Pulmonary Complications of Sickle Cell Disease. UNIFESP experience.

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São Paulo city: 11 millions

- Estimated:
- 6 to 10 thousand sickle cell disease patients

- *Acute manifestations*

Hemocentro - CIAPH

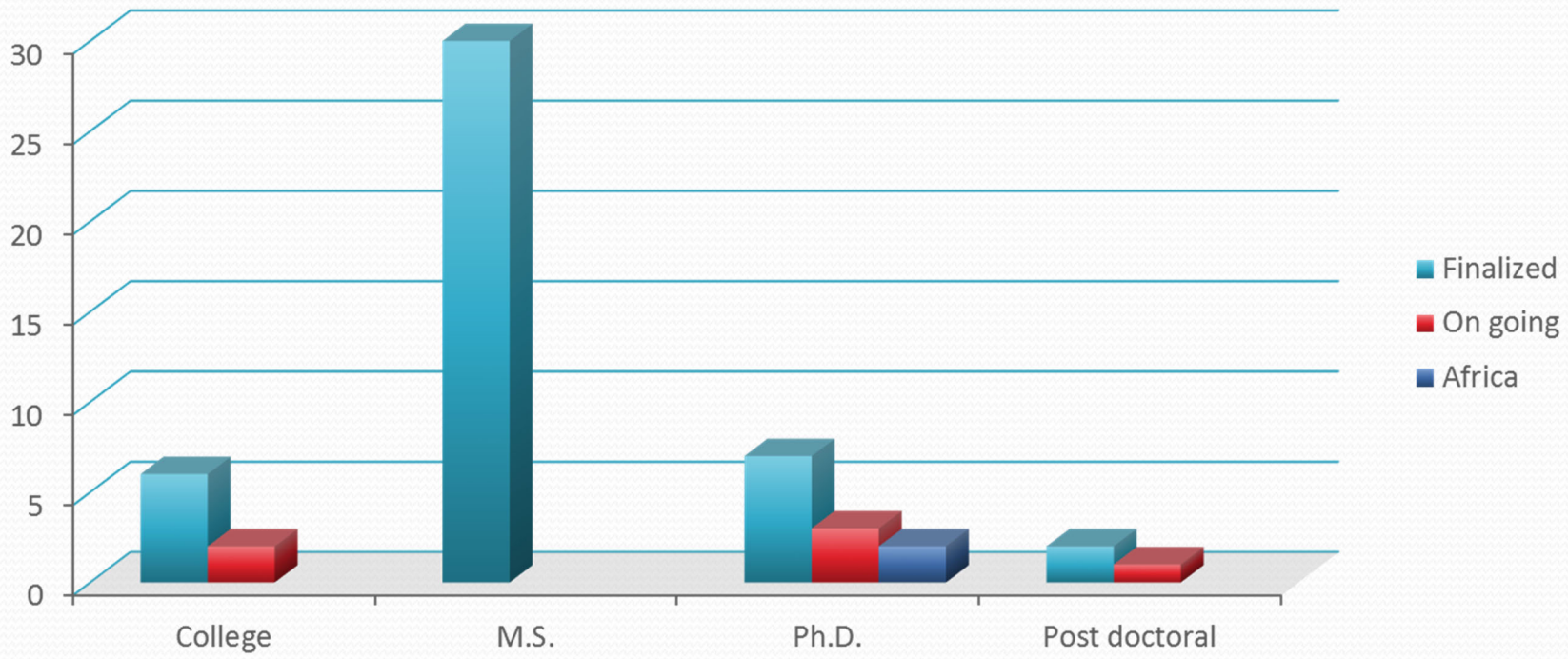
Adults

SCD Phenotype	N (%)
SS	111 (64.5)
Sβ-Thalassemia	8 (4.7)
SC	32 (18.6)
Other Hemoglopinopathies	
CC	9 (5.1)
Cβ-Thalassemia	2 (1.2)
β-Thalassemia	
Major	2 (1.2)
Intermedia	8 (4.7)
TOTAL	**172**

