

I. Csecs<sup>1</sup>, C. Czibalmos<sup>1</sup>, A. Toth<sup>1</sup>, F.I. Suhai<sup>1</sup>, Z. Dohy<sup>1</sup>, M. Toth<sup>2</sup>, Z. Pozsonyi<sup>3</sup>, A. Vereczkei<sup>3</sup>, B. Merkely<sup>1</sup>, H. Vago<sup>1</sup>, <sup>1</sup>Semmelweis University Heart Center - Budapest - Hungary, <sup>2</sup>Semmelweis University, 2nd Department of Internal Medicine, Semmelweis University, Budapest, Hungary - Budapest - Hungary, <sup>3</sup>Medical Center of Budapest, 3rd Department of Internal Medicine, Semmelweis University, Budapest, Hungary - Budapest - Hungary:

## Cardiac fingerprints of histiocytic infiltration - Common cardiac characteristics in a diverse, multisystemic histiocytic disease - Single center data over a 12-year period

### Abstract: P6303

#### Cardiac fingerprints of histiocytic infiltration - Common cardiac characteristics in a diverse, multisystemic histiocytic disease - Single center data over a 12-year period

**Authors:**

I. Csecs<sup>1</sup>, C. Czibalmos<sup>1</sup>, A. Toth<sup>1</sup>, F.I. Suhai<sup>1</sup>, Z. Dohy<sup>1</sup>, M. Toth<sup>2</sup>, Z. Pozsonyi<sup>3</sup>, A. Vereczkei<sup>3</sup>, B. Merkely<sup>1</sup>, H. Vago<sup>1</sup>, <sup>1</sup>Semmelweis University Heart Center - Budapest - Hungary, <sup>2</sup>Semmelweis University, 2nd Department of Internal Medicine, Semmelweis University, Budapest, Hungary - Budapest - Hungary, <sup>3</sup>Medical Center of Budapest, 3rd Department of Internal Medicine, Semmelweis University, Budapest, Hungary - Budapest - Hungary,

**Topic(s):**

Tumors of the heart

**Citation:**

European Heart Journal ( 2017 ) 38 ( Supplement ), 1349

Erdheim-Chester disease (ECD) is a rare, multiorgan non-Langerhans histiocytic disorder, the exact prevalence of the disease is unknown and the cardiovascular manifestation could be underestimated due to the occult nature of the signs and symptoms. Cardiac magnetic resonance imaging (CMR) is a standard technique to assess and characterize a suspected cardiac tumor, because of unrestricted field of view, high temporal resolution and non-invasive tissue characterization.

Between January 2005 and January 2016 at the Heart and Vascular Center of the Semmelweis University we performed 9168 cardiac CMR scans, in 4.5% (412 cases) with the suspicion of intra/pericardiac tumors. We performed our institutional multiparametric CMR imaging protocol to evaluate the signal properties, morphologic characteristics (localisation, size, infiltrative nature, presence of pleural/pericardial effusions) of a suspected cardiac mass. In axial and oblique planes cine movies, T1- and T2-weighted, fat-suppressed triple inversion recovery images were performed. First pass perfusion images, early and late gadolinium enhancement (LGE) images were also performed in the same planes.

During the 12-year-period CMR findings suggested cardiac ECD in four patients (3 male, age: 48.5±12.0 years old). The referring diagnoses were lipomatous hypertrophy of the interatrial septum and right atrial mass evaluated by echocardiography.

In all cases the histiocytes infiltration is mainly affected the right atrial wall, the interatrial septum and the interventricular sulcus (Figure). In half of the cases the aortic wall was also involved and had a special morphology, the coated aorta sign (Patient A and D). Pericardial effusion could also be detected in two cases (Patient B and C). After CMR examination all patients had a histologically proven diagnosis of ECD and underwent a detailed medical check-up looking for extracardiac histiocytic infiltration. Extracardiac manifestations were the following: neurological involvement (n=2) (exophthalmus, chromophobe cell macroadenoma), renal involvement (n=1), multifocal osteolysis (n=1), histiocytic soft tissue mass (n=3) in various localisation (retroperitoneal, gastrointestinal, skeletal muscle). During follow-up (668.4±427.8 days) 3 patients died of the progression of the disease, the overall survival rate was 2 years and 4 months after the diagnosis.

