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EBSTEIN'S ANOMALY AND PATIENT SURVIVAL

Introduction. Ebstein's anomaly (EA) is a heart disease which is considered to be complex and rare in which is involved deformity of the tricuspid valve (TV) along with myopathy of the right ventricle (RV), with variable anatomic and pathophysiologic appearances, thus leads to correspondingly variable clinical consequences. Observation and Medical administration is recommended for patients of asymptomatic type, and may be successful for many years. An adulthood first time appearance of EA is common, with reduced natural history survival exhibition accompanied by biventricular failure [1, p. 28].

Aim. To systematize the available data on the frequency of occurrence, hemodynamic features, rates of successful correction, and outcomes of late discovered «Ebstein's anomaly» operation.

Methods. Analyze systematic reviews, numerous scholar studies, scientific resources and researches results regarding the «Ebstein's anomaly».

Results. Ebstein's disease is counted a rather uncommon pathology, the occurrence of which between all congenital heart defects does not go beyond 1%. This pathology happens in 1 case in 20,000 neonates. On late operative results for EA, one of the biggest published series included the following: overall late survival was 98%, 94%, 90%, 86%, and 76% at 1, 5, 10, 15, and 20 years, correspondingly. Freedom from late reoperation was 97%, 91%,

82%, and 70% at 1, 5, 10, and 15 years, correspondingly [5, p. 139]. Results of Late cone reconstruction are restricted on account of the relatively recent application of this surgical strategy; however, adult-specific freedom from reoperation subsequent to cone reconstruction at 6 years is 98.8% [3, p. 165]. Mitral regurgitation requiring a surgical intervention, RV outflow tract obstruction (RVOTO), elevated haematocrit indicating preoperative cyanosis, > moderate RV dysfunction, and \geq moderate LV dysfunction, are all included as an Independent predictors of late mortality [2, p. 1].

Repair techniques of various type, all of which preserve the atrialized chamber and are used exclusively according to morphology, provide pleasing long-term ventricular function and functional outcome even in severe types of EA.

Factors of risk for a worse outcome included right, and/or left ventricular systolic dysfunction; raised hemoglobin/hematocrit values; male gender; right ventricular outflow tract blockade; or hypoplastic pulmonary arteries [4, p. 1].

Conclusion. In conclusion, our results indicate that EA is of higher survival rates treated in early life stages rather than in adulthood. Management is complex and must be personalized. Detailed knowledge about the diverse anatomic and hemodynamic variables, associated malformations, and management options is critical. Thus, it is important that patients with EA be evaluated regularly by a cardiologist who has expertise in congenital heart disease [6–7].

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