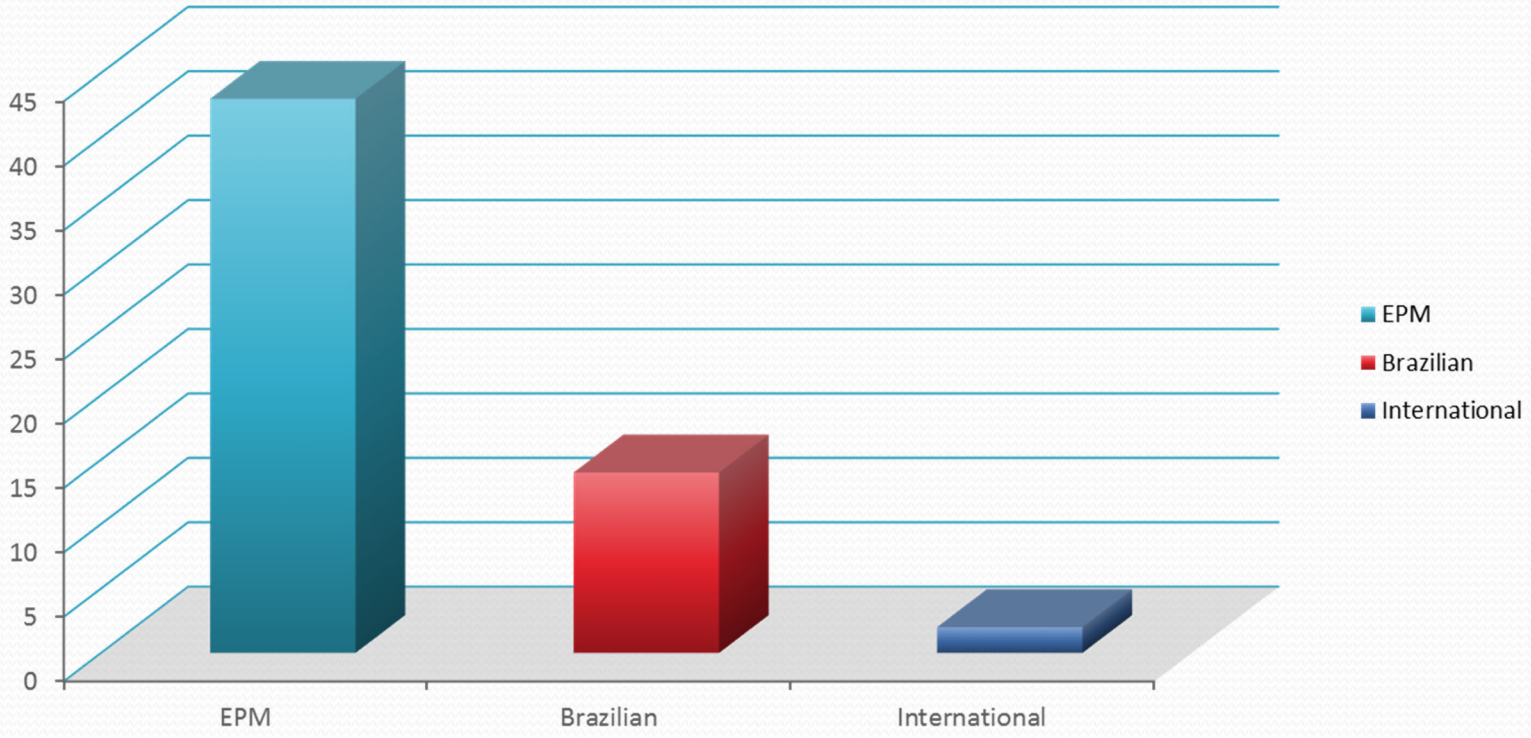
Children & Adolescents

SCD Phenotype	N (%)
SS	214 (48.2)
Sβ-Thalassemia	75 (16.9)
SC	113 (25.5)
Other Hemoglopinopathies	
CC	30 (6.7)
Cβ-Thalassemia	10 (2.3)
β-Thalassemia	
Major	1 (0.2)
Intermedia	1 (0.2)
TOTAL	**444**

Thesis & Research since 1986



Publications since 1989



Pulmonary Complications

First study

Am J Hematol. 2005 Feb;78(2):160-1.

Echocardiographic abnormalities in Brazilian sickle cell patients.

Vicari P, de Cassia Rosario Cavalheiro R, de Gouveia A, Filho Campos O, Figueiredo MS.

