The residual AHI of patients on therapy was on average 2.32/hour (min: 0 and max: 12.5). Of these 44 were in CPAP mode, 179 in Auto-CPAP, 2 in NIV, another 2 in auto-bilevel) and 1 in Servoventilation. The Auto-CPAP Pressure in 95% of the nights (P95) averaged was 12.7 cm of H2O, with a minimum of 6 and a maximum of 15 cm of H2O. In June 2021, at 3 months 30 patients, at 6 months 56 patients , at 12 months 59 and at 24 months 83 patients were included, with the average daily use being, respectively: 4.30, 5.18, 5.40 and 5.47 hours. The mean residual AHI was respectively: 2.09, 1.81, 2.27 and 2.65/hour. We achieved a good adaptation to therapy in 209 patients (91%) and 19 dropped out of therapy.
Conclusions: In patients with a confirmed diagnosis of OSA treated by PAP, the use of telemonitoring should be considered to improve follow-up and therapeutic adjustment, promoting better compliance with the therapy. In this study, Airview® was considered a useful tool, capable of preventing early dropouts , allowed us to understand its influence on the control of residual events and promoting its particular usefulness during a pandemic, often avoiding the need for health professionals to travel to patients homes, without compromising their adherence.

***Keywords**: Telemonitoring. OSA. Airview®.*

PE 030. THE CHALLENGING DIAGNOSIS OF A PATIENT WITH A VERY SEVERE LUNG OBSTRUCTION

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Case report: 36-year-old man, non-smoker, works as a green meat cutter. Episodes of coughing and wheezing in childhood, with asymptomatic period until 2009, when he reported having persistent productive cough, wheezing, exertional dyspnea and chest tightness, with worsening of complaints during the night and morning. After worsening of these symptoms, he was sent to a Pulmonology consultation. He had been medicated for the last 5 years with fluticasone 250 μg and salmeterol 50 μg, with moderate improvement of symptoms. The pulmonary auscultation had a globally diminished ventricular murmur, with crackles in the lower 1/3 bilaterally. He had peripheral oxygen saturation of 91%, and the arterial blood gas analysis revealed a global respiratory failure. Further diagnostic studies identified a very severe lung obstruction with hyperinflation, and a slight decrease in DLCO in the lung function tests. The chest CT scan revealed tubular bronchiectasis in both upper lobes and apical segments of the lower lobes, with thickened walls and mucoid impaction, in addition to centrilobular micronodules with a diffuse bilateral mosaic pattern and mediastinal adenopathies, the largest subcarinal measuring 14 mm. The analytical and autoimmune study only revealed a slight increase in eosinophils. In bacteriological examination of sputum, a multisensitive Pseudomonas Aeruginosa was isolated, and an eradication cycle was prescribed with ciprofloxacin for 21 days. After showing little symptomatic improvement in later evaluations, a sweat test was requested, with NaCl values indicating a probable cystic fibrosis (114 mEq/L NaCl). It was requested a genetic study, with the panel of mutations for the CTFR gene identifying heterozygous genes for c.1521_1523delCTT (formerly F508del) and c. 254G>A (formerly G85E), which confirmed the diagnosis of cystic fibrosis. The patient was later sent for a specialized cystic fibrosis consultation at the Hospital de Santa Maria.
Discussion: The typical form of cystic fibrosis is diagnosed early in life and diagnosis at a later age is often associated with mild lung disease. A high index of suspicion is needed to make the diagnosis, as older patients with cystic fibrosis can appear to be well and have symptoms similar to other diseases. With advances in gene detection, adult cystic fibrosis diagnoses are increasing. A timely diagnosis of cystic fibrosis is important, as it has prognostic and treatment implications, in addition to being able to lead to genetic counseling in families.

