Title: External validation of the REVEAL risk score calculator for PAH survival: A French pulmonary hypertension network analysis

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Body: Introduction: The French Pulmonary Hypertension Network (FPHN) and the Registry to EValuate Early And Long-term Pulmonary Arterial Hypertension (PAH) Disease Management (REVEAL) have recently developed predictive models for survival in patients with PAH. Aims and objectives: The REVEAL risk score calculator was assessed in a FPHN cohort to evaluate its generalizability in a different PAH population. Methods: French validation cohort was built to approximate the inclusion/exclusion criteria originally defined by REVEAL. For each patient, the risk score was evaluated using most recent assessment of risk at any clinic visit from November 2006 or later. Results: The French validation cohort had 1738 patients (pts) with group 1 PAH, mean ± standard deviation 6-min walk distance of 356 ± 130 m and 53% pts in functional class III. Observed 1-yr survival for patients with risk scores of 1-7, 8, 9, 10-11, or 12+ were 96.4%, 88.6%, 86.4%, 78.7%, and 62.1%, respectively (Figure) falling close to the predicted risk for each of these 5 pre-specified risk strata. The c-index for the risk calculator was .729. Predicted versus observed 1-year survival in FPHN.

Conclusion: The REVEAL risk score calculator was accurate and well calibrated in the FPHN, suggesting its
prognostic generalizability in a different PAH population and in everyday clinical practice.