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Acute Vasodilator Response in Paediatric Pulmonary Arterial Hypertension: current clinical practice from the TOPP-registry

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Introduction:
Acute pulmonary vasodilator testing(AVT) in paediatric pulmonary arterial hypertension(PAH) is considered important to identify patients with favourable prognosis using calcium-channel-blocker-(CCB)-therapy. However criteria used to identify acute responders, the prevalence of responders and the use of CCB in paediatric PAH are insufficiently studied.

Methods:
Consecutive PAH-patients, 3months-18years at diagnosis were enrolled in the Tracking-Outcomes-and-Practice-in-Paediatric-Pulmonary-Hypertension-(TOPP)-registry at 31 centres in 19 countries from Jan-2008 to May-2013. Patients who had valid AVT during diagnostic heart catheterization were included in this study. Agents used for this test were recorded. AVT-responder status was identified at the discretion of the treating physician and retrospectively compared to the response according to criteria proposed by Barst for paediatric patients (2012) and Sitbon for adult patients (2005).

Results:
Of 529 PH-confirmed children in the registry, 382 were eligible for inclusion in this study. Of these, 212 had idiopathic/hereditary PAH(IPAH/HPAH) and 105 had PAH associated with congenital heart disease(PAH-CHD). In 70% of the patients, AVT was performed using inhaled NO (+/-O2-suppletion), a variety of other agents were used in the remaining patients. Acute responders were identified in 78 (37%) of IPAH/HPAH patients according to the treating physician, 62 (30%) according to Barst-criteria and 32 (15%) according to Sitbon-criteria. For PAH-CHD patients these percentages were 36%, 13% and 7% respectively. Correlation between assessment by treating physician and by published response criteria was poor.
Regardless of the criteria used, acute responders had favourable baseline hemodynamics compared to non-responders, with lower mean pulmonary arterial pressure, mean right atrial pressure, pulmonary vascular resistance, PVR/SVR and PAP/SAP. There was no association between acute response status and age, sex, 6-minute walking distance or NYHA Functional Class.
Of the IPAH/HPAH patients, judged by the treating physician to be acute responders, only 23% were treated with CCB without additional PAH-targeted therapy. This was 26% in the “Barst-responders” and 47% in the “Sitbon-responders”.

Conclusions:
The current practice of identifying responders to AVT in centres treating children with IPAH/HPAH shows large discrepancies with the assessment by response criteria proposed in literature. Furthermore, in current clinical practice, the majority of paediatric IPAH/HPAH acute responders are not treated with CCB-therapy.