The inaccuracy of Doppler echo estimates of pulmonary arterial pressures in patients with pulmonary arterial hypertension associated with congenital heart disease: Insights from a large multicenter study

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Background: The correlation of Doppler ECHO (DE) estimates with actual systolic and mean pulmonary arterial pressures (PAPs, PAPm) remains controversial. We aimed to assess the accuracy of PAPs and PAPm DE estimates in comparison to heart catheterization (HC) in patients with APAH-CHD in the THALES Registry.

Methods: Patients (>3 months of age) with APAH-CHD (PAPm>25 mmHg, PCWP ≤15 mmHg, and PVRi >3 Wood units.m2), included into THALES Registry, were analysed with regard to correlation between DE and HC. Bland-Altman analysis was performed to test the agreement between HC and DE measures of PAPs and PAPm by 4 tricuspid regurgitation (TRV) based formulas. The 4 formulas used to estimate PAPm based on TRV were: 1.Chemla (0.61xPAPs+2 mmHg), 2.Friedberg (0.69xPAPs - 0.22 mmmHg), 3.Auden (0.70xPAPs), and 4.Bech-Hanssen (0.65xPAPs -1.2 mmHg).

Results: The study population comprised of 1034 APAH-CHD patients (M/F:434/600; mean age±standard deviation (SD) 16.91±17.91 years). The invasively measured mean±SD PAPs and PAPm were 84.5+29.4 and 54.7+22.2 mmHg, transpulmonary and transsystemic pressure gradients (TPPG, TSPG) were 45.0+22.1 and 70.5 +17.6 mmHg, Qp/Qs and PVR/SVR ratio were 3.34+4.84 and 0.38+0.47, respectively. There were mild correlations between DE and HC measures in terms of PAPs (r=0.34; p<0.0001), and PAPm estimated by formula 1 (r=0.31, p<0.0001), 2 (r=0.31, p<0.0001), 3 (r=0.31, p<0.0001) and 4 (r=0.31, p<0.0001), respectively. Using Bland-Altman analysis, the bias for DE estimates with 95% CL for PAPs were -17.83 [95% CL: -84.619 and 48.959] and for PAPm estimated by formula-1 (-14.443 [95% CL: 60.966 and 32.081]), 2 (-9.30, [95% CL: -58.084 and 39.483]), 3 (-8.383 [95% CL: -60.683 and 34.539]) and 4 (-13.072 [95% CL: -57.471 and 40.706]), respectively. The LVEF assessed by echocardiography was 67.4+10.9%, and showed no correlation to any clinical and hemodynamic parameter (p=NS).

Conclusion: DE estimates of PAPs and PAPm weakly correlate with HC in APAH-CHD population. This may be due to inter-center/observer variability and confounding factors associated with volume and pressure overload and may limit the reliability of DE in diagnosis or follow-up of APAH-CHD.