Will SPAR be useful in the usual patients with scleroderma?

We read with great interest the simplified predictive score ‘SPAR model’ by Wu et al.¹ A simplified predictive score which can predict ‘fast progressors’ can be used to target and selectively recruit such patients for drug trials and finally lead to improvement in outcomes in the future. However, certain aspects require clarification.

First, the external validity of the patients recruited is unclear. Although early interstitial lung disease (ILD) as defined in the study (<20% CT involvement) would be common, approximately half of the patients of this study could be having forced vital capacity (FVC) >100% (assuming normality). The latter would be a subgroup which would be uncommon. More interestingly, these patients have never been recruited for interventional studies looking at drugs on ILD—both the scleroderma lung studies (refs ² and ³) recruited patients with FVC ≤83%; thus we have no idea whether they (FVC>100%) respond to therapy. In the latter study (SLS2), only 30 out of 198 screened systemic sclerosis were excluded due to pulmonary function test, which could be higher or lower FVC and diffusing capacity of the lung for carbon monoxide-. The authors may like to provide their cohort numbers and how many of them fulfilled the inclusion criteria.

Second, the authors found ‘arthritis ever’ to be significant after multivariate analysis, though a previous study did not show any association of arthritis and ILD progression.⁴ However, there is no comment on whether arthritis was persistent and erosive, and did it require treatment in their cohort? Was baseline presence of arthritis also significant? In a patient who comes for the first time, history of arthritis would not be available.

Third, the best multivariate predictive model in this study (model 3 (SpO2≤94%+arthritis ever)) has a sensitivity of only 44%, thus more than half of the progressors would not be detected. Even in a 0–2 SPAR score, the most common score is 1, and that would only identify one-third of progressors¹!

Finally, the authors may like to provide any data on other variables expected to predict progression—baseline extent of ILD on CT (varying from 0% to 20%), oesophageal diameter on high-resolution CT (as shown by other studies² ⁵) and nail fold capillaroscopy. The latter becomes important as low oxygen saturation after the 6 min walk test, when severe ILD and pulmonary artery hypertension (PAH) are excluded (as in this study), may reflect early microvasculature changes in pulmonary bed, which as expected to be generalised and would be reflected in the nail bed capillaries also.

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