glasgow, UK

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Introduction Constitutional mismatch repair deficiency (CMMRD) syndrome is a rare autosomal recessive disorder that results from homozygous germline mutations in one of the mismatch repair genes *MLH1*, *MSH2*, *MSH6* and *PMS1*. CMMRD syndrome results in a predisposition to childhood malignancy, with an increased risk of developing central nervous system (CNS), haematological and gastro-intestinal (GI) tract cancers. Colorectal polyps and cutaneous manifestations resembling neurofibromatosis type 1 are common. We present the case of a patient with CMMRD syndrome and bronchiectasis, an association not previously documented in the literature.

Case report A 13-year-old boy of Pakistani origin born to consanguineous parents was referred to the paediatric outpatient clinic with anaemia. Both parents had a diagnosis of Lynch syndrome and elder brother a diagnosis of CMMRD syndrome with a previous history of CNS primary neuro-ectodermal tumour and basal cell carcinoma. Consultation revealed a history of chronic productive cough and recurrent lower respiratory tract infections with evidence of finger clubbing on examination. Cutaneous manifestations included a large scalp congenital melanocytic naevus, multiple CALMs, hypopigmented lesions and axillary and inguinal freckling. Persistent left lower lobe changes were noted on chest X-ray. Bronchoscopy demonstrated normal airway anatomy with increased secretions noted from left lower lobe bronchus. CT chest confirmed a diagnosis of bronchiectasis. Sweat test and ciliary biopsy were normal. Immunoglobulin A was low at <0.7 g/L while lymphocyte subset analysis was unremarkable. Baseline pulmonary function testing demonstrated a moderate restrictive defect with no evidence of obstruction. Genetic screening was performed in light of family history which identified a familial homozygous *MSH6* sequence variant, c.3175del p.(Val1059fs), consistent with a diagnosis of CMMRD syndrome.

Bowel screening was undertaken and colonoscopy identified a large colorectal villous adenoma with focal high-grade dysplasia. The patient subsequently developed focal seizures and was diagnosed with a right fronto-parietal brain tumour with three metastatic lesions. Biopsy, although not conclusive, was felt to be representative of a high-grade glial or embryonal tumour.

Conclusion CMMRD syndrome is associated with a complex array of clinical manifestations and a wide tumour spectrum. Bronchiectasis has not been previously documented in the literature and should be considered where a history of chronic respiratory symptoms exists.

British Association of Paediatricians in Audiology

G464

NATIONAL SURVEY OF THE AETIOLOGICAL ASSESSMENT SERVICE OF PERMANENT CHILDHOOD HEARING IMPAIRMENT IN IRELAND

¹JM Balfe, ²S Doyle, ³D Waldron, ⁴N van der Spek. ¹Department of Developmental Paediatrics and Neurodisability, Tallaght University Hospital, Dublin, Ireland; ²Our Lady's Children's Hospital, Dublin, Ireland; ³St Luke's Hospital, Kilkenny, Ireland; ⁴Cavan General Hospital, Cavan, Ireland

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Introduction The Universal Neonatal Hearing Screening programme was implemented in Ireland in 2014. Approximately 100 infants are identified with a permanent childhood hearing impairment (PCHI) each year, an incidence of 1.5/1,000.

Best practice dictates that all children who are identified with PCHI should have access to prompt paediatric assessment to determine the need for aetiological investigations. The implementation of the UNHS programme did not include any increased resource for paediatrics thus raising concern regarding timely access to medical assessment.

This survey aimed to assess the provision of aetiological assessment services nationally with a view to informing future service development.

Methods All paediatricians involved in aetiological assessment were identified from the national audiology lead. A questionnaire was developed using the BAAP standards and was sent by post with email reminders.

Results were collated on excel.

Results 38 consultants were identified of whom 37 responded. 33 paediatricians reported that they assessed children with PCHI in clinic. Only 6/33 had received any specific training in the assessment of PCHI infants. This work was not included in any job plan and no additional posts had been developed. Only 1 paediatrician reported participating in a regional support network, but the vast majority (92.9%) surveyed expressed an interest in developing a regional network. 5/33 (15%) saw the infants in a dedicated hearing clinic.

