Sotatercept for pulmonary arterial hypertension: something old and something new

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Sotatercept is a drug that targets a novel pathogenic pathway in pulmonary arterial hypertension and has the potential to be an effective new therapy for this condition https://bit.ly/3s1tQb1

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Humbert *et al.* [1] report safety and efficacy results from the long-term open-label extension study of the phase 2 PULSAR placebo-controlled, randomised trial of sotatercept in pulmonary arterial hypertension (PAH) in this issue of the *European Respiratory Journal*. Subjects who had completed the phase 2, 24-week randomised trial were eligible for participation, with ex-placebo subjects re-randomised to receive sotatercept in doses of either 0.3 or 0.7 $mg \cdot kg^{-1}$, while ex-sotatercept subjects remained on their earlier randomised dose. Of the 106 subjects completing the 24-week PULSAR trial [2], 97 enrolled in the extension study, of whom 30 had received placebo. These patients had moderately severe disease, as reflected by their background use of PAH-targeted therapies: over half were already receiving triple therapy, 36% were receiving double therapy, and over one-third were receiving a parenteral prostacyclin agonist as part of their treatment regimen. Over one-half of subjects had idiopathic PAH, while fewer than 20% had connective tissue disease as the aetiology of PAH. This study population reflects a cohort in need of additional therapy at a time when few other options exist, although it is heavily weighted towards subjects with idiopathic PAH, who tend to have better responses to therapy than those with connective tissue disease.