Letter to Editor

Should cardiopulmonary exercise testing become a part of regular evaluation for patients with a family history of pulmonary hypertension?

Regarding “Cardiopulmonary exercise testing reveals onset of disease and response to treatment in a case of heritable pulmonary arterial hypertension”

Editor:

The case presented by Trip et al. wonderfully highlights the importance of cardiopulmonary exercise testing (CPET) for evaluating patients with familial pulmonary hypertension (PH). Nevertheless, it is crucial to understand certain salient features of CPET which can further strengthen the interpretation.

Recent reviews by Arena et al. and Guazzi et al. have described which CPET variables provide important clinical information in patients with PH. The data presented by Trip et al. demonstrate abnormalities in a number of these key CPET variables in their patient with PH.

The prognostic implications of CPET responses in patients with PH are beginning to gain recognition. For the CPET variables reported by Trip et al., we have provided the survival rates based on cut-off values from previous studies. An oxygen consumption (VO\textsubscript{2}) <1.32 l/kg/min has cumulative survival rates of 71\% and an oxygen (O\textsubscript{2}) pulse <12 ml/beat with and without cardiopulmonary disease has a relative mortality risk of 3.4 and 2.2, respectively. The reported O\textsubscript{2} pulse of 9.1 ml/beat reported in the current study suggests greater disease severity with a higher mortality risk, which decreased with initiation of treatment, though not completely ameliorating risk. The Ventilatory efficiency slope (VE/VCO\textsubscript{2} slope) reported in this case was higher than that observed for those with thromboembolic PH after normalizing pulmonary pressures (33).

This information shows that even after beginning therapy, though the CPET variables favorably changed, the risk for adverse events may still remain elevated. This, however, could be due to the short duration of therapy. Perhaps a follow-up CPET after 1 year will demonstrate a better response to therapy. In the setting of PH, the longitudinal use of CPET may play a vital role in determining the onset of disease as well as track disease progression and the response to interventions. Additional information from assessments of dead space to tidal volume assessments will help in identifying the severity of ventilation-perfusion mismatch that exists in these patients.

From the case described, it is seen that the 2009 report was completely normal while in 2012 there was a drastic decrement in CPET parameters. The period between 2009 and 2012 may have had steady decrements in the CPET response, although not to the extent of being symptomatic. Thus, it may be appropriate to advocate yearly CPET evaluations along with a blood workup and echocardiography for those patients with a history of familial PH or bone morphogenetic protein receptor type 2 (BMPR2) mutation. However, only prospective follow-up of generations of offspring from those with BMPR2 mutations will provide an answer to the utility of serial CPET assessments in this patient population.

Abraham Samuel Babu, Ross Arena, Arun G. Maiya, Ramachandran Padmakumar, and Marco Guazzi

Department of Physiotherapy, Manipal College of Allied Health Sciences, Manipal University, Manipal, Karnataka, India; Division of Physical Therapy, Department of Orthopaedics and Rehabilitation, and Division of Cardiology, Department of Internal Medicine, University of New Mexico Health Sciences Center, Albuquerque, New Mexico, USA; Dr.TMA Pai Endowment Chair in Exercise and Health Promotion, Manipal University, Manipal; Department of Cardiology, Kasturba Medical College, Manipal University, Manipal and Cardiopulmonary Laboratory, Cardiology Division, San Paolo Hospital, University of Milano, Milan, Italy

Email: abrahambabu@gmail.com

REFERENCES

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The valuable comments made by Babu et al. are very much appreciated. Babu et al. stress the value of exercise oxygen consumption and oxygen pulse to assess mortality risk.[1,2] Indeed, both CPET parameters were reduced in the presented patient and remained reduced after the start of therapy.[3] Babu et al. hypothesize that the lack of normalization in CPET parameters may have been explained by a short duration (4 months) of therapy. However, during her recent 1 year follow-up evaluation, we observed no further improvements in maximal exercise capacity (from 167 to 162 W), maximal oxygen consumption (from 2.11 to 2.03 l/min), or oxygen pulse (from 12 to 11 ml/beat). It is difficult to determine whether the lack of normalization of CPET parameters translates into a poor prognosis. The prognostic value of changes in CPET parameters after the start of therapy remains unclear. Studies examining the prognostic utility of CPET in pulmonary arterial hypertension (PAH) were performed in a relatively small number of patients,[4] and in these studies, measurements were only done at baseline.

Babu et al. make valuable comments on the usefulness of CPET as a screening tool for patients with a family history of PAH. They suggest that in the presented case, it may have been appropriate to perform yearly CPET along with a blood workup and echocardiography. They hypothesize that the patient may have had steady decrements in CPET parameters between 2009 (no PAH) and 2012 (diagnosis of PAH). Although we agree that steady decrements may have been visualized by repeated tests, we question if performance of CPET would have altered this patient’s management. She was seen by a cardiologist between 2009 and 2012 and repeated echocardiography did not reveal any signs of PH. In addition, the patient only started to experience physical complaints in November of 2011. We would probably not have performed a right heart catheterization in a patient with no physical complaints or signs of PH on echocardiography, even if CPET would have shown some subtle abnormalities. We agree with Babu et al. that only a prospective follow-up study in offspring of heritable PAH patients will determine the value of CPET as a screening tool for PAH.

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