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Medical therapy and mortality in pulmonary arterial hypertension associated with congenital heart disease: Data from the Hellenic pulmonary hypertension registry (HOPE)

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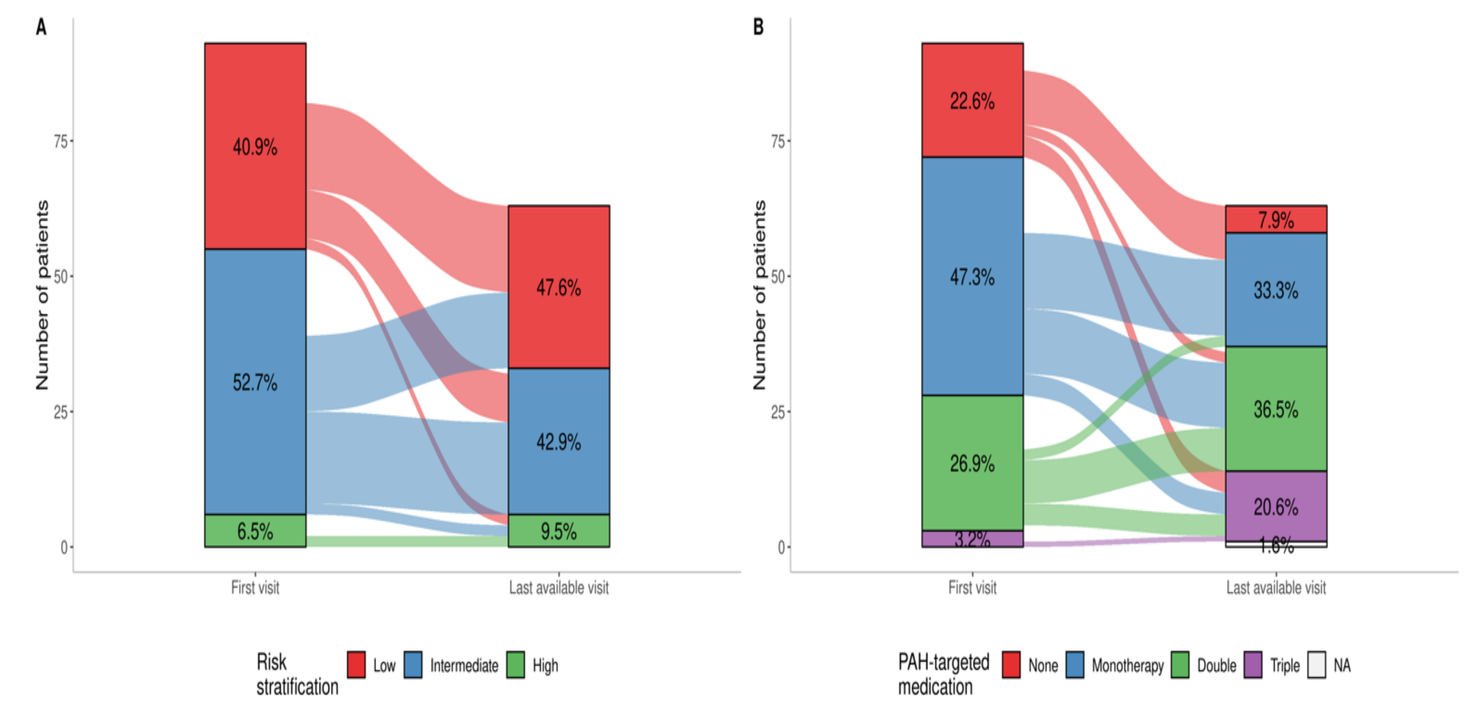
Background: Pulmonary arterial hypertension (PAH) is a common complication among adult patients with congenital heart disease (ACHD).

Purpose: This study presents "real-world" data on risk stratification, pharmacotherapy and mortality in PAH-ACHD

Methods: The Hellenic Pulmonary Hypertension Registry (HOPE) registry was launched in early 2015 and enrols patients with pulmonary hypertension in Greece. This analysis focuses on PAH-ACHD.

