

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