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Refining the 2015 European guidelines risk assessment tool for pulmonary arterial hypertension in adult congenital heart disease

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Background: Current European guidelines advocate a goal-oriented treatment approach in pulmonary arterial hypertension (PAH), based on a comprehensive risk assessment. However, this instrument has been based predominantly on patients with idiopathic PAH and its accuracy has not been well established for PAH associated with congenital heart disease (CHD)–a patient population known to be distinctly different for other PAH aetiologies.
Purpose: To investigate the discriminatory ability of the guidelines risk assessment tool and explore the benefit of including other cut-offs or variables in PAH-CHD.
Methods and results: Data from 112 PAH-CHD patients (age 42.1±16 years, 70% Eisenmenger, 38% Down syndrome) seen between 2004 and 2016 at two specialized adult PAH-CHD expert centres were prospectively collected. Patients were classified as "Low", "Intermediate", or "High" risk following the strategy proposed by Kylhammar (Eur Heart J, 2017) based on N-terminal pro-brain natriuretic peptide (NT-proBNP), 6-minute walk distance, functional class and imaging parameters and analysed by Kaplan-Meier method, truncated at 5 years. At baseline, 25% (28) of patients were classified as "Low risk", 69% (77) as "Intermediate risk" and 6% (7) as "High risk". Although survival was better (P=0.012) for patients with higher proportions of "Low risk" variables, this method did not discriminate well between the three risk groups (Figure 1A, P=0.371). One-year survival estimates corresponded moderately to those proposed by the guidelines, 96.4% in the "Low risk" (vs. >95%), 94.8% in the "Intermediate risk" (vs. 90–95%), and 85.7% in the "High risk" (vs. <90%) baseline cohorts, respectively. Analysis of different cut-off values for NT-proBNP (i.e., "Low", "Intermediate", "High" as <500, 500–1440 and >1400 ng/l, respectively) and use of tricuspid annular plane systolic excursion (TAPSE) measurements ("Low", "Intermediate", "High" as <1.6, 2.6–2.7 and >2.7 cm, respectively) as imaging parameter instead of right atrial area improved discrimination between the risk groups (Figure 1B). With these adjustments to the risk assessment tool, survival differed between all three risk groups (P<0.001).
Conclusion: Our preliminary findings suggest that an updated version of the European guidelines risk assessment tool–with different cut-off values for NT-proBNP and use of TAPSE–discriminates more accurately in the PAH-CHD population. Further analysis will be performed to estimate the prognostic benefit of reaching a "Low risk" profile, as this is the recommended treatment goal.

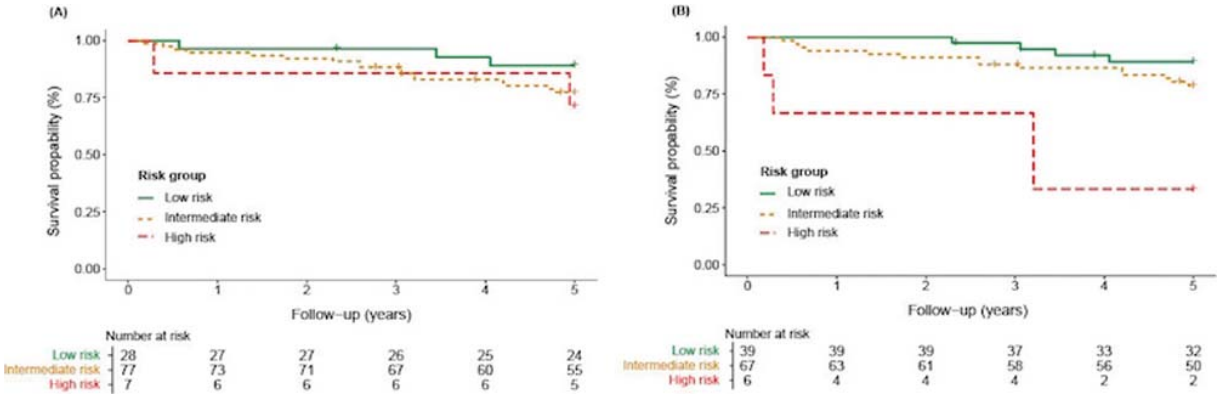

Figure 1