Long term follow up of patients with Ebstein’s anomaly

Hidvégi E.(1), Környei L.(2)
Dr. Jakab &Co Ltd. Pediatric Cardiology, Szolnok, Hungary (1)
Pediatric Heart Center, National Heart Institute, Budapest, Hungary (2)

Introduction Ebstein’s anomaly (EA) is a rare congenital cardiac anomaly characterised by downward displacement of the septal and posterior leaflet of tricuspid valve (TV), which may result in cyanosis, right heart failure and tachyarrhythmia during the newborn period or after adolescence. Due to the rarity of the diseases, the data regarding life-expectancy are still limited, especially in a small country.

Aim To define the demographic parameters, clinical manifestations and long-term outcomes in Hungarian cohort of EA.

Patients and Methods The downward displacement of the TV from the atrioventricular ring ≥ 0.8 cm/m² body surface area was set as the criterion for EA. Patients with EA+congenitally corrected transposition of the great arteries were excluded. Data of 32 (20 male) patients with EA born between January 1994 and December 2012 in Hungary were studied.

Results The frequency of EA during this period was 1.67/100,000 life birth. The clinical signs leading to the diagnosis were: abnormal echo scan in foetus (5), cardiac murmur (13), cyanosis (7), dyspnoe (7), heart failure (6), extracardiac malformation (4) in the perinatal period (all of 26 cases), and cardiac murmur (3), cyanosis (1), dyspnoe (1), cardiac arrythmia (1) in childhood (all of 6 cases). Average gestational age was 38.5±2.2 weeks, birth weight was 3280±667 g. Extracardiac malformations occur in 10 cases. The follow-up time was 87,0 months (1-218). Majority of the associated cardiac anomalies disappeared spontaneously during the follow-up period: ASD-II (28-11), PDA (9-0), VSD (2-0), PS (6-6), PA (1-1). The QRS duration increased from 76.9±17.4 (58-120) ms to 98.4±24.1 (60-161) ms, occurrence of incomplete RBB increased from 10 to 13, complete RBB from 2 to 10, preexcitation from 1 to 3, respectively, during the follow-up period. It happen 2 cardiac intervention, 1 RF ablation, in 8 cases one cardiac operation, in 4 cases two, and in 3 cases three. There were 3 death (49,6, 6-108 months) related to the EA.

Conclusion The occurrence of EA in Hungary in this period is higher, the ratio of extracardial anomalies is higher, the complexity and the mortality is much more lower, than in the literature.