P-162

Combination of Bosentan and Sildenafil therapy in patients with congenital heart defects (CHD): A case series

Rehak T., Nagel B., Fritsch P., Heinzl B., Köstenberger M., Gamillscheg A.
Division of Pediatric Cardiology, Department of Pediatrics, Medical University Graz, Austria

Background:

Data on the effects of an oral combined pulmonary vasodilator therapy in patients with CHD are scarce. Our aim was to evaluate a possible benefit of a combined therapy compared to a monotherapy with either Bosentan or Sildenafil in different forms of operated CHD.

Methods:

Seven patients (median age 13.5 (5.2–27.9) years, 3 f) treated with either Sildenafil or Bosentan were retrospectively analyzed before and during a combination therapy with both Bosentan and Sildenafil. Underlying diagnosis were protein losing enteropathy following Fontan operation (n=2) and pulmonary hypertension secondary to CHD (PAH-CHD) (n=5), patients with Eisenmenger syndrome were excluded : atrioventricular septal defect (n=1), atrial septal defect (n=2), ventricular septal defect (n=2). Invasive pulmonary artery pressures (PAP) and PA resistance, oxygen saturation at rest, NYHA functional class, 6 minute walking distance (6MWD), NT-pro BNP were investigated before start of combination therapy and at latest follow-up. Mean doses of Bosentan were 2.8±1.5 mg/kg/d and Sildenafil 1.7±0.7 mg/kg/d. Mean follow up time was 2.5 years (0.4–5.2 years).

Results:

Mean PAP in PAH-CHD patients decreased from 62±23 to 48±22 mmHg, PA resistance decreased from 18±11 to 14±11 U*m^2 (n.s.). Protein losing enteropathy improved with discontinuation of Albumin replacement. NYHA functional class of all patients improved from 2.4±0.5 to 1.7±0.7 (p=0.003). Oxygen saturation increased from 91±9% to 94±3% (n.s.). 6MWD did not improve significantly over time (534±53m vs. 545±37m). NT-pro BNP levels also did not change. In one patient the Bosentan dose had to be decreased because of elevated liver enzymes, no other major side effects were noted.

Conclusion:

The combination of an oral therapy with Bosentan and Sildenafil was safe in our patient group without major side effects. NYHA functional class significantly improved. However other functional parameters like 6MWD, oxygen saturation and NT-pro BNP as well as invasive hemodynamic parameters in PAH-CHD patients did not change significantly.