6.15 Pulmonary hypertension in Hereditary Haemorrhagic Telangiectasia:Clinical characteristics and treatment patterns in Ireland

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Background: Pulmonary Hypertension (PH) is a serious complication of Haemorrhagic Telangiectasia (HHT) To define the clinical characteristics, treatment patterns and outcomes of patients with HHT referred to the Irish National PH Unit (NPHU) between 2010 and 2022. **Methods**: Patients with HHT and PH referred to the NPHU between 2010 and 2022 were included in this retrospective study. **Results:** Of the 9 HHT patients referred to the NPHU during this study period, 8 were diagnosed with PH. 7 were diagnosed with group 1 PAH and 1 was diagnosed with Group 5 PH. Five patients had a family history of HHT and genetic mutations were identified in 75%; 5 mutations in the ACVRL-1 gene and 1 in the endoglin gene. The median age at PH diagnosis was 56 years and the median pulmonary arterial pressure at diagnosis was 48mmHg. 7 subjects were treated with PH specific therapies. 4 patients were deceased at the end of the study period, with a median duration of 3 years between PH diagnosis and death. **Conclusion:** PH in HHT is frequently multifactorial and is associated with a high morbidity and mortality. We outline the clinical and treatment characteristics of this cohort in this single centre study.
Conflicts of interest: The authors have no conflicts of interest.