Long term outcome of single ventricle physiology with pulmonary restriction not undergoing Fontan repair

Authors:
FB Francisco Buendia¹, BG Gordon Ramirez², PG Pastora Gallego³, JMO Jose Maria Oliver⁴, SM Silvia Montserrat⁵, AO Ana Osa¹, BM Berta Miranda², MJR Maria Jose Rodriguez Puras³, AG Ana Elvira Gonzalez⁶, BC Barbara Carbonell⁵, PM Pablo Meras⁶, AA Alejandro Aduar³, JR Jose Ruiz Cantador⁶, JR Joaquin Rueda Soriano¹, LD Laura Dos², ¹University Hospital La Fe, Adult Congenital Cardiac Unit. Hospital Universitari i Politècnic La Fe, Valencia, Spain - Valencia - Spain, ²University Hospital Vall d'Hebron, Integrated Hospital Vall d'Hebron-Hospital Sant Pau Adult Congenital Heart Disease Unit - Barcelona - Spain, ³Complex Public Hospital Virgen del Rocio Regional - Seviela, Spain - Spain, ⁴University Hospital Gregorio Maranon - Madrid - Spain, ⁵Hospital Clinic de Barcelona - Barcelona - Spain, ⁶University Hospital La Paz - Madrid - Spain,

Topic(s):
Congenital Heart Disease – Clinical

BACKGROUND

Patients with univentricular physiology who do not complete the palliation to Fontan are a heterogeneous group with unknown long term outcome.

AIMS

This study aimed at describing the clinical course and long-term survival of patients with SV physiology with restricted pulmonary flow that had not undergone a Fontan type of repair.

METHODS

From the prospectively maintained databases of the adult congenital cardiac units of five tertiary referral centers, data from all SV physiology patients were obtained. Patients completing a Fontan type palliation or developing Eisenmenger physiology and segmental pulmonary hypertension were excluded. Baseline data were recorded on the first visit at adult congenital heart disease (ACHD) unit. The primary end point was death.

RESULTS

101 patients (50.5% females) were identified. Mean age at end of follow up was 39.3 ± 11.3 years. Of these, 45 (44.6%) were unoperated (group 1, restricted forward pulmonary flow with or without pulmonary banding), 38 (37.6%) had undergone a cavopulmonary shunt as a definitive palliation (group 2) and 18 (17.8%) had aortopulmonary shunts (group 3). The main diagnosis was double inlet left ventricle (DILV) (N: 52, 51.5%) and most of the ventricle was left (82.2%). The principal reason for not performing a Fontan repair was mean pulmonary artery pressure >18 mmHg. At initial visit at the ACHD unit patients were 32.2 ± 11.1 years of age. 35% of the patients were in NYHA class III-IV, with no differences between groups. However, patients in group 2 had worse oxygen saturation (p=002) and higher haemoglobin (p=0.037). After a mean follow-up of 7.3 ± 4.1 years, mortality was 20.8% (21 patients), being sudden death (7p, 6.9%) the most frequent cause. Patients in group 3 showed worse ventricular function (p=0.0001) and a trend to higher mortality that did not reach statistical significance (HR 2.7, CI 95% 0.91-8.14, P=0.07).

CONCLUSIONS

Patients with single ventricle physiology not undergoing Fontan repair are a population of high risk, with sudden
Long term outcome of single ventricle physiology with pulmonary restriction not undergoing Fontan repair

Authors: FB Francisco Buendia 1, BG Gordon Ramirez 2, PG Pastora Gallego 3, JMO Jose Maria Oliver 4, SM Silvia Montserrat 5, AO Ana Osa 1, BM Berta Miranda 2, MJR Maria Jose Rodriguez Puras 3, AG Ana Elvira Gonzalez 6, BC Barbara Carbonell 5, PM Pablo Meras 6, AA Alejandro Adsuar 3, JR Jose Ruiz Cantador 6, JR Joaquin Rueda Soriano 1, LD Laura Dos 2,

1 University Hospital La Fe, 2Adult Congenital Cardiac Unit. Hospital Universitari i Politècnic La Fe, Valencia, Spain - Valencia - Spain, 2University Hospital Vall d’Hebron, Integrated Hospital Vall d’Hebron - Hospital Sant Pau Adult Congenital Heart Disease Unit - Barcelona - Spain, 3Complex Public Hospital Virgen del Rocio Regional - Sevilla, Spain - Spain, 4University Hospital Gregorio Maranon - Madrid - Spain, 5Hospital Clinic de Barcelona - Barcelona - Spain, 6University Hospital La Paz - Madrid - Spain.

Topic(s): Congenital Heart Disease – Clinical

BACKGROUND

Patients with univentricular physiology who do not complete the palliation to Fontan are a heterogeneous group with unknown long term outcome.

AIMS

This study aimed at describing the clinical course and long-term survival of patients with SV physiology with restricted pulmonary flow that had not undergone a Fontan type of repair.

METHODS

From the prospectively maintained databases of the adult congenital cardiac units of five tertiary referral centers, data from all SV physiology patients were obtained. Patients completing a Fontan type palliation or developing Eisenmenger physiology and segmental pulmonary hypertension were excluded. Baseline data were recorded on the first visit at adult congenital heart disease (ACHD) unit. The primary end point was death.

RESULTS

101 patients (50.5% females) were identified. Mean age at end of follow up was 39.3 ± 11.3 years. Of these, 45 (44.6%) were unoperated (group 1, restricted forward pulmonary flow with or without pulmonary banding), 38 (37.6%) had undergone a cavopulmonary shunt as a definitive palliation (group 2) and 18 (17.8%) had aortopulmonary shunts (group 3). The main diagnosis was double inlet left ventricle (DILV) (N: 52, 51.5%) and most of the ventricle was left (82.2%). The principal reason for not performing a Fontan repair was mean pulmonary artery pressure >18 mmHg. At initial visit at the ACHD unit patients were 32.2 ± 11.1 years of age. 35% of the patients were in NYHA class III-IV, with no differences between groups. However, patients in group 2 had worse oxygen saturation (p=0.02) and higher haemoglobin (p=0.037). After a mean follow-up of 7.3 ± 4.1 years, mortality was 20.8% (21 patients), being sudden death (7p, 6.9%) the most frequent cause. Patients in group 3 showed worse ventricular function (p=0.0001) and a trend to higher mortality that did not reach statistical significance (HR 2.7, CI 95% 0.91-8.14, P=0.07).

CONCLUSIONS

Patients with single ventricle physiology not undergoing Fontan repair are a population of high risk, with sudden death as main driver of mortality. Patients palliated with aortopulmonary shunts are prone to worse ventricular function and a trend to higher mortality.