Serial assessment of somatic and cardiovascular growth in patients within the first decade after Fontan procedure

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Background: Surgical modifications lead to improved long-term outcome after Fontan procedure. The effects of altered hemodynamic condition on somatic and cardiovascular growth in Fontan patients are still unclear. Aim of this study was to investigate the long-term growth regarding body weight, height as well as growth of both pulmonary arteries (LPA/RPA), single ventricle systemic valve annulus and single ventricle end-diastolic diameter (SVDDE) of Fontan patients with an extra cardiac conduit.

Methods: Anthropometric and echocardiographic data of 139 patients undergoing Fontan procedure from 1995 to 2018 at the University Children’s Hospital Zurich were retrospectively analyzed. The first 10 years of somatic and cardiovascular growth were determined by z-scores. The time-point of normalization after Fontan procedure was assessed by a year-by-year comparison of z-scores using Wilcoxon signed rank test.

Results: The median age at Fontan procedure was 2.6 years (inter quartile range: 0.9 years) with a dominant left ventricle in 44% of all patients. In all patients a significantly lower weight and height was observed for the first 6 years of life (median z-score -0.4 to -0.9, p<0.05) compared with a healthy control population; afterwards, anthropometric parameters normalize. Similarly, RPA diameter is lower in the first 5 years of life (median z-score -1.1 to -0.6, p<0.05) and LPA in the first 8 years of life (median z-score -1.3 to -0.5, p<0.05). In contrast, SVDDE is increased in the first 4 years (median z-score 0.5 to 1.3, p<0.05) and normalizes afterwards. The median z-score of the systemic annulus is above 3 within the first 10 years of life and does not normalize. Somatic parameters are strongly correlated with cardiovascular ones over the study period (r >0.8, p<0.05).

Conclusions: Patients after Fontan procedure have a delayed somatic and cardiovascular growth (LPA/RPA), with a catch up in between the 5th to 6th year of life. The initially increased SVDDE normalizes after 4 years, the systemic annulus remains strongly enlarged over the first 10 years of life.