

Outcome after surgical repair of atrioventricular canal defects in patients with trisomy 21

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Introduction: Trisomy 21 is often associated with heart defects and especially of the atrioventricular canal (AV-canal). The aim of the study was the analysis of outcomes after correction of AV-canal defects comparing surgical techniques, complications, and results in patients with and without trisomy 21.

Methods: We performed a retrospective study of 241 patients undergoing surgical repair of AV-canal defects during the years 1986 and 2016. One-hundred twenty-four patients (51%) had had trisomy 21 (group D). The data of these patients were compared to the data of patients without trisomy 21 (group non-D; n=117). Mean follow-up was 113 months.

Results: Twenty-one patients had undergone previous cardiac surgery; there was no significant difference between the groups (10.5% group D vs. 6.8% non-D). Trisomy 21 patients had had more often complete AV-canal defect (77.4% vs. 30% group non-D, $p<0.05$) and more often pulmonary hypertension (78% of patients in group D vs. 29% in non-D group, $p<0.05$). Down-patients were significantly younger at the time of surgery (24.8 ± 56.6 months vs. 51.5 ± 109.1 months in non-D-group) and subsequently smaller. After surgery, Down-patients required a longer period of mechanical ventilation (4.9 ± 8.3 days vs. 1.8 ± 3.2 days, $p<0.001$) and they needed catecholamine therapy for a longer time (2.9 ± 3.5 days vs. 1.5 ± 5.1 days, $p<0.05$). The 30-days mortality was comparable between the two groups (n=9 in group D vs. n=5 in group non-D, $p=0.37$). There was also no difference in the need for re-operation or intervention during follow-up (16.1% group D vs. 12% group non-D, $p=0.35$). The most common indications for re-operation were mitral valve regurgitation and left ventricular outflow tract obstruction. Echocardiographic examination at last follow-up appointment demonstrated a better mitral valve function in patients with trisomy 21.

Conclusions: Outcome after repair of AV-canal defects is very good with low re-operation rate during follow-up. Although patients with trisomy 21 had a more complex defect morphology and were younger at surgery, they have an equal chance for good outcome after surgical correction.