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Why patients that die of worsening pulmonary arterial hypertension are not on parenteral prostacyclin analogue treatment?

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We read with great interest the article by Dr Farber et al.(1) on the treatment of patients with pulmonary arterial hypertension (PAH) at the time of death or deterioration to functional class IV, based on the REVEAL (Registry to Evaluate Early and Long-term PAH Disease Management) registry data. The authors concluded that intravenous prostacyclin therapy and combination therapy are not consistently used in patients that either progress to New York Heart Association (NYHA) class IV or died. Although the authors elaborate on the potential explanations of this apparently low use of intravenous prostacyclin analogues or PAH-combination treatment in this particular group of PAH patients, the REVEAL registry did not investigate the particular reasons for not initiating or continuing these PAH-specific treatments.

We have recently presented a study on the causes and circumstances of death in PAH patients in which we carefully investigated the reasons why these patients were not on IV prostacyclin analogues or PAH-combination therapy before death (2). Interestingly, almost all PAH patients (83 of 84) received at least one PAH-specific therapy; however only 50 % of patients (42 out of 84) were on parenteral prostacyclin analogues before death. The proportion of patient receiving the latter treatment increased when considering those that died due to worsening PH (73 %) and were in NYHA class IV (78%). More importantly the reasons for not receiving parenteral prostacyclin analogues included the concomitant presence of advanced interstitial lung disease and hypoxemia in patients with scleroderma (n=2), dementia (n=1), psychiatric disease (n=1), advanced age with inadequate family support (n=1), frequent catheter related infections (n=1), comorbidities with poor prognosis (n=1), suspicion of pulmonary venous occlusive disease (n=1) and patient refusal (n=1).

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Adriano R. Tonelli MD: The author has no significant conflicts of interest with any companies or organization whose products or services may be discussed in this article.

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Most of the PAH patients that died due to worsening PAH and were on NYHA functional class III or IV received PAH combination therapy (n=34, 94 %)

At our institution, as in the REVEAL registry, a significant proportion of PAH patient did not receive treatment before death, but this was because patients had conditions other than PH that led to their demise, were not in NYHA class III or IV or the risk / benefit ratio did not favor initiating this treatment. It would be important to know how the authors adjudicated patients to the “PAH-related death” group, since in our experience this was a very complex task, even when we prospectively reviewed each case in our own program as a group.

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