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Changing patterns in clinical, echocardiographic and haemodynamic characteristics, and management strategies that may be translated to improved survival in pulmonary hypertension

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Background and aims: In this retrospective analysis of a single-center series on pulmonary hypertension (PH), we aimed to evaluate clinical, echocardiographic, and hemodynamic characteristics, management strategies, morbidity, and mortality in PH subgroups across three consecutive periods.

Methods: The study included 1071 patients enrolled in the Evaluation of Pulmonary Hypertension Risk factors Associated with Survival study (EUPHRATES). In reference to enrollment time, patients were assessed in three periods ; before 2016, the era of primarily monotherapy or sequential combinations; between 2016 and 2019, post-approval and reimbursement of macitentan in our country, marking a shift towards more proactive sequential combinations; and after 2019, signifying the initiation of earlier sequential combinations with selexipag in pulmonary arterial hypertension (PAH). The ESC/ERS 2022 risk scoring, COMPERA 2.0 and REVEAL lite 2 models were used for risk predictions. Composite endpoint (CEP) definitions were adopted from the SERAPHIN and GRIPHON trials. Annual incidence curves for mortality and CEP were evaluated, but 2020 analysis was adjusted to account for the possible adverse effects of the COVID-19.

Results: During the 1st, 2nd and 3rd periods, 481, 352, and 238 patients with PH were diagnosed, respectively. Incident cases comprised 91% of them. Idiopathic PAH (IPAH), PAH-associated with congenital heart diseases (CHD-PAH), PAH-associated with connective tissue diseases (CTD-PAH), group II, III, IV and V PH were documented in 24 %, 28 %, 4.6 %, 4.3 %, 11%, 27 % and 0.7 % of patients, respectively. Age and female dominance remained unchanged. Incidences of Group 1 PH and CHD-PAH decreased while IPAH, drug-associated PAH, and Group II and IV PH increased across three periods (p-values varying from <0.001 to 0.03). Although baseline functional class became worse (p=0.013), six-minute walk distance, N-terminal pro brain natriuretic peptide and risk scores showed no change (p=NS). Baseline echo and hemodynamic measures became better, and triple combinations increased after 2019 (p<0.001 for all). For overall PH, rates of all-cause mortality and CEPs at 1st, 2nd and 3rd periods were 52 % and 59 %, 30 % and 35 %, and 11 % and 13 %, respectively (p< 0.001, for all). Mortality rates for 1st, 2nd and 3rd periods were 36 %, 38 % and 26 % in IPAH, 60 %, 28 % and 13 % in CHD-PAH, 55 %, 22% and 22% in CTD-PAH, 38 %, 38% and 24 % in group IV PH, respectively (p< 0.001, for all). Mortality curves showed sudden increases in 2020, but excluding 2020, a steady improvement in the annual mortality was evident.

Conclusions: Our findings suggest a trend towards better clinical, echocardiographic, and hemodynamic presentations and improved survival in the PAH subgroups, and Group IV PH across the three periods that might be attributed to more proactive management strategies favouring earlier initiation of combinations.