Review of the Latest Published Research

Section Editor
Nick Kim, MD

Section Editor
Ioana Preston, MD

Summaries and commentaries from the section editors and invited reviewers present a clinical context for pracitioners' application of the latest published research relevant to the care of patients with pulmonary hypertension. In this issue, Nick Kim, MD, discusses the implications for screening for PH in patients with systemic sclerosis.

Humbert M, Yaici A, de Groote P, et al. Screening for pulmonary arterial hypertension in patients with systemic sclerosis: clinical characteristics at diagnosis and long-term survival. Arthritis Rheum. 2011 July 18. [Epub ahead of print]

Emphasis on early diagnosis and treatment of pulmonary arterial hypertension (PAH) has been commonplace in consensus algorithms and pulmonary hypertension lectures. The subgroup of PAH associated with systemic sclerosis (PAH-SSc) has perhaps garnered the most attention in an effort to make an early diagnosis. The rationale for screening in SSc relates to its relatively high incidence of PAH, recognition that PAH may be the most devastating comorbidity within SSc, and the availability of therapies demonstrating efficacy. To that end, prospective screening may identify patients with early PAH-SSc who may be candidates for PAH-targeted therapy.

In this article, our colleagues from Clamart in collaboration with the ItinerAIR-Sclérodermie program have reported their observations on early diagnosis and treatment of PAH-SSc. They report 2 cohorts of PAH-SSc patients enrolled and followed from 2002 and 2003. The first group, "routine practice" cohort, consists of patients diagnosed following routine workup for symptomatic SSc. The second group, "detected" cohort, represents PAH-SSc patients diagnosed only following systematic PAH detection screening for patients with SSc. Both groups required right heart catheterization in keeping with current guidelines for PAH diagnosis. In order to maintain homogeneity, patients with concomitant significant ventilatory or hypoxic lung disease were excluded. Treatment for PAH-SSc followed standard practice guidelines.

The "routine practice" cohort consisted of 16 incident cases of PAH-SSc from a total consecutive pool of 674 patients enrolled in the French PAH registry. This registry also included 58 prevalent cases of PAH-SSc. The "detected" cohort consisted of 16 incident cases of PAH-SSc and 29 prevalent cases, from a pool of 599 patients with SSc enrolled in the ItinerAIR-Sclérodermie program. This report focused on the 32 total incident cases of PAH-SSc. Compared to the "routine practice" cohort, the "detected" group had lower New York Heart Association functional class (detected group had 50% FC III/IV compared with 87% FC III/IV for the routine practice group), milder hemodynamics, and better survival. The 1-, 3-, 5-, and 8-year survival rates were 100%, 81%, 73%, and 64% respectively for the "detected" group versus 75%, 31%, 25%, and 17% respectively for the "routine practice" group. There was no significant difference in use of PAH-specific therapies between groups.

The authors address the critical questions standard to such reports comparing outcome between proactive screening vs standard practice; i.e., lead-time and selection biases. The survival advantage in the "detected" group may in part be explained by earlier diagnosis rather than true benefit of early detection and intervention. Furthermore, screening may introduce patients with inherently different kinetics in terms of disease progression than typical patients diagnosed with established or more advanced symptoms. Cost of such biases includes not only health care resources but also potential psychological effect of early diagnosis and treatment. However, the evidence from clinical trials in mildly symptomatic PAH patients remains compelling and supports efforts toward early diagnosis. Whether screening-based early diagnosis and treatment will confer disease modification or true survival benefit remains to be elucidated.

Correspondence: h33kim@ucsd.edu