Comment on

Echocardiographic abnormalities in sickle cell disease. [Am J Hematol. 2004]

The present study involved 75 SCA patients (hemoglobin SS), 41 females and 34 males, with a mean age of 27.3 years (range 15–74 years).

Conventional echocardiography was carried out in all patients without painful or hemolytic crisis or infection in the previous 4 weeks. The participants were then retrospectively evaluated from January 1980 to October 2003. PH was diagnosed as pulmonary artery systolic pressure values of 30 mmHg or above.

PH being the most frequent complication shown (26%)

In conclusion, we agree with Ahmed et al. [1] that Doppler echocardiography is useful in evaluating the prognosis of these patients. With respect to risk factors for PH in SCA, however, we did not observe a correlation between this complication and infections and acute chest syndrome.

Exercise-Induced Pulmonary Hypertension in Sickle Cell Anemia: a Study with Exercise Stress Echocardiography

- Authors: N Nunes, O Campos, MS Figueiredo, RD Cançado, P Vicari, ACC Carvalho, JO Bordin

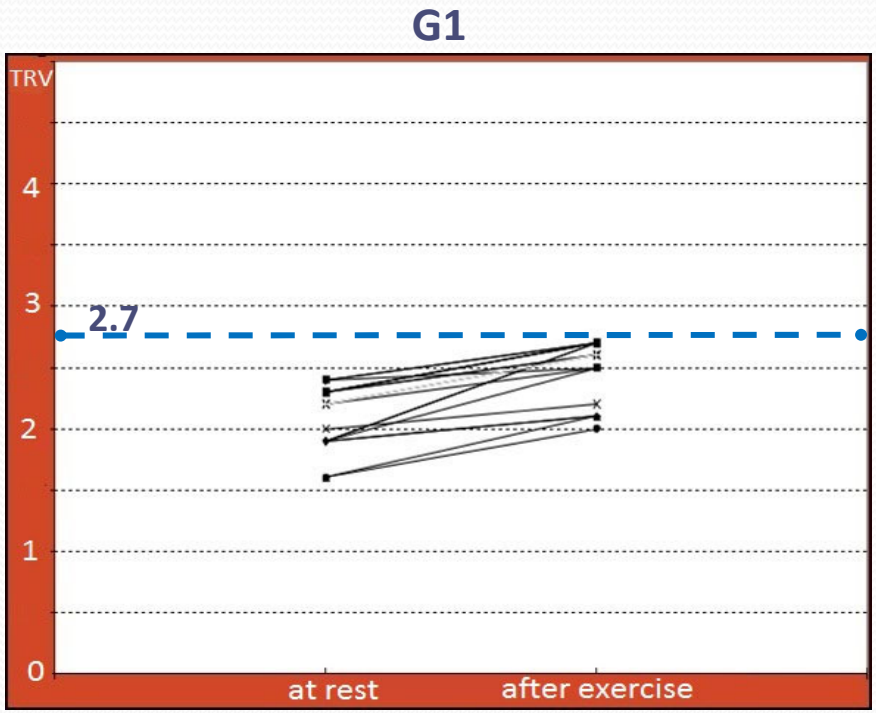
- *Poster at 2008 ASH Annual Meeting*

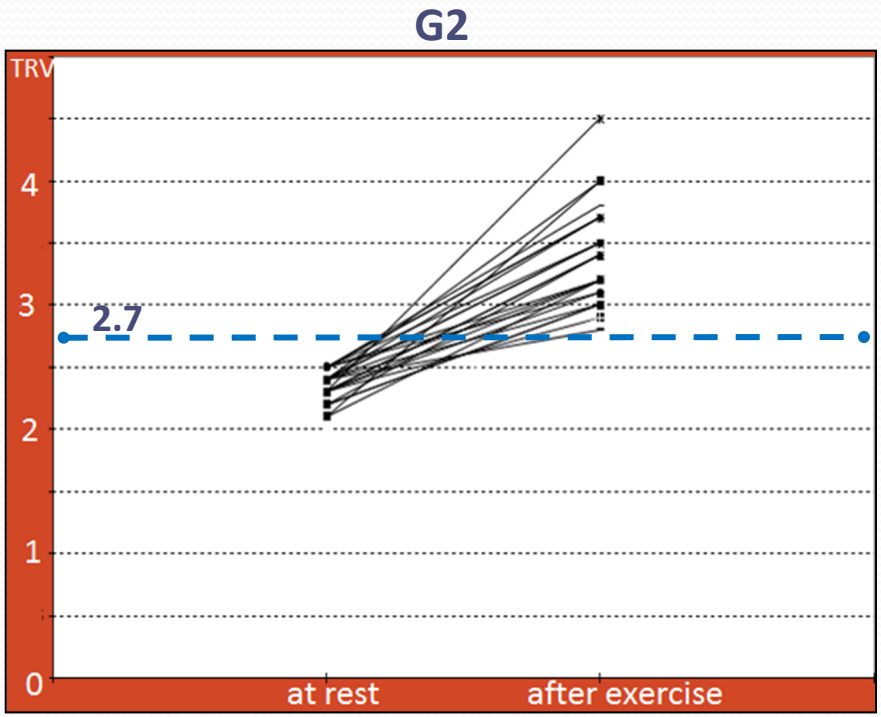
- Exercise treadmill stress echocardiography (ETSE)
  - detect latent PH
  - effective screening tool to assess pulmonary pressure (PP)
- Objectives:
  - To investigate the occurrence of exercise-induced PH in patients with SCA and normal PP at rest
  - To determine its relationship with clinical and echocardiographic variables.

Exercise-Induced Pulmonary Hypertension in Sickle Cell Anemia: a Study with Exercise Stress Echocardiography

- 44 patients with normal PP at rest (TRV < 2.5 m/s).
