Ebstein’s anomaly management in Central and South Eastern European (CESEE) countries: current status and improvement needed

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Background: Ebstein anomaly (EA) of the tricuspid valve (TV) as a rare congenital heart disease (CHD) is characterized by variable morphology, hemodynamics and clinical course, associated heart defects as well. Moreover, whether and when to intervene on TV remains a big challenge. Patients with EA should therefore be managed in expert centres equipped with all diagnostic and treatment tools according to ESC guidelines (ESC-GL), including cardiac magnetic resonance (CMR) as the gold standard.

Aim of the study: The aim of the study was to characterize EA patient population from selected countries within Central-Southern-Eastern Europe (CESEE), evaluate the management in relation to ESC-GL and determine optimal parameters for long-term follow-up (FU) feasible in CESEE.

Material and methods: In retrospective cross-sectional study from 9 CESEE countries, 203 adult patients with EA were included: median age 39y, 58% women. Determined were age at diagnosis, EA complications (associated CHD, arrhythmias), cardiosurgery. ECHO and CMR right ventricular (RV) characteristics, NTproBNP, 6-minute walking distance (6MWD) were analysed as follows: ECHO parameters: long-axis RV end-diastolic dimension (RVEDD, mm), long-axis end-diastolic right to left ventricle diameter ratio (RV/LV), tricuspid annular plane systolic excursion (TAPSE, mm), RV fractional area change (RV FAC, %), RV tissue Doppler myocardial velocity (m/s). CMR-derived analysed parameters were: ejection fraction (EF, %), indexed end-diastolic volume (EDVi, ml/m²).

Results: Patients were diagnosed at age <1y/1-18y/>18y in: 29.6%/31.1%/39.3%. NYHA I/II/III/IV was as follows: 41.6%/37%/22.5%/0%. Median NTproBNP: 160ng/l, 6MWD: 473m. CMR-characteristics were: median RVEF: 43.2%, EDVi: 118ml/m². CMR was absent in 60% cases. Associated CHD/arrhythmias were present at 46.7%/64.4. TV surgery were indicated / underwent 30.7% patients at mean age 32y. Performed was daSilva cone-repair/other valvuloplasty/valve replacement in 36.4%/25.5%/38.2%. CMR-derived EDV correlates significantly (P < 0.001) with ECHO-based RVEDD and RV/LV ratio. No correlations between CMR-derived EF and ECHO functional parameters were found.

Conclusion: Although patients with EA in CESSE region are diagnosed late, the characteristics correspond to data published so far. Despite large heterogeneity including various associated complications, patient’s care in the CESEE region does not fully meet the requirements of ESC-GL. Mainly, CMR as a golden standard for EA evaluation is not widely accessible and so not performed. Therefore, an inconsistency in management and differences in therapeutic approach is a consequence. In these conditions in view of our previously proven experience, that the RV/LV ratio correlates significantly with CMR volume parameters, ECHO could serve as a reliable and decisive imaging method in FU, however insufficient in relation to ESC-GL.