

Fontan Circulation in Adult Patients with Congenital Heart Disease: Results from the CHALLENGE Registry

Apostolopoulou S.C.(1), Ntiloudi D.(2), Giannakoulas G.(2), Vasiliadis K.(3), Frogoudaki A.(4), Ntellos C.(5), Tzifa A.(6), Brili S.(7), Karvounis H.(2), Tsioufis K.(8), Rammos S.(1) on behalf of the CHALLENGE investigators

Onassis Cardiac Surgery Center, Department of Congenital Heart Disease, Athens, Greece (1)

AHEPA General Hospital, Thessaloniki, Greece (2)

General Hospital G. Papanikolaou, Cardiology Department, Thessaloniki, Greece (3)

Attikon University Hospital, Cardiology Department, Athens, Greece (4)

Tzaneio General Hospital of Piraeus, Cardiology Department, Athens, Greece (5)

Mitera General Hospital, Department of Congenital Heart Disease, Athens, Greece (6)

Hippokraton General Hospital, Cardiology Department, Athens, Greece (7)

Hellenic Cardiological Society (HCS), Athens, Greece(8)

Introduction: Patients with structurally or functionally univentricular hearts undergo a series of complicated operations ending, if possible, with creation of palliative Fontan physiology, associated with several cardiac and pulmonary sequelae.

Methods: The purpose of this study was to describe the population and clinical characteristics of adult patients with Fontan circulation included in the national registry of adult congenital heart disease (ACHD) in Greece (CHALLENGE registry) from February 2012 until November 2017.

Results: Within the 2320 total patients included in the CHALLENGE registry from 12 centers, we identified 68 patients (3%) with Fontan type circulation. Age at inclusion in the registry was 26.1±9.5 years, 40% of patients were males, while 45 (66%) had a dominant left and hypoplastic right ventricle. Overall, 30 (44%) of patients had had 1 and 29 (43%) >1 surgeries prior to the Fontan operation, while 34 (50%) had undergone at least one percutaneous intervention. Clinically, 26 (38%) of patients were in WHO class I, 25 (37%) in class II, 14 (21%) in class III and only 3 (4%) in class IV. The majority (84%) of patients received medications, 29% antiarrhythmic, 63% anticoagulant, 53% anticongestive and 6% pulmonary arterial hypertension therapy.

Conclusions: Fontan type circulation patients in our national ACHD registry are in their 2nd to 5th decade of life, have had multiple prior operations and percutaneous interventions, are mostly in class I and II, the majority receiving medications. Collection of additional data such as laboratory, echocardiographic, magnetic resonance imaging and exercise stress test data as well as long follow-up will help to further describe this population and possibly identify important factors for the management and prognosis of this population.