P175 SINGLE CENTRE EXPERIENCE OF THE REAL-LIFE IMPACT OF PIRFENIDONE ON LUNG FUNCTION IN PATIENTS WITH IDIOPATHIC PULMONARY FIBROSIS

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Introduction Idiopathic pulmonary fibrosis (IPF) is a progressively destructive lung disease that culminates in respiratory failure and death. Trials have demonstrated that treatment of IPF patients with Pirfenidone reduces %FVC decline, improves progression-free survival and significantly reduces the risk of all-cause mortality at 1 year. Our anecdotal experience is that a small proportion of patients show improvement of %FVC with treatment.

Objectives To assess the proportion of patients in an ILD specialist centre that improve, stabilise or decline in their %FVCs on Pirfenidone treatment.

Methods In this retrospective study patients with IPF diagnosed according to the ATS/ERS guidelines at the ILD MDT, who were commenced and continued on Pirfenidone for >6 months were included. Data was derived from the clinical records of the Oxford IPF clinic.

Results 100 patient records were analysed and 31 were excluded (n = 15 <6 months' therapy, n = 5 inadequate data, n = 2 death <1 month, n = 9 other). 58 (84.1%) male, 11 (15.9%) female; 38 (55.1%) had Definite IPF, 31 (44.9%) Probable IPF.

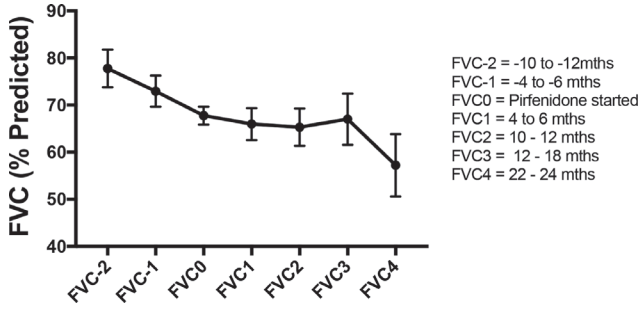
Six months after commencing Pirfenidone (n = 69 patients), 5 (7.25%) experienced significant (>10%) and 9 (13.04%) experienced marginal (5–10%) improvement in %FVC, 33 (47.83%) showed stability (−5% to 5% change %FVC), 10 (14.49%) showed marginal decline (−5% to −10%) and 12 (17.39%) showed significant (>10%) decline in %FVC.

After one year of Pirfenidone (n = 44 patients), 3 (6.8%) showed significant and 5 (11.4%) showed marginal improvement, 18 (40.9%) showed stability, 11 (25%) showed mild and 7 (15.9%) showed significant decline of %FVC.

After 2 years of treatment (n = 15 patients), 1 (6.7%) showed significant and 3 (20%) showed mild improvement, 4 (26.7%) showed stability, 3 (20%) showed mild and 4 (26.7%) showed significant decline of the %FVC.

Among 8 patients who had improvement in %FVC at one year, 6 were males, 6 had definite IPF, median age 77 years (68 – 84) and the median FVC was 73.5% predicted (66 – 79).

Conclusions Real-life use of Pirfenidone shows clear slowing of decline in the %FVC, whereas a clinically significant subset show improvement in FVC. Potentially the beneficial effect is lost after 22–24 months, although small numbers limit this analysis.



Abstract P175 Figure 1 Pre and Post- pirfenidone treatment

Paediatric Respiratory Disease

P176 DIAGNOSING ASTHMA IN CHILDREN USING SPIROMETRY: EVIDENCE FROM A BIRTH COHORT STUDY

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Background NICE draft guidance for the diagnosis of childhood asthma proposes algorithms based on four tests of lung function (FEV1/FVC ratio, bronchodilator reversibility [BDR], FeNO, PEFR variability); a minimum of two tests must be positive to make a diagnosis. For FEV1/FVC ratio, the proposed cut-off for a positive test is <70%, or the lower limit of normal (LLN), which is neither defined nor widely available. In this algorithm, spirometry is the first-line investigation, and children with FEV1/FVC > 70% are not offered BDR. However, the diagnostic test accuracy for FEV1/FVC and BDR is unknown. Within the setting of a population-based birth cohort we investigated the value of FEV1/FVC and BDR in diagnosing asthma.

Methods We assessed study participants at clinical follow-up at age 16 years using validated questionnaires and lung function measurement. Spirometry was measured according to ATS/ERS guidelines. Using the Asthma UK reference equations, we calculated LLN for FEV1/FVC. BDR was considered positive if FEV1 increased by >12% following administration of 400 mg of salbutamol. Current asthma was defined as all three of: (1) doctor-diagnosed asthma ever, (2) wheezing in the previous 12 months and (3) current use of asthma treatment. We assigned children negative to all three features as a non-asthmatic control group.

Results Spirometry was available for 630 children (325 boys, age range 13.1–16.9 years), of whom 74 (11.7%) had current asthma and 403 were assigned as non-asthmatic controls. FEV1/FVC was significantly lower among asthmatics (84.1% *vs.* 89.2%, p < 0.001, Figure 1). Ten children (1.6%) had FEV1/FVC <70% (two in asthma group). Discriminative ability of FEV1/FVC < 70% was poor (Receiver operating characteristic curve, AUC = 0.70; sensitivity = 2.7% [2/74], specificity = 98.8% [398/403]). For the calculated FEV1/FVC LLN (74.8% for boys, 78.2% for girls), 28 children (4.4%) had FEV1/FVC<LLN (11 in asthma group). Discriminative ability of FEV1/FVC<LLN was poor (sensitivity 14.9% [11/74]; specificity 97.0% [391/403]). BDR was positive in 54 children (8.7%), of whom 12 had asthma. Discriminative ability of BDR was poor (AUC = 0.64, sensitivity = 16.2% [12/74], specificity = 93.5% [373/399]). Combining these two tests did not result in a better diagnostic accuracy (sensitivity = 2.7%, specificity = 99.0%).

Conclusions FEV1/FVC < 70% or <LLN, and BDR > 12% have a poor diagnostic accuracy as tests for childhood asthma.

P177 HIGH PREVALENCE OF UNRECOGNISED ASTHMA IN CHILDREN WITH SICKLE CELL DISEASE

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Background Sickle Cell Disease (SCD) affects about 1 in 1,900 children born in the UK. Respiratory morbidity affects children as well as adults with SCD and the burden may have been