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


1The National Institute of Cardiovascular Diseases, ACHD Centre, Departement of Cardiology and Angiology of Medical Faculty, Slovak Medical University, Bratislava, Slovakia
2The National Institute of Cardiovascular Diseases, ACHD Centre, Departement of Cardiology and Angiology of Medical Faculty, Slovak Medical University, Bratislava, Slovakia
3Gottsegen Gy Hungarian Institute Of Cardiology Budapest, Department of Cardiology, Budapest, Hungary
4John Paul II Hospital, Jagiellonian University Medical College, Institute of Cardiology, Department of Cardiac and Vascular Diseases, Krakow, Poland
5University Medical Centre of Ljubljana, Adult Congenital Heart Centre, Ljubljana, Slovenia
6Motol University Hospital, Cardiovascular Centre, Prague, Czechia
7Institute for Cardiovascular Diseases Dedine, Department of Noninvasive Cardiology, Belgrade, Serbia
8St. Ekaterina Hospital, Department of Cardiology, Sofia, Bulgaria
9University Medical Centre of Ljubljana, Adult Congenital Heart Centre and National Centre for Pulmonary Hypertension, London, United Kingdom of Great Britain & Northern Ireland
10Institut of Cardiology, Ukrainian Children’s Cardiac Centre, Kyiv, Ukraine
11Paul Stradins Clinical University Hospital, Riga, Latvia
12Royal Brompton Hospital, Adult Congenital Heart Centre and National Centre for Pulmonary Hypertension, London, United Kingdom of Great Britain & Northern Ireland
13University hospital Münster, Division of Adult Congenital Heart Disease, Department of Cardiovascular Medicine, Muenster, Germany

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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 (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, 6MWDV: 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.