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Results of total correction of complete atrio-ventricular canal (CAVC) depending on coexistence of Down syndrome in children.

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CAVC is predominantly seen in Down syndrome (DS) patients (70%), with correction results in DS and non-DS patients ambiguous due to differences in anatomy and clinical course. The objective was assessment of CAVC correction results and prognosis depending on concomitant DS. Clinical material: 131 patients, with 108 DS - G1 and 23 normal karyotype subjects -G2, operated on from 1990 to 2003. Methods: All patients were subjected to physical examination, ECG, CXR and ECHO. Testing was done postoperatively with the mean follow-up of 9.2 years. Statistical analysis: descriptive statistics, t-Student test, Yates's $\chi 2$ test and Kaplan-Meier survival curve were analyzed. Results: Type A was more common in G2 (69.6% vs. 48.6%, p=0.1), type C in G1 (32.1% vs. 17.4%, p=0.2). Qp/Qs in G2 was significantly higher than in G1 (3.5 vs. 2.4, $p<0.05$), as well as significant common A-V valve regurgitation (AVVR) (34.8% vs. 25%). Age at surgery was x=8.8 months. Postoperatively, significant left AVVR was more common in G2 (17.6% vs 34.8%). Significant residual VSD was equally common (4.6% vs. 3.8%), as was LVOTO (2.8% vs. 4.3%). Eighteen patients (13.7%) died: nine - within 30 days postoperatively and nine - at the mean time of 10.1 months. Mortality rates in G1 were non-significantly higher (14.8% vs. 8.7%, p=0.6), due to infections, CHF and pulmonary hypertension crisis. Early mortality risk factors were younger age at surgery (x=5.4 vs. x=9.0 months, $p<0.05$), lower body mass (x=4.0 kg vs. x=5.6 kg, $p<0.05$) and concomitant PDA (77.8% vs. 27.6%, $p<0.05$). Reoperations included 15 patients (G1: 11.1%, G2: 13%) due to left AVVR, LVOTO and/or residual VSD. The 10-year survival for was 87.3%, being higher in G2 (85.2% vs. 91.3%). Survival without reoperation was seen in 88.5% children, being comparable in both groups (87% vs. 88.9%). Conclusions: CAVC morphology is more favorable for repair in DS children, however higher susceptibility for infections and risk of pulmonary hypertension crisis may affect clinical course and early mortality. 10-year survival and freedom from reoperation are similar in both groups but incidence of significant left AVVR may result in more frequent future re-operations in non-DS patients.