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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FIRST CASE OF CONSTITUTIONAL MISMATCH REPAIR DEFICIENCY SYNDROME ASSOCIATED WITH BRONCHIECTASIS

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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.

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