

**Idiopathic dilatation of right atrium in young children: case report of 7 successful repairs.**

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Objectives: idiopathic dilatation of right atrium is an extremely rare congenital heart disease which is characterized by significant increase of right atrium in absence of other heart malformations.

The aim of our study was to present successful right atrium enlargement repair in 7 young children.

Methods. 7 children with idiopathic dilatation of right atrium (RA) was operated at the median age from 1 month to 5 years, median age 5 months, Interquartile Range (IQR) 14 months, BSA 0,385 m<sup>2</sup> (IQR 0,43) in Bakoulev SCCVS from 01.2005 to 12.2013. Examination included routine physical cardiovascular examination and X-ray finding, ECG with 12 leads and 24 hours monitoring, contrast-enhanced MSCT, EchoCG and angiography.

Results. Right atrium enlargement was diagnosed on 30-32 gestation age (weeks) in four of seven infants, two of them were newborns. Heart failure was preponderous in the clinical picture for all patients, and right atrium and ventricle overload was diagnosed by 12 lead ECG and X-ray finding. NYHA class was 3 (IQR 1). Cardio-thoracic index ranged from 0,57 to 0,91 (median 0,74, IQR 0,12). Arrhythmia in the form of paroxysmal atrial flutter was up to 300 beats/min in 1 of 7 patients and one of 7 infants was diagnosed with supraventricular trigeminy. According to MSCT findings RA volume varied from 145 ml/m<sup>2</sup> to 322 ml/m<sup>2</sup> (median 215,7 ml, IQR 113,7). Partial resection of right atrium to the extent of normal tissue by cardiopulmonary bypass during moderate hypothermia was performed to all patients. Patient mortality was 0%. Morphological examination of resected RA portion uncovered high-grade dysplasia of atrial myocardium with diffuse thinning, endocardial fibroelastosis and local muscle layer atrophy. One of 7 cases was accompanied with calcareous degeneration patch. Three of seven children were under long-term follow-up from 3 months and up to 4 years (median 16 months, IQR 24). Late outcomes were successful.