

Fontan completion – does it really relieve cyanosis?

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Objectives: Total cavopulmonary connection (TCPC) aims for reduction of ventricular volume overload and cyanosis. However, due to the use of fenestration and the development of venovenous (vv.) collaterals, some patients remain cyanotic or desaturate during follow-up (FU). We analyzed the persistence or development of secondary cyanosis during long-term FU after TCPC.

Methods: We retrospectively studied 267 patients (median age 15 (3-57) years), who underwent TCPC at our institution between 1986 and 2015. 16 early deaths were excluded and 251 enrolled in the long-term analysis. During median FU of 6 (0.3-30) years after TCPC, all patients underwent routine investigations in our outpatient department, including 142 cardiac catheterizations for fenestration closure and/or hemodynamic evaluation if indications were given. Transcutaneous oxygen saturation (SaO₂) was noted and compared longitudinally. Cyanosis was defined as SaO₂ ≤ 93%.

Results: Total long-term survival was 90% (N=226). Total long-term mortality (N=25) significantly correlates with cyanosis during FU (20 cyanotic vs. 5 non-cyanotic patients, p< 0.001). On discharge median SaO₂ of the total cohort was 94% (78-100%), 95% (80-100%) in patients without fenestration and 91% (78-98%) in patients with fenestration. In 43 patients fenestration was closed. SaO₂ increased to median of 96% (88-100%) (p<0.01). On last FU SaO₂ was in median 95% (69-100%), 92% (72-99%) in patients who still have an open fenestration and 96% (69-100%) in those without. SaO₂ ≤ 85% was noted in 15 patients, of whom 6 died. 142 patients received catheterization during long term FU; vv. collaterals were identified in 56 and closed in 53. A total of 37% (N=94) were cyanotic at last FU. In 47% (N=44) this can be explained by an open fenestration, in 17% (N=16) by vv. collaterals. In 36% (N=34) no reason for cyanosis could yet be found.

Conclusion: Our study shows that a significant proportion of TCPC patients stay or again become cyanotic at long-term FU. More attention should be directed to the development of desaturation during long-term FU to treat and prevent long-term complications.