The European Cystic Fibrosis Society (ECFS) Neonatal Screening Working Group (2010) developed a framework for the early management of infants during the first year of life.²

One of the many recommendations of this framework is that newly diagnosed infants should receive *frequent monitoring* to ensure good clinical outcomes.

Objective The nurse-led MDT infant clinic was introduced in Tallaght Hospital, Dublin in July 2011 following the introduction of newborn screening for CF in Ireland.

It was introduced to ensure parents/families continued to receive an individualised quality service, with ongoing education and support following the diagnosis of their infant.

The clinic is held once, or twice weekly and the frequency of infant visits is dependent on clinical need.

The clinic is led by a senior CF clinical nurse specialist (CNS) and infants are reviewed by senior members of the CF physiotherapy, dietetic departments & other MDT members, as required.

Methods A retrospective review of the healthcare records & MDT records of all infants diagnosed with CF over a 7.5 year period (1st July 2011– 1st January 2019) was made and information was captured on a data collection form.

Results 36 infants were diagnosed in this period.

A total of 141 appointments were attended with each infant having an average of 4 appointments (R 2–11) prior to their first Consultant-led MDT CF clinic.

A wide range of actions/interventions were carried out at the clinic and parents welcomed the regular return visits.

Conclusion The nurse -led infant clinic in Tallaght Hospital enables regular assessment of newly diagnosed infants -up to the age of 6 months, and provides ongoing parental education and support during this critical period.

The success of this clinic is dependent on having experienced CF team members who are competent to make clinical decisions and treatment changes, as required.

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MANAGEMENT OF CONGENITAL PULMONARY AIRWAY MALFORMATIONS IN A TERTIARY HOSPITAL IN IRELAND – ARE WE FOLLOWING THE RULES?

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Background Congenital Pulmonary Airway Malformations (CPAM) is a congenital disorder in which there is replacement of a portion of lung by non-functioning cystic section of abnormal tissue. Majority of CPAMs are asymptomatic at birth. The aim of this project was to review the current management of CPAM in a tertiary hospital in Ireland.

Methods Retrospective review of cases of CPAM attending the respiratory outpatients department over 4 years (2013–

2017). Data was collected from electronic outpatient letters and hospital radiology system. Data was reviewed against best practice guidelines,^{1,2,3} and analyzed using descriptive methods.

Results In total 16 cases of CPAM attended the respiratory department between 2013–2017. Diagnosis was either made antenatally, in 10 cases (63%), postnatally in 3 cases and unknown in 3 cases. Evidence suggests a chest x-ray should be performed in the postnatal period, followed by a CT Thorax in the following months. In our cohort, 11 (69%) cases had a CXR in the postnatal period, 3 of which were reported normal. 14/16 cases were asymptomatic, the most common symptom being recurrent infections. All cases had a CT Thorax performed- mean timing was 11 weeks (1–56 weeks).⁸ cases had a repeat CT Thorax prior to surgery. Surgery was planned in 11(69%) cases. The mean time to surgery was 18 months (2–31 months).² cases are being followed up medically by the respiratory team with repeated imaging at scheduled intervals and 2 further cases are awaiting surgical review.

Conclusions The management in 9(56%) cases of CPAM in this tertiary hospital met the suggested guidelines in the current literature. It is recommended that children with CPAM are operated on at an early stage, ideally before 2 years of age. This was achieved in just over half of our cohort (55%). Of note, 73% of children who underwent surgery had at least two CT scans prior to surgery.

Recommendations Close follow-up in the postnatal period and early referral to tertiary surgical services for management of their lesion, given the potential future risk for infection and malignancy.²

Education of staff and family regarding the radiation risk of repeated CT imaging in young children.⁴

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IMAGING FINDINGS OF FILAMIN A MUTATION IN CHILDHOOD ONSET RESPIRATORY DISEASE

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FLNA gene on Xq28 encodes Filamin A protein. Mutation in FLNA causes a wide variety of disease including skeletal dysplasia, neuronal migration abnormality, cardiovascular malformation, intellectual disability and intestinal obstruction. Recently childhood onset chronic respiratory disease associated with a range of FLNA mutations has been recognised and reported.

In this poster, we present two further cases of pathogenic FLNA mutation associated pulmonary disease with a focus on the importance of radiology in helping to diagnose this condition and review the literature available on the topic. Previously reported cases showed that patients with FLNA mutation display characteristic findings on chest radiograph, CT thorax and MRI brain; findings that were consistent with the cases we present here.

We will present the clinical history, imaging and genetics of these infants and highlight the radiological findings that

contributed to the diagnosis of FLNA mutation including diffuse ground glass opacification and persistent interstitial changes on Chest Radiographs and CT as well as periventricular heterotopia on MRI Brain.

P549 SHOULD CHILDREN WITH DOWN SYNDROME RECEIVE PROPHYLACTIC ANTIBIOTICS TO PREVENT RECURRENT RESPIRATORY INFECTIONS?

