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).[1] Nevertheless, it is crucial to understand certain salient features of CPET which can further strengthen the interpretation.

Recent reviews by Arena et al.[2,3] and Guazzi et al.[4] have described which CPET variables provide important clinical information in patients with PH. The data presented by Trip et al.[1] 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.,[1] we have provided the survival rates based on cut-off values from previous studies. An oxygen consumption (VO_2) <1.32 l/kg/min has cumulative survival rates of 71%[5] and an oxygen (O_2) pulse <12 ml/beat with and without cardiopulmonary disease has a relative mortality risk of 3.4 and 2.2, respectively.[6] The reported O_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_2 slope) reported in this case was higher than that observed for those with thromboembolic PH after normalizing pulmonary pressures (33).[7]

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 (BMPRI2) 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.

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Quick Response Code:	**Website:** www.pulmonarycirculation.org
	DOI: 10.4103/2045-8932.109927