Heart Catheterization (HC) complications in children with Pulmonary Hypertension (PH) as reported from the global TOPP Registry (Tracking Outcomes and Practice in Pediatric PH)

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Objectives: PH is a significant cause of morbidity and mortality in children. Hemodynamic assessment via HC remains the “gold-standard” confirming diagnosis and evaluating disease severity. We collected data from the global pediatric TOPP registry regarding complications associated with HC in pediatric PH.

Methods: Of 568 patients (age ≤18 years at diagnosis) enrolled in TOPP (HC required to confirm diagnosis; 34 sites, 20 countries) between 1/2008-2/2012, 486 patients (86%) fulfilled the criteria to confirm PH (HC: mean pulmonary artery pressure ≥25 mmHg, pulmonary capillary wedge pressure ≤12 mmHg, pulmonary vascular resistance index ≥3 WU*m2).

Results: 908 HCs were performed in 555 patients (554 diagnostic; 354 follow-up HCs; conscious sedation in 257 (46%), general anesthesia in 291 (54%)). PH etiology: 263 (47%) idiopathic/familial, 202 (36%) congenital heart disease (CHD) associated, 57 (10%) chronic lung disease. 32 patients had either associated pulmonary arterial hypertension other than CHD (n= 27), or Venice Group 4 (n= 2) or 5 (n=3). Clinically significant complications were reported in 37 patients (6.7% of 555): cardiac arrest (n=5), systemic hypotension requiring intervention (n=17) and PH crises (n=10). Frequency of complications was similar in 16 follow-up HC (6.8% of 325 patients). Five HC-related deaths were reported in TOPP (0.6 %). Factors identified with increased risk for complications included younger age (<1 yr 8/73 [11%]; 1-2 yrs 4/46 [8.7%]; 2≤12 yrs 16/288 [5.6%]; ≥12 yrs 9/147 [6.1%]) (P<0.001, Fisher’s Exact test) and worse WHO functional class (FC): FC I 3/72 [4.2%]; FC II 14/259 [5.4%]; FC III 15/185 [8.1%], and FC IV 5/38 [13.2%] (P<0.001, Fisher’s Exact test). Study results are limited as some patients who died during/shortly after HC may not have been included in TOPP due to its design (informed consent required).

Conclusions: In TOPP, <1% HC-related mortality was reported in >900 HCs. Complications occurred in 6.7% of procedures and increased with lower age and worse WHO FC. HC is required to establish valid hemodynamic data for confirmation of diagnosis and evaluation of treatment. To minimize complications, expertise and experience in the pathophysiology and clinical management of pediatric PH are paramount for all members of the pediatric PH team.