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

Tumanyan MR[1,2], Filaretova OV[2], Chechneva VV[1], Levchenko EG[1], Gulasaryan RS[1], Butrim YV[1], Bokeria LA[1,2]

Bakoulev SCCVS, Moscow, Russian Federation
1 Department of Intensive Cardiology for infants with congenital heart disease,
2 The Pediatric Cardiology Course at the Chair of Cardiovascular Surgery

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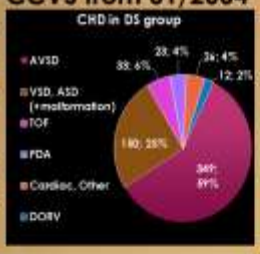
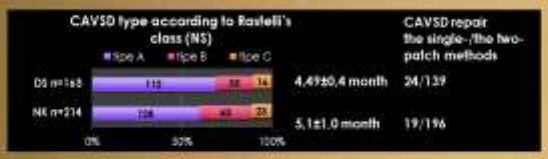
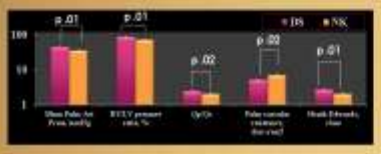
Objectives. A retrospective cohort study was made as well as a comparative analysis of the immediate (up to 30 days) and long-term (56 ±15 months) results of the repair of complete atrioventricular septal defect (CAVSD) in infants with Down syndrome (DS) and normal karyotype/chromosome(NK) set.

Methods. . Surgical correction of congenial heart disease (CHD) during the 1 year of life was performed on 593 children(8.3%) with DS in the Bakoulev CCVS from 01/2004 to 01/2011. Of this number, 349 infants 4.8±2.5 mths. were diagnosed with AVSD. CAVSD occurred in 279 infants, 163 of whom were underwent surgical repair (DS group). The NK group consisted of 214 infants (6.5±3.3 mths) with NK and CAVSD.





Results. In infants with DS abnormalities of the left AV valve (doubling of the mitral valve, single papillary muscle, closely spaced groups of papillary muscles, leaflet or chordal dysplasia, hypoplastic valve ring) occur as statistically significant (5% DS vs 16% NK; p .002) 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 .05) and the risk of 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-dose cardiotonic support (14% DS vs 37% NK; p .01). CAVSD in the DS group was presented preferentially in the respiratory system combined with infective pathology with basic immuno-deficiency (21% DS vs 8% NK; p. 01), which was delimiting the postop. pulmonary ventilation time (DS 5.3÷2.2 vs NK 1.9÷0.6 days; p .05).

Reoperations after repair of CAVDS.
In the early postop. period (up to 30 days) greater proportions of infants with DS, compared with NK, had postoperative complete AV block requiring permanent pacemaker placement (14,1% vs 3,2%; p .0006).
In long-term results there was no significant difference from reoperation after initial repair of CAVSD between DS vs NK (p .13).
In total DS patients were undergoing AV valve repair/replacement rarer then in NK child's (7,5% vs 11,4%; p .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.

prof. Margarita R. Tumanyan MD, DM mrtumanyan@mail.ru & associated prof. Olga V. Filaretova, MD, DM ppcss2004@gmail.com