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Outcomes and long-term results of complete atrioventricular septal defect repair in infants with Down syndrome

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Objectives. A retrospective cohort study was made as well as a comparative analysis of the immediate (up to 30 days) and long-term (56±15;12-75 months) results of the repair of complete atrioventricular septal defect (CAVSD) in infants with normal karyotype/chromosome (NK) set and with Down syndrome (DS).
Methods. Surgical correction of congenial heart disease during the first year of life was performed on 593 children (8.3%) with DS in the Bakoulev CCVS. Of this number, 349 infants aged 4.8±2.5 months were diagnosed with atrioventricular septal defect. CAVSD occurred in 279 infants. According to Rastelli's classification, CAVSD of type A occurred in 71%(198/279); of type B, in 14%(40/279); and of type C, in 15%(41/279) infants. CAVSD repair has been performed on 163 patients with DS using the single-patch (n=40) and the two-patch (n=123) methods. The control group consisted of 214 infants aged 6.49±3.03 months with CAVSD and NK.
Results. In infants with DS abnormalities of the left AV valve (doubling of the mitral valve, single papillary muscle, leaflet or chordal dysplasia, etc.) occur as statistically significant (8%DS vs 12%NK;$p<0.05$) which is rarer than in NK children. The presence of DS increases the risk of complications (mainly in the respiratory area) in the early postop. (48%DS vs 63%NK;$p<0.05$) and significant co-morbid conditions in the long-term period of observation. Squeal structures in groups were differentiated. The early postop. in the DS group was characterized as rarer with high-class heart failure (14%DS vs 37%NK;$p<0.05$). CAVSD in the DS group was presented preferentially in the respiratory system combined with infective pathology with basic immunodeficiency (21%DS vs 8%NK;$p<0.05$), which was delimit the postoperative pulmonary ventilation time (DS 5.1±2.8 days vs NK 1.7±0.8 days;$p<0.05$).
Conclusions. The infants with DS and CAVSD, who underwent surgical repair during the first year of life, have a good prognosis. The presence of chromosomal imbalance in them significantly increases the risk of severe co-morbidities that has a significant impact on the duration of the recovery period, as well as the duration of their life even after successful correction of CHD. For these patients, individual training programs are particularly important.