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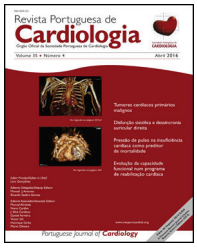

EDITORIAL COMMENT

Lessons from pulmonary hypertension registries

Lições retiradas dos registos de hipertensão pulmonar

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Pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH) are major causes of pulmonary hypertension (PH) that have benefited from novel medical, interventional and surgical strategies in the last two decades.[1–3] Patient registries have provided important information on the clinical characteristics and natural history of different forms of PH.[4,5] Since the US National Institutes of Health (NIH) registry conducted in the 1980s, subsequent registries and databases have yielded additional information on the demographic factors, treatment, and survival of patients with different forms of PH, including PAH and CTEPH.[6–9] These registries have enabled comparisons between populations in different eras and environments.[10,11] In addition, the NIH and French registries have developed equations to predict one-, two- and three-year survival of patients with idiopathic, heritable, and drug-induced PAH.[12–15] These equations have been widely used as comparators in subsequent studies, in order to present indirect evidence of improved outcomes.[4] Since then, the US Registry to Evaluate Early and Long-term Pulmonary Arterial Hypertension Disease Management (REVEAL) has produced different tools, including the REVEAL score and the REVEAL score calculator, that can be used to predict one-year survival of PAH patients.[15,16] More recently, three large European registries have tested a risk assessment instrument derived from the 2015 European Society of Cardiology/European Respiratory Society PH guidelines[1,2] in large cohorts of PAH patients, underscoring the importance of well-designed multicenter registries to support clinical research in PH.[17–20]

An important observation of PAH and CTEPH registries is that survival in the modern treatment era has improved compared with that observed previously.[5,14,21] In addition, PAH registries consistently show that outcomes vary markedly between different PH etiologies, PAH complicating the course of connective tissue diseases being associated with worse outcomes than idiopathic PAH.[13,16,22] Continuing systematic clinical surveillance of PH is essential as treatment evolves.

In the current issue of the *Journal*, Santos et al. present original data on the long-term survival of PAH and CTEPH patients diagnosed between 2005 and 2016 in a Portuguese PH referral center (Hospital Santo António, Centro Hospitalar do Porto).[22] These data indicate that there is a trend for better outcomes in Portuguese PH patients treated in an expert center, but they also confirm that PAH and CTEPH remain disabling and life-limiting conditions.[22] The authors should be congratulated for their results, and also for developing dedicated PH software

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(PAHTool®, Inovultus Lda., Santa Maria da Feira, Portugal). This software is a major achievement of the Portuguese PH research community and is currently disseminated and used worldwide on a daily basis. For example, the PAHTool® is used in the 25 centers of the French Registry, and has been instrumental in the generation of recent data on PH risk assessment.[19]

Research and care for rare diseases is a timely topic in Europe, with the recent approval by the European Commission's Board of Member States of 24 European Reference Networks (ERN), including one for rare respiratory diseases (ERN-LUNG).[23] ERN-LUNG is currently made up of 60 centers in 12 countries and is organized into nine core networks representing the diversity of diseases and conditions affecting the respiratory system, including PH. In the PH Core Network, Portugal is represented by the Centro Hospitalar do Porto. Of note, ERN-LUNG has won the competition for a grant from the European Union for establishing registries within ERN-LUNG where they are still lacking, and for making existing registries, such as PH registries, fully interoperable.[24] ERN-LUNG has proposed building a comprehensive infrastructure for patient data management within ERN-LUNG, and PH will be a key condition tested in this registry warehouse.

In the past 20 years, major changes have taken place in the epidemiological and treatment landscape of PAH and CTEPH. Santos et al. have shown improvements in survival of PH patients in the modern management era in Portugal. The next challenge will be to further improve PH patient outcomes, resulting from better implementation of diagnosis and treatment guidelines and stronger support for basic, translational and clinical research at the national and international level.

Conflicts of interest

The author has no conflicts of interest to declare.

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