Emergency Department (ED). 50% (3/6) of patients had acute symptoms prior to the fatal episode. None of the patients were attending secondary/tertiary services at the time of death. 60% (3/5) had a written personalised action plan. 75% (3/4) patients had been reviewed by the GP in the previous six months for an acute exacerbation and all had been prescribed oral steroids.

Conclusion Data suggests that most patients present to ED late, in extremis and with little warning signs of severity of the attack. Identifying those at risk is difficult. Better education on recognition of symptoms and initiation of action plan is required.

REFERENCES

P544 REVIEW OF NON-INVASIVE VENTILATION SERVICE IN AN IRISH PAEDIATRIC CENTRE
A AlShahrabally*, P Greally, C Careig, M Devitt, M McDonald, B Elnazir. Tallaght University Hospital, Dublin, Ireland

Introduction Non-invasive ventilation represents a major advance in the treatment of acute respiratory failure with well-established clinical utility.

Aim This study aimed to review number, indication and adherence to NIV in an Irish paediatric centre between February 2011 and December 2018.

Methods A retrospective review of pediatric patients requiring non-invasive home ventilation was collected from 2011 to 2018. Adherence was recorded by downloads from the PAP device. ‘Adequate’ adherence was defined as ≥4 h/night for 50% of days

Results Over a period of almost 8 years (2011–2018) there were 97 pediatrics patients needed to be started on home NIV for a diversity of medical reasons. 31% were due to non-syndromic adenotonsillar hypertrophy, 30% were due to trisomy 21 with upper airway obstruction, 18% were having Prader Willi Syndrome, 15% were having neuromuscular diseases, 6% were due to obesity.

The average age was 11.5±5 years with male: female ratio 1:1.3.

The distribution of the patient according to the type of NIV were 89% on CPAP and 11% on BiPAP.

Of those 97 patients on home NIV, 50% (n=48) were non-compliant with NIV, 42% (n=41) were compliant with NIV, 8% (n=8) were stopped using NIV for different reasons like improvement after adenotonsillectomy, non-tolerance, refusal and for non-clear reasons.

The reasons for non-adherence included, 31% (n=15) were due to refusal, 27% (n=13) were due to intolerance, 17% (n=8) were due to behavioral reasons, same percentage was for the sensory reasons, 8% (n=4) were due to parental issues.

Conclusion In this Irish paediatric cohort (predominantly non-obese), the majority of our patients were on CPAP. The adherence to Bi-level was 73% whereas, it was 33% for CPAP. Adherence issues remains a major challenge in our population despite close monitoring.

P545 EPIDEMIOLOGY AND CLINICAL MANIFESTATIONS OF ACUTE VIRAL RESPIRATORY INFECTIONS IN PEDIATRIC PATIENTS IN UKRAINE
Oleksandr Volosovets, 1Sergii Krypoustov, Viktoria Khomenko*, Oksana Iemets, Tetiana Umanets. 1O.O. Bogomolets National Medical University, Kyiv, Ukraine; 2Institute of Pediatrics, Obstetrics and Gynecology named by academician O. Lukyanova of the National Academy of Medical Sciences of Ukraine, Kyiv, Ukraine

Background Acute respiratory tract infections (ARTI) in children are the leading cause of morbidity in Ukraine. The role of respiratory viruses in the clinical manifestations of ARTI in children in Ukraine has not been sufficiently studied.

The aim of study
To investigate the etiology of ARTI and compare the clinical features of different virus infections in children during the period from September 2018 to January 2019.

The methods Nasopharyngeal swabs, collected from ARTI children aged 2 months - 16 years, who received outpatient treatment or were hospitalized to Eurolab clinic (Kyiv, Ukraine) were examined. They were screened for 7 respiratory viruses using Multiplex PCRs - Respiratory Syncytial virus (RSV), Parainfluenza virus (PIV), Adenovirus (AdV), human Metapneumovirus (hMPV), Rhinovirus (RV), human Bocavirus (hBoV) and Coronavirus (CoV). Although rapid influenza diagnostic test was used.

Results Respiratory pathogens detected in 125 of the 147 (85,0%) samples. hMPV was detected in 33 children. Clinical manifestation of hMPV infection were: tracheobronchitis - 13, pneumonia - 6, obstructive bronchitis - 7, bronchiolitis - 3, rhinopharyngitis -3, laryngitis -3. Influenza A (IVA) was detected in 28 children with ARTI: tracheobronchitis - 13, pneumonia - 6, obstructive bronchitis - 2. Half of children with IVA also have symptoms of rhinopharyngitis. RV was detected in 21 children, 12 of them have symptoms of rhinopharyngitis, 3 - croup and 3 - wheezing, 2 - bronchitis and 1 - laryngitis. Clinical characteristics of others viruses are following: RSV was detected in 10 children, it caused pneumonia (3 cases), obstructive bronchitis (5 cases). hBoV was detected in 7 children and caused rhinopharyngitis, laryngitis (6 cases), tracheobronchitis (2), two child had viral exanthema. AdV was detected in 5 children and caused rhinopharyngitis with lymphadenopathy in 3 cases, pneumonia in 2 cases. PIV during season caused rhinopharyngitis, laryngitis (2 cases), croup (1 case), obstructive bronchitis (1 case). The coinfection percentage was 13, 5%.

Conclusions During epidemic season in Ukraine the most prevalent viruses were hMPV -33 (26,3%), IVA -28 (22,4%), RV - 21 (16,8%). Using Multiplex PCR assay can be helpful in prognosing of probable clinical course of disease, for optimization therapy.

P546 A REVIEW OF THE NURSE-LED MDT CYSTIC FIBROSIS (CF) INFANT CLINIC IN TALLAGHT HOSPITAL
Gerardine Leen*, Geraldine Connell, Basil Elnazir, Peter Greally. National Childrens Hospital, Dublin, Ireland

Background Newborn screening (NBS) for CF offers the opportunity for early intervention and improved outcomes.

A 1:1.3. 

The distribution of the patient according to the type of NIV were 89% on CPAP and 11% on BiPAP.

Of those 97 patients on home NIV, 50% (n=48) were non-compliant with NIV, 42% (n=41) were compliant with NIV, 8% (n=8) were stopped using NIV for different reasons like improvement after adenotonsillectomy, non-tolerance, refusal and for non-clear reasons.

The reasons for non-adherence included, 31% (n=15) were due to refusal, 27% (n=13) were due to intolerance, 17% (n=8) were due to behavioral reasons, same percentage was for the sensory reasons, 8% (n=4) were due to parental issues.

Conclusion In this Irish paediatric cohort (predominantly non-obese), the majority of our patients were on CPAP. The adherence to Bi-level was 73% whereas, it was 33% for CPAP. Adherence issues remains a major challenge in our population despite close monitoring.
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.

REFERENCES

P547 MANAGEMENT OF CONGENITAL PULMONARY AIRWAY MALFORMATIONS IN A TERTIARY HOSPITAL IN IRELAND – ARE WE FOLLOWING THE RULES?
Rebecca Finnegan*, Oneza Almaren. Department of Respiratory, Childrens University Hospital, Temple Street, Dublin, Ireland

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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.3

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

P548 IMAGING FINDINGS OF FILAMIN A MUTATION IN CHILDHOOD ONSET RESPIRATORY DISEASE
Ann T Foran*, Enna Sasaki, William Reardon, Angela T Byrne. Our Lady’s Children’s Hospital, Dublin, Ireland

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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