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Introduction Trisomy 21, also known as Down syndrome (DS), is a clinical disorder where a third copy of chromosome 21 is present. Approximately 95% of DS are due to a meiotic nondisjunction, with the remaining 5% caused by chromosomal translocation or mosaicism. Children with DS are predisposed to recurrent respiratory infections due to a number of anatomical and immunological features. Our project investigated whether there is evidence supporting or refuting the use of antibiotic prophylaxis for recurrent respiratory infections in the DS population.

Methods A systematic literature review was conducted of published medical literature within the following databases: MEDLINE, Science Direct, and The Cochrane Library. A systematic search for ongoing clinical trials and guidelines/consensus statements was performed using various clinical trial registers and professional organisation websites. Search terms included 'DS', 'Trisomy 21', 'paediatric', 'respiratory infections', 'recurrent respiratory infections', 'prophylaxis' and 'antibiotics'. Systematic reviews, meta-analyses, randomised controlled trials, case-control studies and case-series were considered.

Results A systematic search revealed 0 published articles and 0 clinical trials meeting the necessary inclusion criteria. 1 guideline was found meeting our inclusion criteria; the Nottingham Guideline which outlines the role of prophylactic antibiotics in the DS population. Given the dearth of evidence in this area, we formulated a clinical trial to investigate the utility of prophylactic antibiotics for current respiratory infections in the DS population. Azithromycin was chosen as the antibiotic of choice for its anti-inflammatory and immunomodulatory properties. Primary endpoints would be the number of respiratory infections experienced over the course of the treatment period requiring a GP or ED attendance. Secondary endpoints include the severity of respiratory infections, both the number and severity of adverse events experienced over the period of the trial, along with the patient and parent/legal guardian self-reported quality of life.

Conclusion There is a current lack of evidence supporting or refuting the use of prophylactic antibiotics for recurrent respiratory infections in DS. Basic scientific studies need to be performed elucidating the role of anatomical and immunological features in predisposing children with DS to recurrent respiratory infections. Clinical trials are needed to elucidate whether prophylactic antibiotics are useful in this cohort and to investigate the optimal timing and combination of antibiotics. Guidelines are needed to support physicians in clinical decision making.

P550 PNEUMONIA IN CHILDREN: DIAGNOSTIC POSSIBILITIES IN UKRAINE

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Background Pneumonia is the third most common fatal cause for children under age five in Ukraine. The increase in the incidence of acute respiratory infections requires the determination of the role of pathogens in the etiology of pneumonia. The spectrum of etiological agents of community-acquired pneumonia (CAP) has not been studied in Ukraine.

The aim of study

To identify and improve the understanding of the causative role of infectious pathogens in etiology childhood CAP during the epidemic season.

The methods 49 children from 2 months to 16 years old with pneumonia were examined and treated at the Eurolab clinic (Kyiv) from September to January 2018/2019. The diagnosis was based on clinical examination, chest radiography, blood test, Multiplex PCRs for throat and nasals wabs, rapid influenza diagnostic tests (RIDTs).

Results Pneumonia was confirmed with a chest x-ray in 36 patients. The diagnosis was determined without radiographic findings, based on clinical symptoms (cough, localized crackles, or decreased breath sounds, fever, tachypnea) and hypoxemia in 13 children over 5 years old. Viral pneumonia was diagnosed in 31 patients (63%) with normal WBC count. It was characterized by hyperinflation with bilateral interstitial infiltrates and peribronchial cuffing on chest x-ray in 23 children. PCR tests were used in 21 children to determine etiology: *Metapneumoviruses (hMPV)* - 6; *Respiratory syncytial virus (RSV)* - 3; *Adenoviruses* - 2; *Mycoplasma pneumoniae* - 2 (confirmed by seroconversion in IgG); *Chlamydophila pneumoniae* - 6 (3 - with Ig G seroconversion); *Mycoplasma pneumoniae* and *Chlamydophila pneumoniae* - 1; *Adenovirus* and *Influenza A* - 1. In 6 children with viral pneumonia, influenza A was identified by RIDTs. Measles was the cause of pneumonia in 2 patients.

Conclusions The study suggests that viruses (primarily, hMPV, influenza and RSV) play a major role in childhood CAP. Future research is required to understand viral and bacterial colonization of the respiratory tract and the relevance of the detection of pathogens in the etiology of community-acquired pneumonia, which will reduce chest X-rays and optimize antibiotic therapy for pneumonia in children.

P551 LONG-TERM EFFECTS IN CHILDREN WHO UNDERWENT RESUSCITATION IN NEONATAL PERIOD

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Background Despite significant progress in nursing premature babies, neonatal pneumonia, along with artificial lung ventilation (ALV), is a significant factor in formation of chronic non-specific lung diseases later in life.