  - 22 men (50%)
  - Mean age = 25.3 y.o.
- Divided into 2 groups:
  - G1: normal PP after ETSE (TRV ≤ 2.7 m/s)
  - G2: exercise-induced PH (TRV> 2.7 m/s)
- Exercise-induced PH ⇨ 57% of patients (G2)
  - G1: TRV = 2.5 ± 0.3m/s
  - G2: TRV = 3.4 ± 0.4m/s
  - $p < 0.001$

Exercise-Induced Pulmonary Hypertension in Sickle Cell Anemia: a Study with Exercise Stress Echocardiography





Exercise-Induced Pulmonary Hypertension in Sickle Cell Anemia: a Study with Exercise Stress Echocardiography

- Exercise-induced PH was significantly related to
  - higher creatinine levels
  - larger right ventricular diastolic area
  - TRV ≥ 2.3m/s at rest
  - left atrial volume index ≥ 41mL/m^2
  - E/E' ratio ≥ 6.3
- Multivariate analysis: independently related to exercise-induced PH
  - TRV at rest [OR 6.5 (1.3-33.6), p = 0.03]
  - left atrial volume index [OR 6.4 (1.2-33.5), p = 0.03]
  - E/E' ratio [OR 5.6 (1.2-26.5], p = 0.03]

Clinical data

	2005-2007 N (%)	2011-2013 N (%)
Number of patients	107	119
	93 + 14 (Santa Casa)	6.7% S-β thalassemia
Age (years)	24 (13-67)	28 (18-70)
Females	60 (56.0)	72 (60.5)
History of ACS	21 (19.6)	60 (50.4) ?
Use of hydroxyurea	24 (22.4)	80 (67.0)
Chronic transfusion	Not included	9 (7.6)

Echocardiography

	2005-2007 N (%)	2011-2013 N (%)	P value
Echocardiography	99 (92.6)	86 (72.3)	
Estimated PAP	23 (24.0)	73 (84.9)	
VRT determined	0 (0)	52 (60.5)	
PH	29 (27.1%)	11 (9.2%)	NS
	mean (SD)	mean (SD)	
PAP	24.9 (15.2)	27.8 (7.0)	NS

PH ⇨ PAP ≥ 35mmHg

Pulmonar Hypertension

	2005-2007		2011-2013	
	Without PH	With PH	Without PH	With PH
	N = 99		N = 82	
%	70.7	29.3	86.6	13.4
Age (years)	23.5	32	27	31
Range	13 – 48	16 – 67	20 – 60	21 – 70

