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Humbert M, Sitbon O, Chaouat A, et al. Survival in patients with idiopathic, familial, and anorexigen-associated pulmonary arterial hypertension in the modern management era. *Circulation*. 2010;122(2):156-163.

This study reports the observed survival of patients with idiopathic, familial (heritable), and anorexigen-associated pulmonary arterial hypertension (PAH) from the French Network on Pulmonary Hypertension. This registry draws from 17 pulmonary vascular centers in France. A total of 354 consecutive patients (56 incident, 298 prevalent) were enrolled from October 2002 to October 2003, and followed prospectively. Of the prevalent cases, only those with diagnosis less than 36 months from the time of enrollment were included in the survival analysis. The final cohort of 190 patients was followed for 3 years.

The baseline characteristics of the 190 patients revealed a mean age of 52.5; 63% were female. The majority of patients were in NYHA functional class III (68.4%). Only 17.4% were

in either functional class I/II. PAH treatment was administered with the following distribution: epoprostenol 14.7%, bosentan 35.3%, sildenafil 2.1%, iloprost 2.1%, beraprost 2.6%, treprostinil 0.5%, combination 12.6%. PAH targeted therapy was observed in 76.8% of the cases. Conventional therapy, consisting of calcium channel blockade (13.8%) or patients unable to receive a PAH targeted agent, was observed in 29.5% of the cohort. Background therapy included: warfarin 90.5%, diuretics 69.5%, and oxygen 30.3%.

For the 190-patient cohort, survival at 1, 2, and 3 years was 82.9%, 67.1%, and 58.2%. These rates were roughly 10% higher than those estimated based on the NIH registry formula. Cox analysis found the following variables associated with improved survival: female gender, greater 6-minute walk distance (6MWD), lower right atrial pressure, and higher cardiac output. Although NYHA functional class I/II had a hazard ratio of 0.402, the 95% CI was 0.158-1.019 with a *P* value of 0.06. Specific cutoff or change in 6MWD or cardiac output was not reported, which correlated with improved outcome.

The authors concluded that this represented a “real-world” observational study on PAH survival (as opposed to formulary predictions) in the era of modern therapies. Although survival is better than predicted, incident cases of idiopathic, heritable, or anorexigen-associated PAH remains a progressive and often fatal disease. This observation, however, does not expand or elaborate on the seemingly low rate or timing of intravenous prostanoid therapy. Also, with 21.4% of incident cases and 13.8% of combined cases receiving calcium channel blockade therapy, argument can be made that this cohort was a transition group over-represented with conventional therapy.

Benza RL, Miller DP, Gomberg-Maitland M, et al. Predicting survival in pulmonary arterial hypertension: insights from the Registry to Evaluate Early and Long-Term Pulmonary Arterial Hypertension Disease Management (REVEAL). *Circulation*. 2010;122(2):164-172.

This report utilized data generated from the Registry to Evaluate Early and Long-Term Pulmonary Arterial Hypertension Disease Management (REVEAL) to discern predictors of survival in pulmonary arterial hypertension (PAH). The currently available prognostic tool based on the original 1980s NIH Registry of Primary Pulmonary Hypertension has limitations when applied to other PAH subtypes and in the current era of multiple PAH targeted therapies.

This study analyzed data from 2716 registered patients from US treatment centers. These represent both prevalent (86.5%) and incident (13.5%) cases. The primary goal was to come up with predictors of 1-year survival. Based on the predictors, a formula was derived and presented for clinical application. Table 1 details the baseline characteristics of the 2716 patients at the time of enrollment into the registry. Mean age was 50. More female and Caucasian patients were represented—79% and 73% respectively. Nearly half of the patients were functional class III (48.2%) at the time of enrollment. Only 5.5% were functional class IV. Idiopathic PAH represented nearly half of all patients (46.5%). Connective tissue disease associated PAH was the second largest cohort, representing 23.9%. The distribution of PAH

targeted therapies was relatively even across the 3 classes: prostacyclin analogs 41.6%, endothelin receptor antagonists 46.9%, and phosphodiesterase-5 inhibitors 49.6%. Combination therapy was being used in 40%; intravenous prostacyclin therapy was observed in 26.2%. Calcium channel blockade therapy was observed in 9.2% as monotherapy, or 15.8% as combination therapy.

The observed 1-year survival was 91.0% (95% CI 89.9-92.1). Multivariable analysis identified several independent risk factors. Greater than 2-fold increase in mortality was associated with PAH associated with portal hypertension (HR 3.6, 95% CI 2.4-5.4), family history of PAH (HR 2.2, 95% CI 1.2-4.0), men >age 60 (HR 2.2, 95% CI 1.6-3.0), NYHA functional class IV (HR 3.1, 95% CI 2.2-4.4), and pulmonary vascular resistance >32 Wood units (HR 4.1, 95% CI 2.0-8.3). Other variables associated with mortality included the following: PAH associated with connective tissue disease, renal insufficiency, NYHA functional class III, resting systemic systolic blood pressure <110 mm Hg, resting heart rate >92 bpm, 6MWD <165 m, BNP >180 pg/mL, presence of pericardial effusion, predicted DLCO ?32%, and mean right atrial pressure >20 mm Hg. Four variables associated with improved 1-year survival were: NYHA functional class I, 6MWD ?440 m, BNP <50 pg/mL, and percent predicted DLCO ?80%. The report discusses numerous other parameters that were not found to be independent risk factors by the current analysis.

The authors concluded that this study identified key predictors of survival leading to a contemporary prognostic equation for PAH. The authors acknowledged the potential survival bias by analyzing mostly prevalent cases (86.5%). They recommended ongoing reassessment and external validation of the observed predictors and the proposed equation. Whether the fore-knowledge of these risk factors will alter outcome is unknown.

Comments

Related Resources

- Prognostication in Pulmonary Arterial Hypertension and Use of Current Risk Prediction Models
- Survival of Patients with Pulmonary Arterial Hypertension (PAH) Associated with Anorexigens vs. Survival Predicted From NIH Registry on Primary Pulmonary Hypertension (PPH) Observations
- Treatment Patterns and Predictors of Drug Therapy in Pulmonary Arterial Hypertension (PAH) Between 1995 and 2005
- Predicting Survival in Pulmonary Arterial Hypertension Using the REVEAL Database
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John Newman, MD

Director, Pulmonary & Critical Care Fellowship Program
Vanderbilt University School of Medicine
Nashville, Tenn.

801 Roeder Road, Ste. 1000, Silver Spring, MD 20910
301-565-3004 | PHAOnlineUniv@PHAssociation.org