

Pulmonary Hypertension in Paediatric Patients: data from the COMPERA-KIDS registry

*Gorenflo M. (1), Pittrow D. (2), Huscher D. (3), Ziesenitz V. (1), Pattathu J. (1), Ewert P. (4), Apitz C. (5), Stiller B. (6), Weil J. (7), Abdul-Khaliq H. (8), Berger F. (9), Breuer J. (10), Hansmann G. (11), Michel-Behnke I. (12), Stein J.-I. (13), Hoepfer M. M. (11), Kaemmerer H. (4)
Univ. Hospital Heidelberg, Germany (1); Techn. Univ. Dresden, Germany (2); Charité Hospital, Berlin, Germany (3); German Heart Centre Munich, Germany (4); Univ. Hospital Gießen & Marburg, Germany (5); Univ. Hospital Freiburg, Germany (6); Univ. Heart Centre Hamburg, Germany (7); Univ. Hospital of Saarland, Homburg, Germany (8); German Heart Centre Berlin, Germany (9); Univ. Hospital Bonn, Germany (10); Med. Univ. Hannover, Germany (11); Med. Univ. Vienna, Austria (12); Med. Univ. Innsbruck, Austria (13).*

Background: Pulmonary hypertension (PH / PAH) can have many possible causes in childhood. The aim of the COMPERA registry is the characterization of patients of all ages with PH / PAH and their treatment patterns. Methods: Since June 2013, paediatric patients can be included in the COMPERA registry (ClinTrials.gov: NCT01347216) which has originally been established for adult patients with pulmonary hypertension in 2007. Results: Until November 2015, 102 patients <18 years with pulmonary hypertension were enrolled. Of these paediatric patients, 65.7% had PAH due to congenital heart disease (PAH-CHD), 18.6% had idiopathic PAH (iPAH), 6 had persistent PH of the newborn (PPHN), and 2 had PH associated with interstitial lung disease, valvular heart disease or congenital malformations, respectively. The patients were 5.2 ± 5.9 years old, 58 female; NYHA functional class I / II in 59%, III in 36%, and IV in 5%. The average disease duration after diagnosis was 31.7 ± 52.0 months. Invasive measurement data by right heart catheterization were available for 78% of the patients. The mean pulmonary artery pressure was 45 ± 20 mmHg, the right atrial pressure was 7.9 ± 8.0 mmHg, cardiac index was 3.7 ± 1.3 l/min/m². Monotherapy was received by 66% of the patients, 28% of the patients had a dual and 4% a triple therapy. Phosphodiesterase-5 inhibitors were administered to 81% of the patients, 33% of the patients received endothelin receptor antagonists and 3% received prostacyclins. Conclusion: The most common form of PH in this study cohort is PAH due to congenital heart disease, followed by idiopathic PAH. Invasively measured right heart pressure data were available for 80% of the paediatric patients. Treatment options for children primarily comprise phosphodiesterase-5 inhibitors, although endothelin receptor antagonists are increasingly used. Only a small number of paediatric patients receive anticoagulation therapy.