***Keywords**: Cystic fibrosis. Lung obstruction. Genetic study.*

PE 031. ACCESS TO PULMONARY REHABILITATION: PERSPECTIVES OF PATIENTS, LOVED ONES AND HEALTHCARE PROFESSIONALS

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Introduction: Improving access to pulmonary rehabilitation (PR) is an international priority, but due to several constraints, healthcare professionals are often faced with the challenge of having to prioritise patients. Evidence from quantitative research suggests that symptoms, functional and health status and not lung function should guide referrals to pulmonary rehabilitation (PR). Whether these criteria are corroborated by the opinions of different stakeholders remains unknown. This qualitative study explored criteria, barriers and facilitators to access PR from the perspectives of people with chronic respiratory disease (CRD), loved ones and healthcare professionals.
Methods: An exploratory, cross-sectional qualitative study was carried out. Focus groups were conducted separately with people with CRD, loved ones and healthcare professionals; transcribed verbatim and analysed thematically. All participants had previous experience with PR.
Results: Seven focus groups were conducted: four with people with CRD (24 with chronic obstructive pulmonary disease and 5 with interstitial lung disease, 75.9% male, 68.4 ± 7.5 years); one with loved ones (n = 5, 100% female, 66.6 ± 7.7 years) and two with healthcare professionals (n = 16, 25% male, 38 ± 9.2 years). Perspectives among stakeholders were mostly consensual and organised in three themes: all people with CRD should have access to PR and as early as possible "Universal access"; if prioritisation is needed then priority should be given to those motivated, with high symptom burden and impaired functional status "Priority to those struggling and motivated"; and education about PR and continuity and communication between care settings and professionals are lacking to improve access to PR "Communication, dissemination and organisation as main keys".
Conclusions: Our findings corroborate previous evidence and provide new and complementary in-depth understanding to design interventions to improve access to PR in line with the perspectives of different stakeholders.

***Keywords**: Access to pulmonary rehabilitation. Chronic respiratory disease. Informal caregivers. Qualitative methods.*

PE 032. MEASUREMENT TOOLS TO ASSESS EDUCATION AND PSYCHOSOCIAL SUPPORT OF PULMONARY REHABILITATION IN PEOPLE WITH CHRONIC OBSTRUCTIVE PULMONARY DISEASE – A SYSTEMATIC LITERATURE REVIEW

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Introduction and objectives: Education and psychosocial support is a core component of pulmonary rehabilitation (PR). Nevertheless, measurement tools used to assess the effects of this component of PR have been scarcely investigated. Thus, this systematic literature review aimed to identify which measurement tools have been used to assess education and psychosocial support of PR in people with chronic obstructive pulmonary disease (COPD.)
Methods: A systematic search was conducted on PubMed, Scopus and Web of Science in February 2021. Articles were screened and inclu-

sion decided by two independent researchers. Randomised control studies, quasi-experimental and pre-post studies, which included people with COPD enrolled in a PR programme, with exercise and education and psychosocial support, were included. The same two independent researchers extracted the data to a standardised table.
Results: A total of 7112 studies were screened and 9 studies were included. 1121 people with COPD (68.69 ± 8.98 years old, 30.78% female, FEV1pp 53.25 ± 19.04%) were enrolled in the included studies. A total of 9 measurement tools were identified and the most reported was the Bristol COPD Knowledge Questionnaire (BCKQ) (n = 3, 33.3%). Other measurement tools were found, such as the Patient Activation Measure (PAM) (n = 1, 11.1%), the Education Specific Questionnaire (ESQ) (n = 1, 11.1%), the Lung Information Needs Questionnaire (LINQ) (n = 1, 11.1%), the COPD Knowledge Test (n = 1, 11.1%), the Pulmonary Disease Knowledge Test (PDKT) (n = 1, 11.1%), the Health Education Impact Questionnaire (heiQ) (n = 1, 11.1%), the Understanding COPD Questionnaire (UCOPD) (n = 1, 11.1%) and the Knowledge Assessment Questionnaire (n = 1, 11.1%).
Conclusions: This review has shown that there are nine measurement tools available to assess the effects of education and psychosocial support of PR in people with COPD. The most reported has been the BCKQ however, few studies assessing this component in PR programmes have been conducted. Given the importance of education and psychosocial support of PR for people with COPD, future research is urgently needed to provide recommendations on the most suitable measurement tools and promote its routine assessment.

Keywords: *Respiratory rehabilitation. COPD. Education program. Psychosocial intervention.*

