Causes and management of systemic arterial desaturation in children with congenital heart defects and functionally single ventricle after the Fontan procedure. A single center experience.

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Introduction: Important complications in children with CHD and single ventricle after the Fontan procedure (FP) include systemic arterial desaturation (SaO2). Objective: to assess the SaO2<90% causes and management in post-Fontan pts, we analyzed cardiac catheterization (CC) and intervention results in a total 52 children, between January 2000 and December 2012.

Material/Methods: SaO2:56%-89% was seen in 19 (36.5%) pts aged 1.5-21 years (x=7.5 ±5.76 years), body mass: x=21.9 ±12 kg. Depending on CHD type, the pts were divided into two groups: Group-I: HLHS (n=8), and Group-II: other complex CHD’s (n=11). The pts underwent staged transition to their completed Fontan circulation, mostly (18) with fenestration. Results: SaO2 in Group-I was x=84.8%±3.7% while in Group-II: x=80.1% ±9.1%. Age at the Fontan procedure in Group-I (x=3.73±2.64 yrs) vs. Group-II (x= 4.4±1.7 yrs) and time between surgery and catheterization (Group-I: x=2.4±4.3 yrs vs. Group-II: x=4.5±4.9 yrs) differed insignificantly (p=0.5, p=0.3, respectively). In Group-I, 6 pts showed significant LPA stenosis treated with balloon angioplasty (BPA) and stents implantation, 1 - restrictive ASD demanding reoperation, 1 – recanalization of LSVC to LA, 1 - large fenestration (both closed percutaneously). One teenager with severe RV dysfunction was qualified to heart transplantation performed 3 mo later. In Group-II: 3 pts showed intracardiac tunnel-RA shunts (closed in 1 percutaneously, in 2 surgically), 3 had PA branch stenosis (PBS) (treated with BPA and stents implantation), 3 – multiple intrapulmonary venous fistulas and 1 - SVC-to-PV fistula (closure possibility in 1 child). Fenestration occlusion was performed in 1 of 2 qualified to this procedure. In all pts treated interventionally, SaO2 increased from x=82.5%±4.6% to 89.2±5%. Conclusions: Most reasons causing systemic arterial desaturation in post Fontan pts was possible to correct interventionally. Their different nature: more frequent PBS in Group I (easier to treat) vs multiple v-v fistulas in Group II (difficult to eliminate) despite similar time from FP to CC was a result of CHD’s type and initial stage of operation.