Results:

- 93 PAH-ACHD patients, 8 centers, 2015- 2023
- 58 women (62%)
- A follow-up visit available in 63 patients (68%)
- Median age at first visit: 37 (25, 60) years
- Pre-tricuspid shunt (n=35, 38%) / post-tricuspid shunt (n=40, 43%)/ complex ACHD (n=18, 19%)
- Eisenmenger physiology was present in 32 patients (35%)
- NYHA I/II at baseline (n=59, 63%) / NYHA I/II at follow-up (n=38/63, 60%)



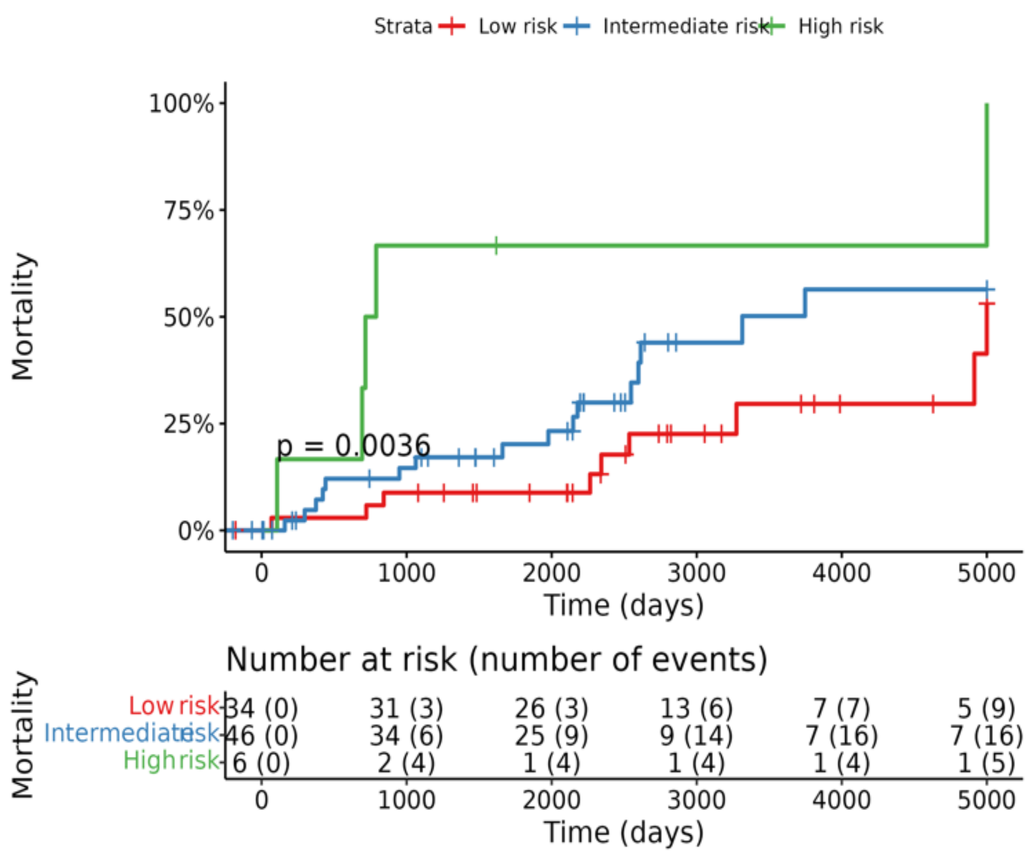


Figure 1

A. Risk stratification of patients with PAH-CHD at baseline (n=93) and last follow up (n=64) according to the latest 2022 ESC/ERS Guidelines on Pulmonary Hypertension.

- Three strata ESC risk score: 52.7% intermediate risk at baseline, 47.6% low risk at follow up.

B. Use of PAH targeted pharmacotherapy at baseline and at last follow up among patients with PAH-CHD.

- PAH monotherapy at baseline: 47.3%. Combination therapy at follow up: 57.1%

Colored lines depict the changes in risk stratification and PAH pharmacotherapy between baseline and last follow up visit.

Figure 2

Mortality of PAH-CHD cohort according to the 1-year mortality risk as assessed at baseline.

- During a median follow up time of 6.9 years, 30 deaths were reported (34.8%).
- 1-, 3- and 5- year mortality rates were 4.8%, 17.3% and 18.8%, respectively.
- Mortality rate was higher in high-risk patients compared to moderate and low-risk at baseline

Conclusions: Overall, there was a tendency towards double or triple combination therapy at follow-up, which might be the cause for a reduction in 1-year ESC mortality risk. However, overall mortality rate remains high among PAH-ACHD patients.

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