limits, a known low-grade metastasis disease in papillary thyroid cancer, a chest X-ray undergone and the description in the scientific letter of iodine-131 uptake in focal bronchiectasis mimicking metastatic thyroid cancer induced to monitor clinical status without further investigations. Her serum TG remained undetectable at six-month follow-up and I-131 whole-body at one-year follow-up showed the same lung focal hot spots. A chest computed tomography scan was performed confirming the presence of bronchiectasis involving both lung fields without evidence of pulmonary metastasis. Abrupt onset of radioactive iodine is present not only in known inflammatory disease of the lung, aspergilloma and respiratory bronchiolitis but false positive iodine uptake can be present also in case of pulmonary involvement in CF.

**P68**

**PHARYNGEAL SINUS MUCOCELE IN A CYSTIC FIBROSIS CHILD**

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A six years old child affected with cystic fibrosis presented to the ENT examination with a history of naso-obstruction and two episodes of intense headache. A nasal flexible endoscopy was performed showing a bulging of the all superior wall of the nasal cavity, a displacement of the middle turbinate’s root and a reduction of the choana’s size, due to the mass occupying space. The CT scan showed a homogeneous mass involving entirely the sphenoidal sinus, with an enlargement of the whole cavity, the posterior portion of the ethmoidal sinuses, the superior and the middle portion of the nasal cavity, and a partial portion of the choanal lamina. Enhanced MRI was performed excluding a vascular pattern of the lesion. Thus, the radiological features of these examinations suggested a big sphenoidal mucocele. The patient was treated with antibiotics prophylaxis i.v. before the surgical treatment. An endoscopic approach (FESS) in achieving marsupialization and drainage of the sphenoidal mucocele was carried out. A sharp removal of anterior and inferior bone walls was performed and a gentle suction of its content of suppressing thick secretions was actuated. The mucocele within the mucocele cavity was sterile at Gram staining investigation. The follow up included a clinical evaluation with iotrofiboscopy at 1–3–6 months and an MRI at 6 months. The marsupialization and drainage of the mucocele is believed to be the mainstay of treatment and the endoscopic treatment seems to be the most successful tool in carrying out this type of surgical management, according with the authors’ opinion that a mini-invasive treatment is the best choice for mucoceles in children and young adults, where an incision of an external approach can bring to cosmetic distortions. Moreover this approach results in lower morbidity and complications than external unmodified and the description in the scientific literature of iodine-131 uptake in CF for neoplastic pulmonary involvement, followed by positive SPECT/CT test (103mIq/g) and extensive molecular analysis of the CFTR gene showing a compound heterozygosity (R334W/211Sdel4). A CT scan was performed in the neonatal period and it revealed a left pulmonary sequestration. The CF diagnosis induced a more prudential approach and surgery was delayed in favour of a more conservative treatment as a first choice.

At 1 year of age, a CT angiography displayed intralobar pulmonary sequestration, receiving blood supply from a large vessel originating from descending aorta. The infant was in good nutritional status, with no respiratory symptoms but occasional rhinitis. In regular chest physiscopy with PEI-mask, presenting no bacterial respiratory infection. At 16 months of age, coil embolization of the abnormal feeding artery was successfully performed, obtaining a 80–90% loss of vascularization. At 30 months of age the infant is in good clinical conditions, with normal growth and no respiratory symptoms.

The case is presented for the rarity of the association of the two pathologies (lung malformation and CF) and because of the concern in the decision-making process regarding treatment. Chest surgery in a CF patient should be avoided, if not strictly needed; thus embolization was considered as the choice treatment in our case.

**P70**

**METHICILLIN-RESISTANT STAPHYLOCOCCUS AUREUS IN CYSTIC FIBROSIS**

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**Background:** Methicillin-resistant Staphylococcus aureus (MRSA) is an increasing problem in both the general population and cystic fibrosis (CF) patients. MRSA is recognized as a bacterial pathogen in CF patients, although its clinical effects can be variable. The isolation of MRSA in the sputum of patients with CF had questionable significance; in many instances, it was considered to be related to colonization rather than being directly related to deterioration in the course of pulmonary disease.

**Aim:** To determine determine whether MRSA infection has a deleterious effect on the clinical status of patients with CF.

**Methods:** Patients with MRSA in respiratory cultures during a three year period, September 2004-September 2007, were identified and compared with controls matched for age, sex, and respiratory function. Respiratory function tests, anthropometric data, symptoms and clinical signs, chest x-ray score were considered in the evaluation of the two groups.

