Journal of Cardiovascular Magnetic Resonance



Poster presentation

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Normalized ranges for right ventricular volumes and function in thalassemia major

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from 13th Annual SCMR Scientific Sessions Phoenix, AZ, USA. 21-24 January 2010

Published: 21 January 2010

Journal of Cardiovascular Magnetic Resonance 2010, 12(Suppl 1):P279 doi:10.1186/1532-429X-12-S1-P279

This abstract is available from: http://jcmr-online.com/content/12/S1/P279

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Introduction

Beta-thalassemia major (TM) patients have a severe anemia requiring lifelong transfusion to allow normal development and prolong survival. Left ventricular volumes and ejection fraction in these patients differ from the normal population because of chronically increased cardiac output, and this can affect the interpretation of scan results. However, normal ranges for RV parameters and function are unknown.

Purpose

To define the normal ranges for RV volumes and ejection fraction (EF) in non-iron overloaded transfusion-dependent TM patients.

Methods

Our study population consisted of 80 transfusion-dependent TM patients (40 males and 40 females) with no evidence of cardiac iron loading (cardiac T2* > 20 ms). Only those over 18 with no evidence of significant cardiopulmonary pathology were included. To control for different iron chelators and possible treatment effect, patients included in the analysis were presenting for their first scan and were only taking a single chelation agent (deferoxamine). Forty age- and sex-matched patients acted as controls. RV volumes and EF were measured from short-axis steady-state free precession cine images using CMRtools (Cardiovascular Imaging Solutions, London). Groups were compared using a two-tailed, two sample t-test.

Results

Both groups were well matched with respect to age and gender but the TM patients (as would be expected) had significantly lower height, weight and body surface area than non-anemic controls (P < 0.05). Body mass index was equivalent in females but slightly lower in male TM patients compared to controls. TM patients had a significantly higher resting heart rate (P < 0.05). Mean RV end-diastolic volume index was higher in TM patients than controls (but this did not reach significance for females). Although the absolute value for mean RV end-systolic volume index was lower in TM patients, there was no significant difference for either gender. In both males and females, the TM patients had a higher RV stroke volume, RVEF, cardiac output and cardiac output index (P < 0.05). See Tables 1, 2, 3 and Figure 1.

Conclusion

Our findings have confirmed that not only LV but also RV 'normal ranges' differ between patients with TM and normal, non-anemic controls. This partly explains why ventricular impairment appears to occur late in iron overload cardiomyopathy. The lower limit of RVEF in TM patients is significantly higher than in normal controls and therefore, if the wrong reference range is used, a cardiomyopathy may exist even though the RVEF appears to be 'normal'. It is important to take this into account when assessing RV function in TM patients.

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Table I: Right ventricular parameters normalised to body surface area for males

Males	TM patients Mean ± SD [95% CI]	Controls Mean ± SD [95% CI]	P value 0.03	
RVEDVI (mL/m²)	97.7 ± 17.1 [92.2-103.2]	88.4 ± 11.2 [83.2-93.7]		
RVESVI (mL/m ²)	33.1 ± 8.0 [30.5-35.6]	33.8 ± 5.0 [31.4-36.1]	0.73	
RVSVI (mL/m ²)	64.4 ± 11.1 [60.9-68.0]	54.7 ± 10.3 [49.9-59.5]	0.002	
RVEF (%)	66.2 ± 4.1 [64.8-67.5]	61.6 ± 6.0 [58.8-64.4]	0.001	
CO (L/min)	7.8 ± 1.9 [7.2-8.4]	6.6 ± 1.6 [5.8-7.4]	0.02	
COI (L/min/m ²)	4.7 ± 1.0 [4.4-5.1]	3.4 ± 0.7 [3.0-3.7]	< 0.001	

RV: right ventricular. RVEDVI: RV end diastolic volume index. RVESVI: RV end systolic volume index. RVSVI: RV stroke volume index. RVEF: RV ejection fraction. CO: cardiac output. COI: cardiac output index.

Table 2: Right ventricular parameters normalized to body surface area for females

Females	TM patients Mean ± SD [95% CI]	Controls Mean ± SD [95% CI]	<i>P</i> value 0.11	
RVEDVI (mL/m²)	86.3 ± 13.6 [81.9-90.6]	80.3 ± 12.7 [74.3-86.3]		
RVESVI (mL/m ²)	29.1 ± 7.2 [26.8-31.4]	30.3 ± 8.6 [26.3-34.3]	0.56	
RVSVI (mL/m ²)	57.3 ± 9.3 [54.3-60.2]	50.0 ± 7.8 [46.3-53.6]	0.004	
RVEF (%)	66.2 ± 5.2 [64.6-67.9]	62.6 ± 6.4 [59.6-65.5]	0.02	
CO (L/min)	6.8 ± 1.7 [6.2-7.3]	5.5 ± 1.5 [4.7-6.2]	0.004	
COI (L/min/m ²)	4.5 ± 0.8 [4.2-4.8]	3.2 ± 0.8 [2.9-3.6]	< 0.001	

Table 3: Patient demographics

	Males			Females		
	TM patients Mean ± SD	Controls Mean ± SD	P value	TM patients Mean ± SD	Controls Mean ± SD	P value
Age (years)	30 ± 8	30 ± 5	0.94	30.4 ± 8.5	30.1 ± 5.0	0.88
Height (m)	1.65 ± 0.1	1.80 ± 0.09	< 0.001	1.54 ± 8.9	1.67 ± 9.2	< 0.001
Weight (kg)	59.1 ± 8.9	75.8 ± 9.9	< 0.001	53.3 ± 10.1	61.4 ± 11.3	0.006
BMI (kg/m²)	21.9 ± 3.1	23.6 ± 3.1	0.048	22.5 ± 4.5	22.0 ± 2.8	0.68
BSA (m ²)	1.64 ± 0.16	1.94 ± 0.15	< 0.001	1.50 ± 0.15	1.68 ± 0.18	< 0.001
Heart rate (min-1)	72.7 ± 10.4	64.0 ± 9.5	< 0.05	78.8 ± 10.1	64.8 ± 13.6	< 0.001

Comparison of indexed RV volumes and ejection fraction for males and females

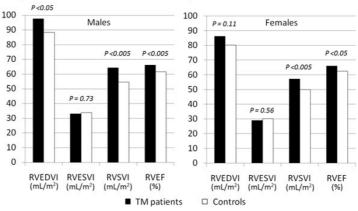


Figure I