The diagnosis of ECD is challenging and cardiac manifestation suggest poor prognosis. Special morphological CMR findings could raise the suspicion of histiocytic infiltration. Besides single case reports national and international registers are needed for the better understanding of ECD.

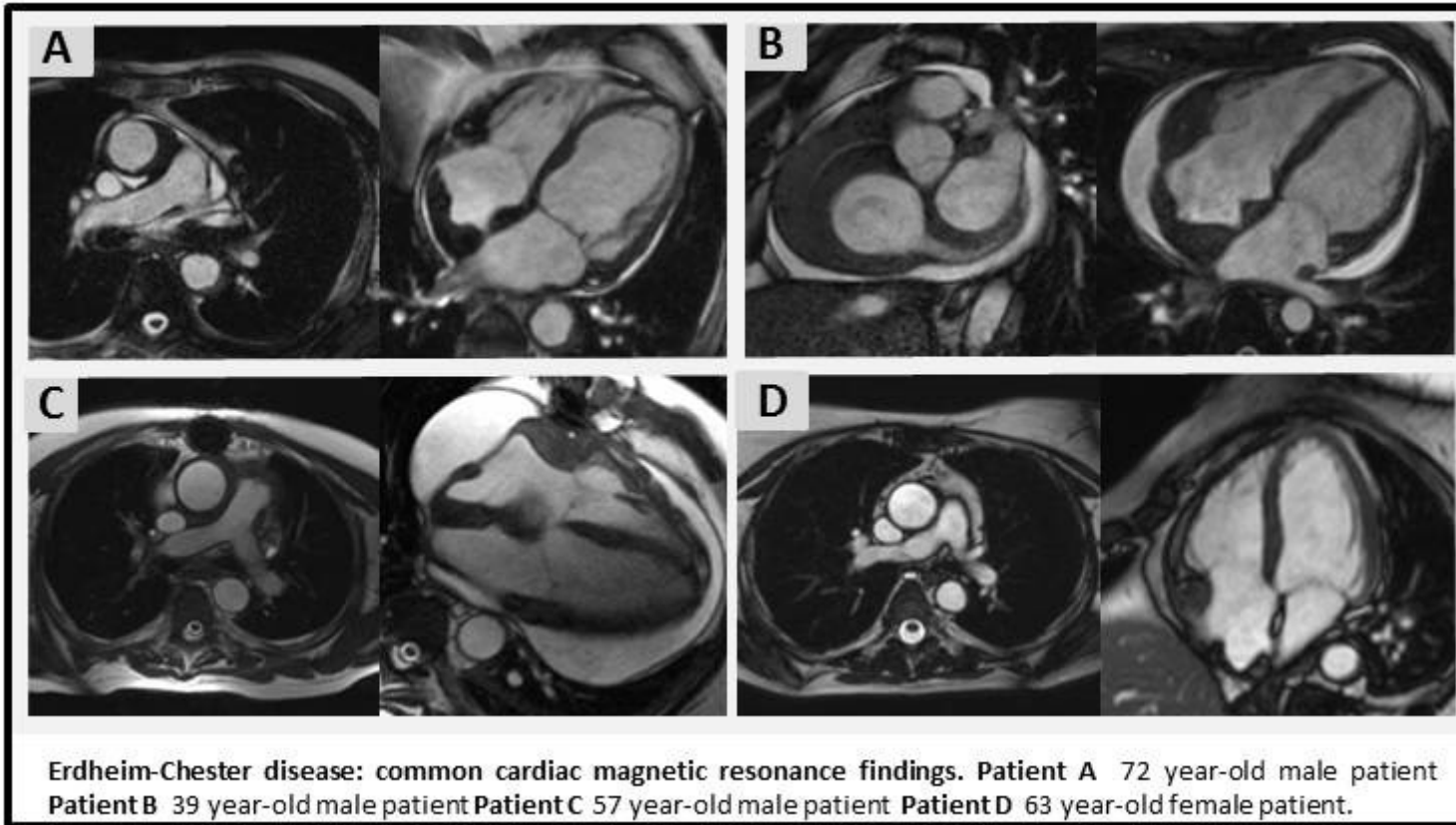


Figure 1