Morphology, Dimensions, Volumes and Mass

Normal CMR bi-atrial and biventricular reference values in sickle cell disease patients without heart damage

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Background. Cardiac function indices in patients with hemoglobinopathies are different from those in healthy population, mainly due to chronic anemia. Normal reference values specific for SCD patients are not available by CMR.

Aim. We aimed to define the normal cut-off value in SCD patients for bi-atrial and biventricular cardiac magnetic resonance (CMR) parameters.

Methods. We considered forty-eight adult SCD patients with no known risk factors or cardiac disease, normal electrocardiogram, no macroscopic myocardial fibrosis, and all cardiac segments with T2*≥20 ms, consecutively enrolled in the MIOT network (Myocardial iron overload in thalassemia). SCD patients were compared with ninety-six healthy controls and 96 thalassemia major (TM) patients without cardiac damage, both matched for age and gender. Nine pediatric SCD patients were also analysed in comparison with 9 TM patients and 9 healthy subjects matched for age and gender. Cine images were acquired to quantify biventricular function parameters: LV and RV end-diastolic volume (EDV), end-systolic volume (ESV) and stroke volume (SV) were normalized for body surface area (EDVI, ESVI, SVI), as well as biventricular mass and atrial areas. Myocardial iron overload was assessed by segmental T2* technique. Late gadolinium enhancement (LGE) images were acquired for evaluation of macroscopic myocardial fibrosis.

Results. In all three groups males showed higher biventricular volumes and mass indexes than females. SCD male patients had significantly higher LVEDVI ($p < 0.0001$), LVESVI ($p = 0.010$), LVSVI ($p = 0.003$), cardiac index ($p = 0.002$), LV and RV mass index ($p = 0.008$ and $p = 0.001$, respectively) and left and right atrial areas ($p < 0.001$ and $p = 0.011$) than healthy subjects. No significant differences were found in RVEDVI, EVESVI and biventricular EF. Compared to healthy volunteers, females with SCD showed a larger LVEDVI ($p = 0.020$), LVSVI ($p = 0.039$), RV mass index ($p = 0.002$) and left atrial area ($p = 0.008$).

SCD and TM patients showed comparable values of bi-atrial and biventricular volumes and function. When compared to TM, SCD patients showed a larger LV ($p < 0.001$) and RV mass index ($p = 0.001$) in male group and a larger RV mass index ($p = 0.001$) in female group. Table 1 shows the cut-offs for bi-atrial and biventricular MR parameters for adult SCD patients by gender.

No significant differences in MR parameters were found among the pediatric groups.

Conclusions. Normal reference ranges of bi-atrial and biventricular MR parameters for adult males and females SCD patients were established. The use of these reference values will prevent possible misdiagnosis of cardiomyopathy in patients with SCD.

Abstract Figure.

	Mean ± SD	Normal values
Males		
Left atrial area (cm^2/m^2)	13.1 ± 2.7	[7.7 - 18.5]
Right atrial area (cm^2/m^2)	12.6 ± 1.4	[9.8 - 15.4]
LV EDVI (ml/m^2)	101.5 ± 20.7	[60.1 - 142.9]
LV ESVI (ml/m^2)	38.9 ± 12.2	[14.5 - 63.3]
LV SVI (ml/m^2)	62.4 ± 12.3	[37.8 - 87]
LV Mass I (g/m^2)	77.7 ± 11.3	[55.1 - 100.3]
LV EF (%)	62.1 ± 6.9	[55.2 - 69]
Cardiac output (l/min)	7.4 ± 1.9	[5.5 - 9.3]
Cardiac index ($l/min/m^2$)	4.1 ± 1.0	[3.1 - 5.1]
RV EDVI (ml/m^2)	92.6 ± 17.0	[58.6 - 126.6]
RV ESVI (ml/m^2)	33.7 ± 6.8	[20.1 - 47.3]
RV Mass I (g/m^2)	29.0 ± 1.4	[26.2 - 31.8]
RV EF (%)	63.0 ± 5.6	[57.4 - 68.6]
Females		
Left atrial area (cm^2/m^2)	13.5 ± 2.7	[8.1 - 18.9]
Right atrial area (cm^2/m^2)	11.2 ± 1.6	[8.0 - 14.4]
LV EDVI (ml/m^2)	85.0 ± 13.4	[58.2 - 111.8]
LV ESVI (ml/m^2)	30.6 ± 5.9	[18.8 - 42.4]
LV SVI (ml/m^2)	53.9 ± 11.0	[31.9 - 75.9]
LV Mass I (g/m^2)	50.0 ± 8.3	[33.4 - 66.6]
LV EF (%)	63.3 ± 5.6	[57.7 - 68.9]
RV EDVI (ml/m^2)	76.2 ± 14.1	[48 - 104.4]
RV SVI (ml/m^2)	28.0 ± 10.2	[7.6 - 48.4]
RV Mass I (g/m^2)	20.9 ± 7.6	[5.7 - 36.1]
RV EF (%)	64.2 ± 8.1	[56.1 - 72.3]