

Congenital Heart Defect Specific Volumetric Data in Children with Hypoplastic Left Heart Syndrome Measured by CMR

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→ BACKGROUND

For each congenital heart defect (CHD) knowledge of morphological and functional variations is crucial to assess the individual case and initiate specific therapy, if necessary. Data for a specific CHD condition are widely lacking. CMR-based values exist only for left ventricular (LV) and right ventricular (RV) volumes of children and adults with normal anatomy. Therefore, we sought to provide reference values for RV volumes, mass, and function in children with Hypoplastic Left Heart Syndrome (HLHS).

→ METHODS

We performed a CMR study in 66 children (Age 6.4 2.3-14.2y) with HLHS were evaluated after surgical completion of Fontan-circulation in a single center. Seven Children with hemodynamic relevant valvular lesions were excluded. All examinations were performed in freely breathing deeply-sedated children. Short axis planes as a stack of at least 10 contiguous, parallel slices covering the full length of the ventricle were acquired as fast field echo cine images (TR/TE/α=1.1/1.6/60; FOV: 240x260, Achieva 3T, Philips, Leiden, Netherlands). With dedicated software (ViewForum, Philips, Leiden, Netherlands) RV volumes and mass were calculated in the end-systolic and end-diastolic phases of each slice (see fig. 1). Statistical analysis was performed with MedCalc (MedCalc Software, Mariakerke, Belgium).

→ RESULTS

Demographic data for the patient cohort are summarized in table 1. Average volumetric data (mean±SD) for all children splitted in males and females are summarized in table 2. We found the majority of patients (n=45) in the age group from 3 to 8 years. In order to rule out potential age related confounding factors we displayed the results for that group separately. The end-systolic (ESV), end-diastolic (EDV) and stroke (SV) volume, as well as the myocardial mass (MM) were indexed to body surface area. No significant gender specific differences were found in young HLHS patients except for the MM of the RV.

Compared to normal values from healthy children from the literature^{1,2} and from our own data base, children with HLHS have a higher myocardial mass (p<0.0001), a larger end-systolic volume (p=0.03), a lower ejection fraction (EF, p=0.01) and a lower cardiac index (CI, p=0.04) of the systemic ventricle. All remaining parameters, especially the end-diastolic volume and the stroke volume are not significantly different compared to age matched control groups.

Intra- and inter-observer variability of the measurements was in the range of 4% and 9% respectively.

→ CONCLUSION

CMR data on ventricular volumes and function specific to patients with HLHS fill an important void in the current literature. These reference values allow improved assessment of an individual patient to assess his congenital heart defect. Additionally, these data may serve as a baseline data for longitudinal studies.

	all (n=59)	male (n=42)	female (n=17)	
Age (years)	5.6 (2.3-14.3)	5.5 (2.3-14.3)	6.2 (3.4-14.0)	ns
Weight (kg)	19 (12-49)	18.9 (12.5-49)	19 (12-40)	ns
Height (cm)	111 (88-158)	111 (88-158)	112 (94-154)	ns
BSA (m ²)	0.76 (0.55-1.47)	0.75 (0.55-1.47)	0.77 (0.57-1.3)	ns
Heart rate (bpm)	81 (41-121)	78 (41-121)	83 (66-115)	ns

Table 1: Demographic data presented as median (range)

		RV-EDV _i [ml/m ²]	RV-ESV _i [ml/m ²]	RV-SV _i [ml/m ²]	CI [l/min/m ²]	RV-EF [%]	RV-MM [g/m ²]
all	Male (n=42)	73 ± 15	34 ± 11	39 ± 9	2.9 ± 1.0	54 ± 8	81 ± 24
	Female (n=17)	68 ± 18	36 ± 15	39 ± 7	3.3 ± 0.8	55 ± 9	68 ± 14
	Difference (%)	7	6	0	12	2	16
	Significance (p)	0.2931	0.5024	0.9893	0.2031	0.6102	0.0380
3 – 8 years	Male (n=32)	73 ± 14	34 ± 10	40 ± 9	3.0 ± 1.0	54 ± 8	79 ± 23
	Female (n=13)	64 ± 18	36 ± 17	38 ± 7	3.2 ± 0.9	56 ± 9	65 ± 12
	Difference (%)	17	6	5	7	4	18
	Significance (p)	0.1039	0.5704	0.6017	0.5197	0.6362	0.0482

Table 2: Average volumetric data (mean +/- SD); RV: right ventricle, EDV: end-diastolic volume; ESV: end-systolic volume; SV: stroke volume; MM: total myocardial mass; CI: cardiac index; EF; ejection fraction; all data indexed to BSA except for EF.

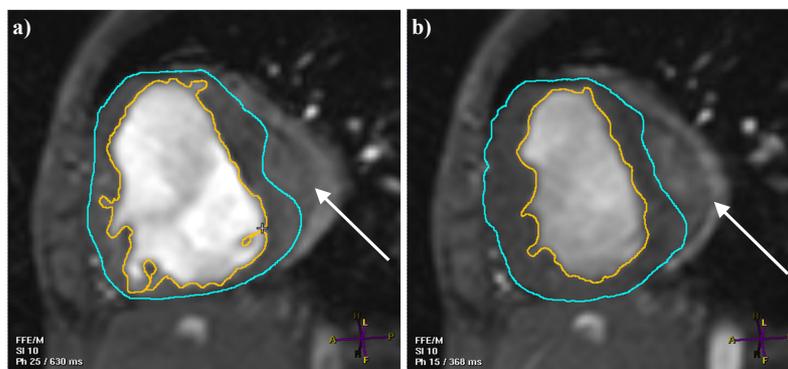


Fig. 1: Volumetric analysis: a) In end-diastolic phase the endo- and epi-myocardial borders are drawn. b) The same is done in the end systolic phase. Note the rudimentary hypoplastic left ventricle in both images (white arrow) as characteristic for HLHS

Lit: 1) Sarikouch S et al., Circ Cardiovasc Imaging. 2010 Jan;3(1):65-76
2) Buechel EV et al., J Cardiovasc Magn Reson. 2009 Jun 21;11:19

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