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Sickle Cell Anemia Verse Beta-Thalassemia: Cardiovascular Affects of Chronic Anemia by Cardiac Magnetic Resonance Imaging

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Background

Sickle Cell Anemia (SCA) and Beta Thalassemia Major (BTM) are both autosomal recessive hemoglobinopathies which cause chronic anemia. Being in a prolonged anemic state leads to a compensatory increase in plasma volume with an increase in cardiac output and eventual LV dilation with subsequent cardiac dysfunction. Left ventricular ejection fraction (LVEF) is a late finding and is dependent on loading conditions which are altered in chronic anemia. Global longitudinal strain (GLS) is thought to be an earlier marker of cardiac dysfunction and precedes global circumferential strain (GCS) and is abnormal in patients with SCA. It is unclear if patients with BTM have the same response to chronic anemia, therefore we sought to determine the cardiovascular differences in SCA and BTM patients.

Objective

To quantify cardiovascular changes including ventricular volume, function by ejection fraction and myocardial deformation from cine cardiac magnetic resonance (CMR) in patients with SCA versus patients with BTM.

Methods

- Single institution retrospective study of SCA and BTM patients with CMR evaluation.
- GLS and GCS were obtained from long axis views and short axis views using a commercially available CMR-FT software (Medis Medical Imaging, Qstrain 3.2, Leiden, Netherlands).
- Demographic, laboratory and clinical CMR data including indexed right and left ventricular end diastolic, end systolic volume, (LVEDVi and LVESVi), cardiac output (LVCi, RVCi) and function by LVEF as well as liver and myocardial iron (T2*) was collected for statistical analysis using Student's T-test.

CMR-FT Strain Analysis using QStrain

Results

- 27 SCA and 8 BTM patients were identified with complete CMR datasets with no difference in age
- Table 1 shows the laboratory and CMR findings.
- SCA had higher reticulocyte % but lower liver iron and higher T2* and slightly lower hemoglobin than BTM patients.
- Both groups had preserved biventricular but SCA had higher biventricular cardiac output and preserved GCS
- However, SCA patients had significantly lower and abnormal GLS magnitude compared to BTM patients.

Table 1: Demographic, Laboratory and Cardiac MRI Findings in Sickle Cell Disease versus B-Thalassemia

Parameters	Sickle Cell Anemia (N=27)	B-Thalassemia (N=8)	P-value
Age (years)	14.5±5.8	11.7±7.8	NS (0.13)
Hemoglobin (g/dL)	8.9±1.1	9.6 ±1.2	0.06
Reticulocyte %	**11.9±4.8**	**0.9±1.5**	**<0.0001**
Liver iron (mg/g of liver)	6.1±3.4	9.9±4.5	0.06
Myocardial Iron (T2* in ms)*	**38±6**	**28.6±9.9**	**0.003**
Height (cm)	**148.9±27**	**126.7±28**	**0.03**
Weight (kg)	**50.6±22**	**32±17.6**	**0.02**
Body Surface Area (kg/m2)	**1.4±0.4**	**1.0±0.4**	**0.01**
Male/Female	13/27 (48%)	2/8 (25%)	N/A
Indexed LV end diastolic volume (mL/m2	**110.3±20**	**85.9±19.2**	**0.002**
LV ejection fraction (%)	59.5±5.9	60.2±3.7	NS (0.4)
LVCi ($L/min/m^2$)	**5.4±1.5**	**4.4±1**	**0.04**
Indexed RV end diastolic volume (mL/m2)	**110.8±19.3**	**89.5±19**	**0.005**
RV ejection fraction (%)	59.4±6.7	58.3±3.2	NS (0.3)
RVCi ($L/min/m^2$)	**5.4±1.5**	**4.4±0.98**	**0.04**
GLS (%)	**-17.7±3.1**	**-19.9±3.4**	**0.04**
GCS (%)	-20.8±4.1	-22.9±3.4	NS (0.1)

¥ - T2* less than 20 ms is abnormal, normal cardiac index CI is 2.4 to 4 L/min/m2, GLS = Global longitudinal strain, GCS = Global circumferential strain, LVCi = Left ventricular cardiac index, RVCi = Right ventricular cardiac index

Conclusion

- Both SCA and BTM patients have conditions leading to chronic anemia and both responded with increase biventricular cardiac output.
- Compared to BTM patients, SCA patients developed biventricular dilation with preserved ejection fraction.
- SCA patients have occult decline in contractility with abnormal GLS.
- The mechanism of why SCA and BTM responded differently is unclear as both can develop both diastolic and systolic dysfunction.
- The higher cardiac output may explain biventricular dilation in SCA compared to BTM patients. However, future larger and longitudinal multicenter studies are needed to better understand this.