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| **Telomere Length in Australian Adults with Pulmonary Fibrosis** |
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| **Introduction/Aim:** Telomere shortening is identified in a significant number of adults with various forms of pulmonary fibrosis (PF). Telomere length less than the tenth centile for age has been associated with worse outcomes across idiopathic pulmonary fibrosis (IPF), fibrotic hypersensitivity pneumonitis (fHP) and unclassifiable interstitial lung disease (uILD). The aim of this study was to define the prevalence and impact of telomere shortening in adults referred to a tertiary Australian PF clinic. **Methods:** Telomere length was measured using an in-house Flow-FISH assay, both prospectively and retrospectively, in adult patients with PF referred to The Prince Charles Hospital, Brisbane, Australia. Demographic details and variables from the ILD multi-disciplinary meeting (MDM) were collected. Short telomere length was defined as less than the tenth centile for age.**Results:** Telomere length was measured in 212 individuals, including 136 IPF, 25 fHP and 39 uILD. One hundred and thirteen (53%) individuals had short telomeres, with similar prevalence across the three main diagnostic groups. Individuals with short telomeres tended to be younger (median age 62 years (IQR 54-69) compared with 64 years (IQR 60-71)), however were similar in terms of the presence of family history of ILD (23% versus 24%), macrocytosis (21% versus 18%) and thrombocytopenia (11% versus 8%). No difference was observed for transplant-free survival between those above and below the tenth centile for the group as a whole or within IPF. Twenty-three individuals initially diagnosed with uILD were subsequently re-classified as IPF, 15 (65%) of whom had short telomeres.**Conclusion:** Shortened telomeres were highly prevalent amongst adults with PF. While those with short telomeres were typically younger at diagnosis, there was no difference in the prevalence of features suggestive of a telomere biology disorder or outcome. Short telomere length may assist in the diagnosis of ILD where the initial MDM discussion yields an uILD diagnosis. **Key words:** Telomere, pulmonary fibrosis, Flow-FISH**Grant Support:** This project was supported by the Centre of Research Excellence in Pulmonary Fibrosis which is funded by the NHMRC (GNT1116371 and GNT2015613), Lung Foundation Australia, Boehringer Ingelheim, and anonymous philanthropy. |