Revista Portuguesa de Cardiologia 41 (2022) 749–750

Revista Portuguesa de
Cardiologia
Portuguese Journal of Cardiology
www.revportcardiol.org

EDITORIAL COMMENT

Therapeutic options for thromboembolic pulmonary hypertension: An overview of the situation in Portugal

Opções terapêuticas para a hipertensão pulmonar tromboembólica crónica: Um olhar sobre a realidade portuguesa

Filipa Ferreira

Serviço de Cardiologia, Hospital Garcia de Orta, Portugal

Available online 27 May 2022

Chronic thromboembolic pulmonary hypertension (CTEPH) is a specific form of pulmonary hypertension characterized by fibrothrombotic obstruction of the pulmonary arteries. Although rare, it is the most frequent form of precapillary pulmonary hypertension after exclusion of lung disease.

When untreated, CTEPH has a poor prognosis, with a three-year mortality that can exceed 50%.[1] Because of this unfavorable prognosis early diagnosis is essential. CTEPH should be actively sought in all patients after acute pulmonary embolism (PE) who remain symptomatic after three months of effective anticoagulation. However, a significant number of patients are diagnosed without a known prior acute PE, so this diagnosis also needs to be considered when investigating patients who present with pulmonary hypertension.

There is a potentially curative surgical treatment that should be considered for all patients. Pulmonary endarterectomy (PEA) is the treatment of choice, by removing obstructive thromboembolic material from the pulmonary arteries in order to reduce pulmonary vascular resistance, improving survival and quality of life. In the latest retrospective case series, in-hospital mortality ranged from 2.2% to 7.0% depending on the center's experience, and long-term survival was excellent, with three-year survival of 90%.[2] Persistent pulmonary hypertension after PEA is common and reported in about one third of patients.[3] Recurrent pulmonary hypertension is less common and is often caused by a further thromboembolic episode after a successful PEA clearance.

Although PEA is an excellent option, the European CTEPH registry indicated that up to 40% of patients diagnosed with CTEPH were considered inoperable,[2] even when assessed in expert centers.

For inoperable, residual or recurrent disease, pulmonary vasodilator therapy should be considered, particularly with riociguat (a soluble guanylate cyclase stimulator), which is the only pulmonary vasodilator approved in this setting, although other pulmonary vasodilators are commonly used off-label.[3]

Additionally, balloon pulmonary angioplasty (BPA) has attracted increasing interest in recent years and has evolved into an important component of the CTEPH treatment algorithm. BPA was first described by Feinstein in 2001, but because of high rates of serious complications, it was necessary to refine the technique, which then made headway worldwide. Since 2013, BPA has been performed in certain European centers. This percutaneous technique was first

DOI of original article: https://doi.org/10.1016/j.repc.2021.05.018
E-mail address: filipamaferreira@hotmail.com

https://doi.org/10.1016/j.repc.2022.04.007

introduced in Portugal, as a sustained program, in 2017 by Cale et al.,[4] and their results have recently been published, demonstrating efficacy and safety.[5]

The paper by Samouco et al. published in this issue of the *Journal*[6] assesses the results of a national pulmonary hypertension referral center in the last 10 years, in which overall reported survival at one, two and three years was 93.3%, 82.4% and 75.9%, respectively. Since this is a retrospective study, it has several limitations, which are clearly acknowledged by the authors. However, it is important since it demonstrates that although PEA was not available in Portugal for many years, it was possible to work together with other European referral centers, and thereby avoid depriving patients of the best possible treatment. This was also clearly demonstrated in a recently published paper by another Portuguese pulmonary hypertension referral center.[7] Since 2019, there has also been a recognized national reference center for PEA, whose initial surgical experience was published in 2020.[8] Although they reported high periprocedural mortality, this could be partially explained by the difficult cases initially referred and by the learning curve inevitable in such a complex surgical procedure.[9] It will be important to document reduction of in-hospital mortality in the future, as well as to assess hemodynamic results in medium- and long-term follow-up.

This paper reveals a more favorable outcome in CTPEH patients who underwent PEA surgery, which is in agreement with reported European and US multicenter experiences.[2,10] A high rate of operable patients (72%) was reported; however, 44% of patients did not undergo multidisciplinary assessment. Referral to foreign centers with time-consuming contacts and a high administrative burden can probably explain this undesirably high figure. Of the 30 non-operated patients, only two underwent BPA. It is not clear whether BPA was offered to or considered for every inoperable patient. Since BPA is available at a national level, referral for BPA should nowadays be offered and significantly fewer patients should be treated with medical therapy alone.

To conclude, we currently have at our disposal all the therapeutic strategies at a national level for the treatment of CTEPH: PEA surgery, BPA and medical therapy. These strategies are not mutually exclusive: an optimal treatment strategy might be a combination of two or even all three approaches. It is important to ensure continuous training of experienced multidisciplinary teams, as well as to recognize the advantages and limitations of each technique, so that the best possible treatment can be offered to the individual CTEPH patient.

Conflicts of interest

The author has no conflicts of interest to declare.

References

1. Lewczuk J, Piszko P, Jagas J, et al. Prognostic factors in medically treated patients with chronic pulmonary embolism. Chest. 2001;119:818–23.
2. Delcroix M, Lang I, Pepke-Zaba J, et al. Long-term outcome of patients with chronic thromboembolic pulmonary hypertension: results from an international prospective registry. Circulation. 2016;133:859–71.
3. Ghofrani HA, D'armini AM, Grimminger F, et al. Riociguat for the treatment of chronic thromboembolic pulmonary hypertension. N Engl J Med. 2013;369:319–29.
4. Cale R, Ferreira F, Pereira AR, et al. Balloon pulmonary angioplasty protocol in a Portuguese pulmonary hypertension expert center. Rev Portug Cardiol. 2021;40:653–65.
5. Calé R, Ferreira F, Pereira AR, et al. Safety and efficacy of balloon pulmonary angioplasty in a Portuguese pulmonary hypertension expert center. Rev Portug Cardiol. 2021;40:727–37.
6. Samouco G, Fonseca M, Correia JB, et al. Chronic thromboembolic pulmonary hypertension: a 10-year analysis from a Portuguese referral center. Rev Portug Cardiol. 2022;41:741–8.
7. Plácido R, Guimarães T, Jenkins D, et al. Chronic thromboembolic pulmonary hypertension: initial experience of patients undergoing pulmonary thromboendarterectomy. Rev Portug Cardiol. 2021;40:741–52.
8. Fragata J, Telles H. Pulmonary thromboendarterectomy in Portugal: initial experience. Rev Portug Cardiol. 2020;39:505–12.
9. Madani M, Auger W, Pretorious V, et al. Pulmonary endarterectomy: recent changes in a single institution's experience of more than 2,700 patients. Ann Thorac Surg. 2012;94:97–103.
10. Kerr K, Elliot CG, Chin K, et al. Results from the United states chronic thromboembolic pulmonary hypertension registry: enrollment characteristics and 1-year follow-up. Chest. 2021;160:1822–31.