O9-1

Clinical Characteristics of Pediatric Pulmonary Hypertension: Results from the global registry Tracking Outcomes and Practice in Pediatric Pulmonary Hypertension (TOPP)

Berger R.M.F. (1), Humpl T. (2), Schulze-Neick I.(3), Raskob G (4)., Ivy D.D (5)., Jing Z-C. (6), Bonnet D. (7), Beghetti M. (8), Barst R.J. (9)
Pediatric Cardiology, Beatrix Children's, Hospital, University Medical Center Groningen, Netherlands (1); Pediatrics, University of Toronto, Toronto, Ontario, Canada (2); Pediatrics, Great Ormond Street Hospital for Children, London, United Kingdom (3); University of Oklahoma, Oklahoma, United States (4); Pediatrics, University of Colorado Denver School of Medicine, Aurora, CO, United States (5); Department of Cardio-Pulmonary Circulation, Shanghai Pulmonary Hospital, Tongji University School of Medicine, Shanghai, China (6); Pediatrics, Université Paris Descartes, Necker Enfants Malade, Paris, France (7); Pediatrics, Hôpital des Enfants, Geneva, Switzerland (8) and. Pediatrics, Columbia University, New York, NY, United States (9)

Introduction
Pediatric and adult pulmonary hypertension (PH) share similar pathology and clinical characteristics yet several features appear different. However, pediatric data are lacking. TOPP, a global, observational study, provides demographic, clinical and treatment patterns in pediatric PH.
Methods
Consecutive patients are screened and consenting patients with PH (mean pulmonary artery pressure [mPAP] ≥25 mmHg, pulmonary capillary wedge pressure ≤12 mmHg and pulmonary vascular resistance index [PVRI] >3 WU.m2) are enrolled at 31 centers in 20 countries.
Results
Of 456 patients (age at diagnosis <18 yrs), enrolled between January 2008 and February 2010, 362 had confirmed PH. Of the PH-confirmed patients, median age at diagnosis was 7 yrs with a female preponderance (1.4:1). Mean time from symptom onset to diagnosis (17 mos; 95%CI 14-20 mos) was shorter than reported in adults. The majority (88%) had Group 1 PH: pulmonary arterial hypertension (PAH), of which 57% had idiopathic or familial PAH (IPAH/FPAH) and 43% had PAH associated with other conditions (APAH). Of the APAH patients, 85% had associated congenital heart disease (CHD). The remaining children (12%) had PH associated with lung diseases and/or hypoxemia, with bronchopulmonary dysplasia (BPD) the most frequent association. Co-morbid conditions, including chromosomal abnormalities, other syndromes and anomalies, were reported in 24%. As in adults, dyspnea and fatigue were the most frequent presenting symptoms. In contrast to adult PH, syncope was reported frequently, especially in children with IPAH/FPAH (31%) or repaired CHD (18%); in contrast, no children with unrepaired/residual shunts had syncope reported. Functional class (FC) at diagnosis was predominantly FC II (12%/51%/30%/7%, I/II/III/IV, respectively) consistent with preserved right heart function despite severe PAH, i.e. mPAP (58±19 mmHg) and PVRI (16±10 WU.m2).
Conclusions
TOPP demonstrates that pediatric PH has important characteristics that differ from adults, including syncope as a frequent presenting symptom in IPAH/FPAH despite preserved cardiac output. Additionally, the distribution of etiologies, the occurrence of specific associated pediatric conditions, including BPD and a high rate of co-morbidities appear different than in adults. As we track outcomes in these children, we hope to define prognostic parameters for risk stratification to guide therapeutic approaches in pediatric PH.