PE 033. LEIRIA´S MUTATION

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Alpha-1 antitrypsin (AAT) deficiency is an autosomal codominant disease caused by multiple mutations in the SERPINA 1 gene. Belongs to the group of Protase Inhibitors (PI) and Pi*MM corresponds to homozygosity of the normal gene. The deficit appears in adults, essentially, with Chronic Obstructive Pulmonary Disease (COPD) and Emphysema in adults and these changes are potentiated by smoking. The minimum serum concentration considered protective is 57 mg/dl by nephelometry and 80 mg/dl by radial immunodiffusion. To date, more than 120 mutations in SERPINA1 are described in the literature. Female, 46 years old, ex-smoker (37 PPY). No known family history. In a Pulmonology consultation since 2012 for Asthma and COPD and alpha-1 antitrypsin deficiency, referred to Leiria Hospital for change of residence. Medicated with Formoterol, Salbutamol, Ipratropium Bromide and Aminophylline with a moderate/severe annual flare-up. In consultation, had complaints of dyspnea (mMRC2) and recurrent wheezing. Respiratory function tests showed very severe obstruction (FEV1 30%; FEV1/FVC 45%; DLCO 40%) with hypoxemia (pO2 66 mmHg). Chest computed tomography showed centrilobular and panlobular emphysema with greater expression in the lower lobes. Serum AAT concentration was < 5 mg/dl and the phenotype study Pi*MZ. As there appeared to be some disagreement between the patient's symptoms and the AAT levels, a genotypic study was continued which showed a ZQ0L mutation. The allele targeted by Q0L corresponded to a substitution of glutamic acid for lysine at position 342 of the protein. It is a severe deficiency allele associated with intracellular accumulation in the endoplasmic reticulum and reduced protein secretion by about 80%. It corresponds to a new variant characterized by the occurrence of a Glu (GAG) -Stop (TAG) mutation in an M3 base allele. This mutation is not described in the literature.

Keywords: *Alpha-1 antitrypsin. Mutation.*

PE 034. ALPHA-1 ANTITRYPSIN DEFICIENCY AND PREGNANCY – A CASE REPORT

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Introduction: Alpha-1 antitrypsin deficiency (A1AD) is the most recognized genetic cause of chronic obstructive pulmonary disease (COPD). There is some evidence about the potential role of alpha-1 antitrypsin (AAT) in the regulation of the immune system and in gestation. A severe deficit may lead to a decline in maternal lung function and fetal growth impairment. Few cases of pregnancy in women with A1AD were reported, but there is some evidence pointing to a higher abortion rate, preterm labor, and worse maternal health related outcomes.
Case report: The authors present the case of a 39-year-old primigravida, previous smoker, with a history of severe A1AD (ZZ genotype) and COPD with severe flow obstruction. At the time of diagnosis, she was medicated with inhaled bronchodilators and corticosteroid. She had serum levels of AAT of 15 mg/dL and forced expiratory volume in the first second (FEV1) of 990 ml (less than 40% of predicted). At 30 weeks of gestation she complained of increased shortness of breath. Lung function tests and arterial blood gas were stable. 24-hour oximetry showed desaturation (~85%) with exertion and in the nocturnal period, so supplementary oxygen (O2) was initiated. At 33 weeks of gestation she was admitted to the Obstetrics ward due to perception of diminished fetal movements and fetal growth restriction. Polysomnographic study showed mild sleep apnea (AHI = 13.7/h); treatment with APAP was initiated, with partial improvement of nocturnal hypoxemia, so supplemental O2 was added. Transthoracic echocardiogram was normal. Due to fetal and maternal risk, an elective cesarean section was performed at 36 weeks, with no reported complications. The newborn presented with low birth weight (2170 g), and an Apgar score of 9/10/10 regarding the 1st, 5th and 10th minutes, respectively, with normal blood gas. Shortly after delivery the patient presented with mild clinical deterioration, but additional therapy was not needed. She was discharged with an O2 prescription for exertion and APAP with O2 supplement. In the following Obstetric and Pulmonology appointments she showed progressive improvement of her respiratory complaints, and lung function was overall stable. The patient is currently undergoing further evaluation of her respiratory disease. She is awaiting approval for AAT augmentation therapy and was referred to a respiratory rehabilitation program.
Discussion: This case illustrates the difficulties of managing an uncommon combination of pregnancy in a patient with severe A1AD and functional respiratory impairment. Patients with A1AD in a reproductive age should receive counselling regarding potential risks associated with pregnancy and the importance of smoking cessation. Follow-up of these patients should focus on monitoring symptoms and preventing future exacerbations.

Keywords: *Alpha-1 antitrypsin deficiency. Pregnancy. COPD.*

PE 035. GENERALISED BRONCHIECTASIS AND SEVERE ALPHA-1-ANTITRYPSIN DEFICIENCY: CONCERNING A CLINICAL CASE

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Introduction: Alpha-1 antitrypsin (AAT) deficiency is the most frequent inherited abnormality in adults and a risk factor for disease in several organs, the most frequent being chronic pulmonary disorders. There are more than 100 described mutations in this gene, but the homozygous genotype of the Z allele (PiZZ) contributes to the majority of clinically identified severe deficits. Although the