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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

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conditions.[22] The authors should be congratulated for their results, and also for developing dedicated PH software (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.

References

1. Galiè N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC). International Society for Heart and Lung Transplantation (ISHLT). Eur Heart J. 2016;37:67–119.
2. Galiè N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC). International Society for Heart and Lung Transplantation (ISHLT). Eur Respir J. 2015;46:903–75.
3. Weatherald J, Taniguchi Y, Humbert M. Novelties in the treatment of pulmonary hypertension. Arch Bronconeumol. 2017;53:235–6.
4. McGoon MD, Benza RL, Escribano-Subias P, et al. Pulmonary arterial hypertension: epidemiology and registries. J Am Coll Cardiol. 2013;62:D51–9.
5. Lau EMT, Giannoulatou E, Celermajer DS, et al. Epidemiology and treatment of pulmonary arterial hypertension. Nat Rev Cardiol. 2017;14:603–14.
6. Rich S, Dantzker DR, Ayres SM, et al. Primary pulmonary hypertension. A national prospective study. Ann Intern Med. 1987;107:216–23.
7. Humbert M, Sitbon O, Chaouat A, et al. Pulmonary arterial hypertension in France: results from a national registry. Am J Respir Crit Care Med. 2006;173:1023–30.
8. Badesch DB, Raskob GE, Elliott CG, et al. Pulmonary arterial hypertension: baseline characteristics from the REVEAL Registry. Chest. 2010;137:376–87.
9. Pepke-Zaba J, Delcroix M, Lang I, et al. Chronic thromboembolic pulmonary hypertension (CTEPH): results from an international prospective registry. Circulation. 2011;124:1973–81.
10. Frost AE, Badesch DB, Barst RJ, et al. The changing picture of patients with pulmonary arterial hypertension in the United States: how REVEAL differs from historic and non-US Contemporary Registries. Chest. 2011;139:128–37.
11. Sitbon O, Benza RL, Badesch DB, et al. Validation of two predictive models for survival in pulmonary arterial hypertension. Eur Respir J. 2015;46:152–64.
12. D'Alonzo GE, Barst RJ, Ayres SM, et al. Survival in patients with primary pulmonary hypertension: results from a national prospective registry. Ann Intern Med. 1991;115:343–9.
13. Humbert M, Sitbon O, Yaici A, et al. Survival in incident and prevalent cohorts of patients with pulmonary arterial hypertension. Eur Respir J. 2010;36:549–55.
14. 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;122:156–63.
15. 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;122:164–72.
16. Benza RL, Gomberg-Maitland M, Miller DP, et al. The REVEAL Registry risk score calculator in patients newly diagnosed with pulmonary arterial hypertension. Chest. 2012;141:354–62.
17. Kylhammar D, Kjellström B, Hjalmarsson C, et al. A comprehensive risk stratification at early follow-up determines prognosis in pulmonary arterial hypertension. Eur Heart J. 2017 June 1, http://dx.doi.org/10.1093/eurheartj/ehx257 [Epub ahead of print].
18. Hoeper MM, Kramer T, Pan Z, et al. Mortality in pulmonary arterial hypertension: prediction by the 2015 European pulmonary hypertension guidelines risk stratification model. Eur Respir J. 2017;50:1700740.
19. Boucly A, Weatherald J, Savale L, et al. Risk assessment, prognosis and guideline implementation in pulmonary arterial hypertension. Eur Respir J. 2017;50:1700889.
20. Benza RL, Farber HW, Selej M, et al. Assessing risk in pulmonary arterial hypertension: what we know, what we don't. Eur Respir J. 2017;50:1701353.
21. Farber HW, Miller DP, Poms AD, et al. Five-year outcomes of patients enrolled in the REVEAL Registry. Chest. 2015;148:1043–54.
22. Santos M, Gomes A, Cruz C, et al. Long-term survival in pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension: insights from a referral center in Portugal. Rev Port Cardiol. 2018;37:753.

23. Humbert M, Wagner TO. Rare respiratory diseases are ready for primetime: from Rare Disease Day to the European Reference Networks. Eur Respir J. 2017;49:1700085.

24. http://ern-lung.eu/inhalt/wp-content/uploads/2017/10/ERN-LUNG-Newsletter-Issue-6-October-2017_final.pdf. [accessed 09.04.18].