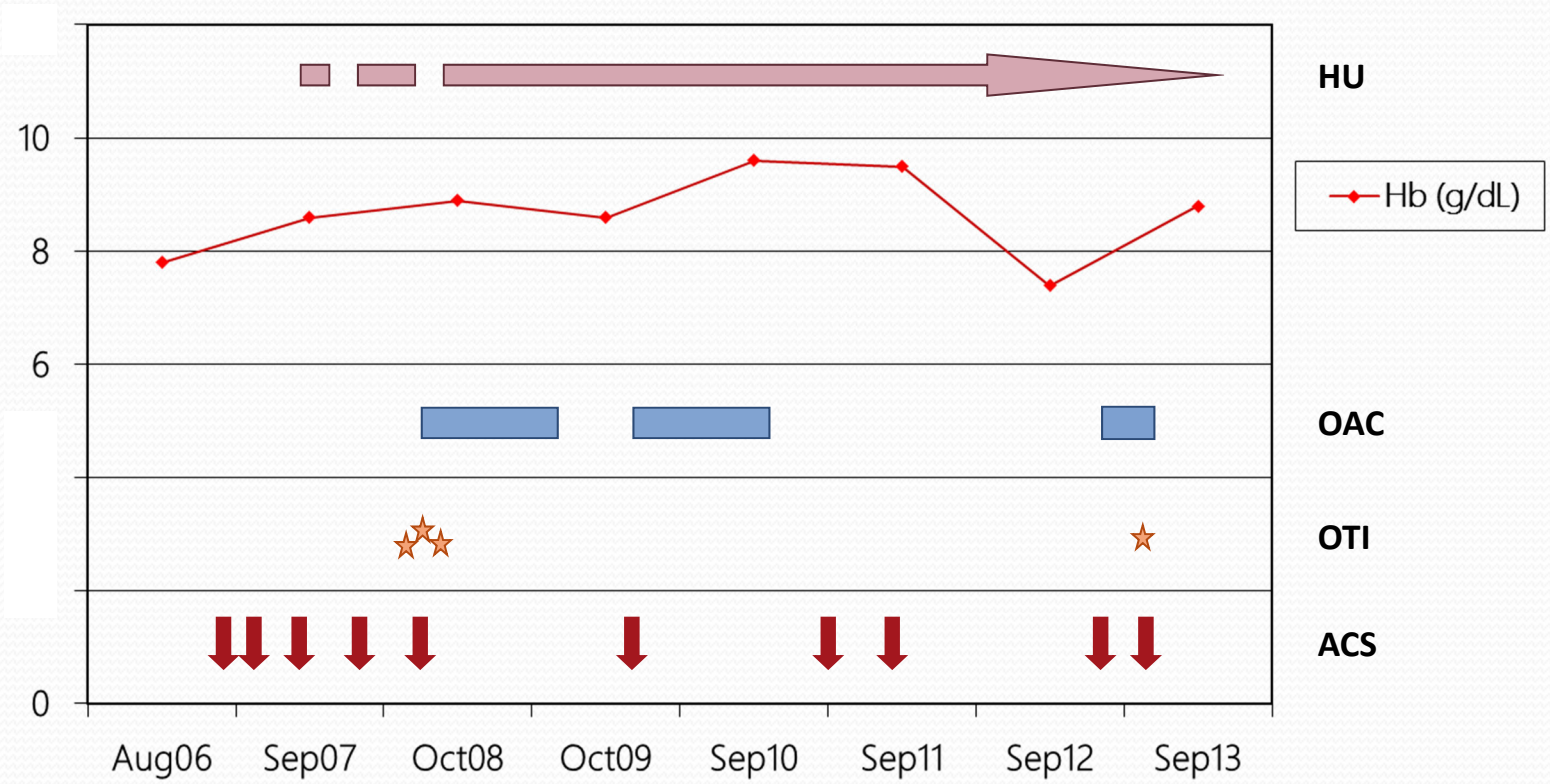
PH ⇨ PAP ≥ 35mmHg

Case Report

CMD, 28 y.o., female

Sickle cell anemia at 2 years of age

Case Report


HU
Hb (g/dL)
OAC
OTI
ACS
10
8
6
0
Aug06
Sep07
Oct08
Oct09
Sep10
Sep11
Sep12
Sep13

Conclusions

- Geographical distribution (patients and Hospitals)

- Improvement of ACS diagnosis (TEP)
- Better relationship with cardiologists
- Improvement PH diagnosis
- Improvement of Clinicians' knowledge about ACS

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