**Results:** From a clinic population of 86, 32 CF patients (22 boys, 10 girls, median age 14 years) had positive sputum or cough swab cultures for MRSA. Thirty-three age-sex-matched MRSA-negative controls were identified. Only 2 of 32 (6.3%) MRSA-positive patients showed chronic colonization. In 30 of 32 (93.7%) patients who were not receiving specific antibiotic therapy MRSA has been eradicated. Prior to isolation infection 27/30 (90%) MRSA-positive CF patients received more antibiotic treatments (oral quinolones in 70% of cases, oral cotrimoxazole in 51%, intravenous aminoglycosides in 37%, oral quinolones associated with nebulized tobramicin in 7%, oral/intravenous cephalosporins in 6% of children). Three of 30 (10%) MRSA-positive patients were treated with no antibiotic therapies before MRSA acquisition. There were no significant differences between the two groups with respect to change in weight, body mass index, decline in chest x-ray appearance, pulmonary function.

**Conclusion:** MRSA infection in children with CF does not significantly affect respiratory function; acquisition did not appear to directly affect the course of the pulmonary disease in these patients, even though no patient received any treatment for their MRSA. MRSA decolonization can be successful in a high proportion of CF patients without resorting to antibiotic protocols. Although more studies are needed to evaluate the full significance of MRSA colonization among patients with CF, a judicious use of antibiotics and a segregation of in and out patients should lead to atelectasis resolution without the use of invasive approaches.

**P69**

**PULMONARY MALFORMATION AND CYSTIC FIBROSIS (CF): WHICH DIAGNOSIS, HOW TO TREAT AND TIMING OF PROCEDURE (CASE REPORT)**

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Pulmonary malformations are comprised in the spectrum of CCAM including bronchial cysts and pulmonary sequestration. They all can complicate with repeated infections, abscesses, or hemothorax, therefore elective removal is desirable as soon as possible. In case of sequestration the resolution can be achieved also with simple embolization of the feeding artery. Concern about diagnosis, timing and modality of treatment can arise from associated pathology. Case report: CC, female, born from healthy non-consanguineous parents, had a prenatal diagnosis of CCAM in the left lung (magnetic resonance at 22 weeks). Then surgery was scheduled soon after birth. She subsequently received a diagnosis of CF for neonatal hypotonia, rhinorrhea, followed by positive SPUT/CT test (103mIq/g) and extensive molecular analysis of the CFTR gene showing a compound heterozygosity (R334W/211Sdel4). A CT scan was performed in the neonatal period and it revealed a left pulmonary sequestration. The CF diagnosis induced a more prudential approach and surgery was delayed in favour of a more conservative treatment as a first choice.

At 1 year of age, a CT angiography displayed intralobar pulmonary sequestration, receiving blood supply from a large vessel originating from descending aorta. The infant was in good nutritional status, with no respiratory symptoms but occasional rhinitis. She is on regular chest physiscopy with PEI-mask, presenting no bacterial respiratory infection. At 16 months of age, coil embolization of the abnormal feeding artery was successfully performed, obtaining a 80–90% loss of vascularization. At 30 months of age the infant is in good clinical conditions, with normal growth and no respiratory symptoms.

The case is presented for the rarity of the association of the two pathologies (lung malformation and CF) and because of the concern in the decision-making process regarding treatment. Chest surgery in a CF patient should be avoided, if not strictly needed; thus embolization was considered as the choice treatment in our case.
be the key of any strategies designed to reduce the risk of MRSA acquisition by patients with CF.

