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Title: Natural history of idiopathic pulmonary fibrosis: Are slowly progressive and rapidly progressive really steady conditions?

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Body: Idiopathic pulmonary fibrosis (IPF) is a devastating lung disease with heterogeneous clinical course. Some patients experience an accelerated disease progression (rapid progressors) while other remain relatively stable over time (slow progressors). The aim was to investigate the different course of the disease in relation to survival. The study population included 55 IPF patients (age at diagnosis 53±1) categorized in rapid progressors and slow progressors by two distinct criteria: pre-diagnosis criteria (time from symptoms onset and IPF diagnosis) or by post-diagnosis criteria (decline in FVC%pr. over 12 months). When stratified by pre-diagnosis criteria 18% were rapid progressors while 66% were slow ones.When stratified according to post-diagnosis criteria 67% were rapid progressors and 33% were slow ones. The coefficient of agreement between the two criteria was 70% and 75% for slow and rapid progressors respectively indicating that up to 30%of patients did not maintain the same label. Stratification by pre-diagnosis criteria was not related to survival. Conversely, stratification by post-diagnosis criteria had a prognostic significance; indeed, rapid progressors had decreased survival as compared to slow ones (28±1Vs49±8mo.p=0.02). Of interest, rapid progressors according to post-diagnosis criteria, often display an unstable decline alternating periods of functional stability to a rapid deterioration. In conclusion our data suggest the need to be cautious in labelling IPF patients to a fixed phenotype from the beginning of symptoms till death. It is possible that IPF patients show a variable and unpredictable clinical course rather than a steady condition.