

P134 COMPARISON OF TWO PROGNOSTIC TOOLS FOR IDENTIFYING HIGH RISK PATIENTS WITH IDIOPATHIC PULMONARY FIBROSIS

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Introduction Idiopathic pulmonary fibrosis (IPF) is a progressive disease with median survival of 2–3 years. Clinical course and survival is variable. Whilst no universally accepted tool is accessible to identify those with poorer prognosis, du Bois et al⁽¹⁾ and Ley et al⁽²⁾ propose individual 4 point scoring systems to calculate mortality. Our aim was to compare both tools and assess for superiority.

Methods Data was collected on 101 patients retrospectively, 13 were excluded. The du Bois score included age, respiratory hospitalisation, baseline forced vital capacity (FVC) and 6 month change in FVC.⁽¹⁾ The GAP index used gender, age, FVC and DLCO.⁽²⁾ Predictive values were assessed using area under the receiver operator characteristic curve (AUC). We used GAP index >1 and du Bois score >22 to indicate high risk for calculation of performance characteristics. Patients were followed up for 3 years or until death.

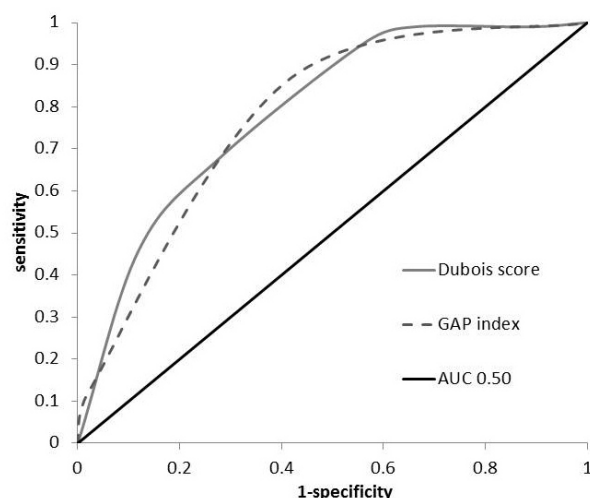
Results Median age was 69 years (interquartile range 63–76). 57.5% were male. 1 year mortality was 10.2%, compared with 18.2% at year 3. For prediction of 1 year mortality, the AUC was 0.78 (0.69–0.87) for the du Bois score and 0.73 (0.63–0.84) for the GAP index. For 3 year mortality, the AUC was 0.73 (0.65–0.80) for both systems. There was no significant difference between the scores for either outcome ($p=0.5$ and $p=0.9$ respectively). The receiver operator characteristic curves for 1 year mortality are shown: figure 1.

For 1 year mortality, the GAP index had a sensitivity of 87%, specificity 55% compared to 66.7% and 71.1% for the du Bois score. For 3 year mortality, the GAP index had a sensitivity of 86.7%, specificity 58.9% compared to 62.5% and 73.7% for the du Bois score.

Conclusions This study shows that both systems are of equal value at identifying patients with IPF at high risk of mortality.

References

1. du Bois RM et al. Ascertainment of individual risk of mortality for patients with idiopathic pulmonary fibrosis. *Am J Respir Crit Care Med* 2011; 184:459–466.
2. Ley B et al. A multidimensional index and staging system for idiopathic pulmonary fibrosis. *Ann Intern Med.* 2012; 156:684–691.



Abstract P134 Figure 1

P135 QUALITY OF LIFE AND ADVERSE EFFECTS IN PATIENTS TAKING PIRFENIDONE FOR IDIOPATHIC PULMONARY FIBROSIS

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Introduction and Objectives Pirfenidone is currently the only disease-modifying therapy for idiopathic pulmonary fibrosis. However the CAPACITY¹ study demonstrated frequent mild adverse effects. No data is available on the overall effect of pirfenidone on quality of life. Our aim was to describe patients' experience of pirfenidone, taking these factors in to account.

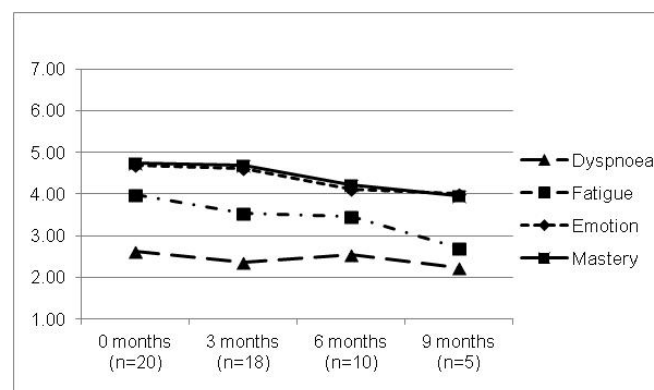
Methods This was a single-centre observational study of patients participating in the pirfenidone named patient programme. Participants completed the Self-Report Chronic Respiratory Disease Questionnaire (SR-CRQ), a quality of life assessment with four domains, at three monthly intervals. Adverse effects were also recorded. Patients were followed up until death or cessation of pirfenidone.

Results 26 patients were included, with 9 patients reaching 3 months, 6 reaching 6 months, and 5 reaching 9 months during the study period. Mean age was 67 ± 7 years, and 77% were male. There was a downward trend in all domains of the SR-CRQ, as illustrated in figure 1. However there were no significant differences in scores from baseline compared with individual time points. No relationship was found between SR-CRQ scores and adverse effects.

Adverse effects were experienced by 14 patients (54%). These included gastrointestinal symptoms ($n=12$), photosensitivity ($n=2$) and fatigue ($n=3$). 5 patients stopped pirfenidone as a result of adverse effects. The mean weight in kilograms of those patients experiencing adverse effects was lower than those who did not (72.5 ± 12.7 vs 86.1 ± 13.2 , $p=0.014$). Age in years was higher in those with adverse effects (70 ± 6 vs 64 ± 8 , $p=0.035$). Patients with more severe disease ($FVC < 50\%$, $DLCO < 35\%$) were also significantly more likely to have adverse effects ($p=0.005$).

Conclusion The data from this study is limited due to small numbers, however we have not observed a significant change in quality of life from baseline in those treated with pirfenidone. Adverse effects were greater in older, lighter patients with more severe disease.

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Abstract P135 Figure 1 Mean SR-CRQ scores in patients with at least 3 months follow-up