**P71** CF NEWBORN SCREENING AND ITS PREVENTIVE EFFECT: 7 YEARS EXPERIENCE IN THE UMBRIA REGION

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In the Umbria Region the CF newborn screening (NBS) was carried out since the year 2000. The tests were run in the “CRI Laboratory” in Rome using the IRT (OLAA3) and PAP protocol [1]. For all positive subjects more detailed tests were suggested in order (a) to find mutations not contained in the OLAA3 panel, (b) to study the available family members to find CF traits and at risk couples. The whole CF gene was studied by DGGF analysis and by sequencing in patients with positive or borderline sweat test. In the 7 years 15 CF patients out of 50,941 newborns were found (1:3465), 8 with the classic and 7 with the mild form. The IRTI and OLA positive cases for at least one mutation or IRT2 positive cases identified were 114: 79 (69%) of them did not come to our observation. Of the 35 examined subjects 13 were found to be CF affected (6 classic and 7 mild) and 22 were heterozygotes. In 6 (4 classic and 2 mild) out of 13 CF patients the genotype was already known by the OLA panel while in 5 CF patients (1 classic and 4 mild) only one mutation was detected and in 2 CF patients (1 classic and 1 mild) the OLA panel was negative. The other 2 patients had a clinical diagnosis as the IRT test was negative: they showed a classic CF and the diagnosis was confirmed by sweat test and identification of the CFTR gene mutations. The IRT assay, even with the 63% of false positive results, was very useful in the identification of the CFTR gene mutations in both the homozygous and heterozygous subjects. In our experience the IRT positive cases gave us the possibility to perform the appropriate genetic counselling followed by the cascade screening and 6 new at risk couples and 2 atypical CF were detected. On the contrary the IRT test failed to detect 2 out of 8 classical CF and the OLA in our population was able to find 65.38% of the mutations (17 out of 26) in 13 patients with the IRT positive test. Our data suggest that the NBS for CF has a great potentiality in the prospective prevention by means of the genetic counselling followed by cascade screening. The need exists for a better knowledge of the prevention power of the NBS program by the medical staff and by the families of the screened subjects. Even if in the last few years the number of the people coming to have more detailed genetic analysis after a positive NBS test is increasing, we think that more attention has to be spent to suggest further investigation for the maximum preventive effect.

**Reference(s)**


**P72** MEASUREMENT OF PANCREATITIS-ASSOCIATED PROTEIN IN COMBINATION TO IMMUNOREACTIVE TRYPsinOGEn FOR NEONATAL SCREENING STRATEGY OF CF: A MULTICENTER FEASIBILITY STUDY

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The most widespread strategy of neonatal screening for CF consists in the measurement of immunoreactive trypsinogen (IRT) from dried blood spots coupled with CFTR mutation analysis. However, this strategy is complicated by the high cost of DNA analysis, the need for an informed consent and of genetic counselling. Moreover, the false positive rate of the IRT assay is quite high (1–2%) resulting in too many unnecessary DNA tests. Thus, alternative markers are being investigated to avoid these drawbacks. Pancreatitis-associated protein (PAP) is a stress protein synthesized by the diseased pancreas, shown to be elevated in newborns with CF too many unnecessary DNA tests. Thus, alternative markers are being investigated to avoid these drawbacks. Pancreatitis-associated protein (PAP) is a stress protein synthesized by the diseased pancreas, shown to be elevated in newborns with CF.

The tests were run in the “CRI Laboratory” in Rome using the IRT (OLAA3) and PAP protocol [1]. For all positive subjects more detailed tests were suggested in order (a) to find mutations not contained in the OLAA3 panel, (b) to study the available family members to find CF traits and at risk couples. The whole CF gene was studied by DGGF analysis and by sequencing in patients with positive or borderline sweat test. In the 7 years 15 CF patients out of 50,941 newborns were found (1:3465), 8 with the classic and 7 with the mild form. The IRTI and OLA positive cases for at least one mutation or IRT2 positive cases identified were 114: 79 (69%) of them did not come to our observation. Of the 35 examined subjects 13 were found to be CF affected (6 classic and 7 mild) and 22 were heterozygotes. In 6 (4 classic and 2 mild) out of 13 CF patients the genotype was already known by the OLA panel while in 5 CF patients (1 classic and 4 mild) only one mutation was detected and in 2 CF patients (1 classic and 1 mild) the OLA panel was negative. The other 2 patients had a clinical diagnosis as the IRT test was negative: they showed a classic CF and the diagnosis was confirmed by sweat test and identification of the CFTR gene mutations. The IRT assay, even with the 63% of false positive results, was very useful in the identification of the CFTR gene mutations in both the homozygous and heterozygous subjects. In our experience the IRT positive cases gave us the possibility to perform the appropriate genetic counselling followed by the cascade screening and 6 new at risk couples and 2 atypical CF were detected. On the contrary the IRT test failed to detect 2 out of 8 classical CF and the OLA in our population was able to find 65.38% of the mutations (17 out of 26) in 13 patients with the IRT positive test. Our data suggest that the NBS for CF has a great potentiality in the prospective prevention by means of the genetic counselling followed by cascade screening. The need exists for a better knowledge of the prevention power of the NBS program by the medical staff and by the families of the screened subjects. Even if in the last few years the number of the people coming to have more detailed genetic analysis after a positive NBS test is increasing, we think that more attention has to be spent to suggest further investigation for the maximum preventive effect.

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