Waiting time for assessment was beyond the recommended timeframe in all cases, (range 2–52 weeks, average of 14.4 weeks). Only 8/29 (27%) assess by 6 weeks of age, the time limit for treatment of congenital CMV infection. Access to MRI is limited, only 12 (28.6%) reported having access to MRI within 12 weeks of age thus the majority of MRI require sedation. 77% routinely refer all children with PCHI to ophthalmology.

Conclusion The above survey highlights significant deficits in the paediatric component of the UNHS. Identified gaps include; inadequate appropriately trained staff and lack of timely access to necessary investigations and tertiary services.

A national multi-disciplinary working group has been developed to address the deficits and ensure an improvement in service provision.

G465

A REGIONAL QUALITY IMPROVEMENT PROJECT FOR CONGENITAL CYTOMEGALOVIRUS ASSOCIATED SENSORINEURAL HEARING LOSS

^{1,2}T Holland Brown, ³FE Walston, ⁴KEM McDewitt. ¹Department of Community Paediatrics and Child Health, Cambridgeshire Community Services, Cambridge, UK; ²Department of Research and Development, Cambridgeshire Community Services, Cambridge, UK; ³Department of Neonatology, Norfolk and Norwich University Hospitals, NHS Foundation Trust, Norfolk, UK; ⁴Department of General Paediatrics, North West Anglia NHS Foundation Trust, Peterborough, UK

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Aims Treating congenital Cytomegalovirus (cCMV) associated sensorineural hearing loss with ganciclovir has been shown to improve hearing outcomes.^{1,2} This project was set up to introduce a region-wide approach to testing, diagnosing and offering treatment to infants with cCMV associated sensorineural hearing loss before 28 days of life.

Methods A region wide study day on cCMV associated hearing loss was organised to which all paediatricians, hearing screeners, audiologists, ENT surgeons and virologists at the region's hospitals were invited. Speakers included international researchers in the area of cCMV, parents of children with hearing loss and clinicians who had set up a cCMV pathway elsewhere. Following this, a subgroup of interested clinicians, laboratory scientists and screeners was set up to investigate the potential for a region wide approach. The group met three times and was truly multi-disciplinary. Between meetings different multi-disciplinary groups worked on elements of the pathway. Beyond those involved in the subgroup, other clinicians and specialists such as ophthalmologists, radiologists and pharmacists contributed to the final pathway.

Results a) audits confirmed that, prior to implementation, cCMV was diagnosed too late, if at all, for ganciclovir treatment b) the regional virology services introduced CMV PCR testing on salivary samples with appropriate validation c) hearing screeners confirmed the acceptability of screeners taking salivary samples d) a generic regional pathway was introduced allowing for local adaptation and implementation

Conclusions Truly multi-disciplinary, cross-region working allows for rapid design and implementation of a project in a consistent and timely manner. Although clinician led, the regional hearing screening teams were instrumental in getting the project off the ground.

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George Still Forum (National Paediatric ADHD Network)

G466

THE NEED AND RISE OF ADHD SERVICE IN A DGH SETTING

S Nelapatla, V Pulla. *Paediatrics, Northern Lincolnshire and Goole NHS Foundation Trust, Scunthorpe, UK*

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Aim There are no national or regional models of care...in CAMHS or Paediatrics.

To share our experience of our model of care in setting up and running ADHD service (fully compliant with NICE guidelines) for nearly 20 years, actively managing about 200 children per year in a borough with child population of 40 000.

We will be presenting care pathway, customised templates, assessments, monitoring, resources, stake holder involvement, specialist nurse role, specialist and speciality trainee involvement and transition arrangements to adult care.

Conclusion Mental Health – The new epidemic needs novel ways and models of care.

Our model demonstrates collaborative working, efficient use of limited NHS resources in ever increasing demand on Paediatric and CAMHS services.

G467

MANAGEMENT OF SLEEP DIFFICULTIES AMONG A COHORT OF CHILDREN WITH ADHD IN A SCOTTISH LOCAL AUTHORITY

MO Ogundele. *Community Paediatrics Unit, NHS Fife, Glenwood Health Centre, Glenrothes, UK*

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An estimated 25%–50% of children and adolescents with ADHD are known to experience problems with sleep, which is approximately five-fold that of healthy controls. ADHD-related sleep problems include high rates of daytime sleepiness, increased risk of sleep disordered breathing (50% vs 22% in controls), restless legs syndrome and periodic leg movement disorder. Sleep deprivation may mimic and