# ENDOCRINE AND METABOLIC COMPLICATIONS IN CHILDREN AND ADOLESCENTS WITH SICKLE CELL DISEASE: AN ITALIAN COHORT STUDY

**E. Bigi[1], P. Bruzzi[2], B. Predieri[2], L. Lucaccioni[3], M. Lodi[3], A. Pancaldi[3], G. Palazzi[1], M. Cellini[1], L. Iughetti[1,2]**

1. Oncology-Hematology Pediatric Unit Department of Medical and Surgical Sciences for Mothers, Children and Adults, University of Modena and Reggio Emilia, Modena, Italy
2. Pediatric Unit, Department of Medical and Surgical Sciences for Mothers, Children and Adults, University of Modena and Reggio Emilia, Modena, Italy
3. Post Graduate School of Pediatrics, Department of Medical and Surgical Sciences for Mothers, Children and Adults, University of Modena and Reggio Emilia, Modena, Italy

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## Introduction and objectives

- Children with Sickle Cell Disease (SCD) show endocrine complications and metabolic alterations.
- The physiopathology of these conditions is not completely understood: iron overload, ischemic damage, and inflammatory state related to vaso-occlusive crises may be involved [1]
- We aim to evaluate the growth pattern and the endocrine and metabolic alterations in a cohort of children with SCD and to detect the relationship between these conditions and the SCD severity.

## Methods

- Study population: 52 patients [38 homozygous (HbSS) and 14 heterozygous (HbSC); age range 3-18 years]
- Anthropometric [height, body mass index (BMI), arm span, sitting height, target height (TH), and pubertal status] and laboratory [blood cell counts, hemolysis indices, metabolic and nutritional status indices and hormonal blood levels] data were evaluated
- The SCD severity was defined according to hematological and clinical parameters.
- Statistical analysis was performed using the program STATISTICA, StatSoft Inc, Tulsa, OK, USA

## Results

### 1. Anthropometric parameters in HbSS patients vs HbSC patients

| Anthropometric parameters | HbSS Mean ± SD | HbSC Mean ± SD | *P-value* |
|---|---|---|---|
| Age (yr.) | 10,44 ± 4,55 | 13,05± 4,47 | *0,08* |
| Height-SDS adjusted for TH (SDS) | 0,3 ± 0,9 | 1,0 ± 0,6 | *0,027* |
| z-score BMI (SDS) | -0,7 ± 1,4 | 0,9 ± 1,1 | *0,004* |

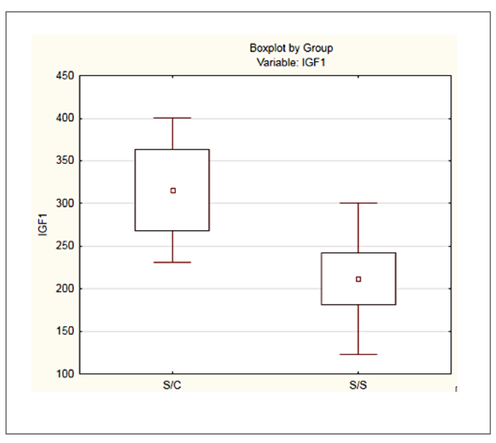
Height-SDS adjusted for TH and z-score-BMI were significantly higher in HbSC children than in patients with HbSS

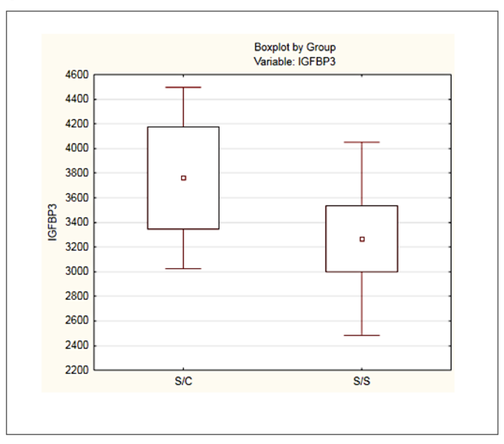
### 2. Prevalence of endocrine and metabolic alterations

| Endocrine/metabolic complications | N°/52 | % | M/F | SS/SC |
|---|---|---|---|---|
| Vitamin D insufficiency (10-30 ng/ml) | 33 | 63.5% | 16/17 | 24/9 |
| Vitamin D deficiency (<10 ng/ml) | 11 | 21.2% | 7/4 | 7/4 |
| GHD | 2 | 3.8% | 2/0 | 2/0 |
| Subclinical hypothyroidism | 2 | 3.8% | 1/1 | 2/0 |
| Hypergonadotopinic hypogonadism | 1 | 1.9% | 1/0 | 1/0 |
| Ovarian insufficiency | 1 | 1.9% | 0/1 | 1/0 |
| Insulin resistance | 6 | 11.5% | 2/4 | 4/2 |

The 92% show at least one metabolic and/or endocrine alteration:
- insufficiency/deficiency of vitamin D (84.7%)
- insulin resistance (11.5%),
- growth hormone deficiency (3.8%),
- subclinical hypothyroidism (3.8%)
- hypogonadism (1.9%)

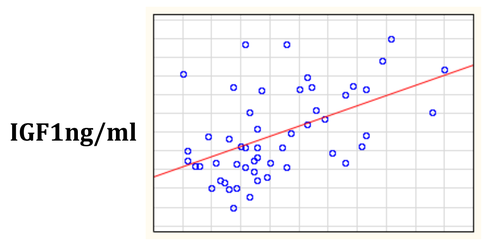
### 4. Values of IGF-1 and IGFBP3 in patients with HbSS and HbSC

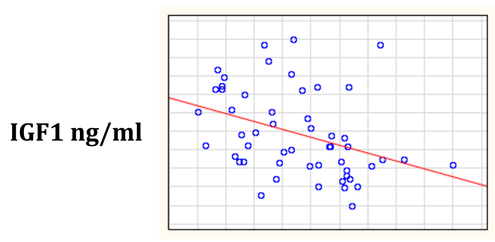



Subjects with HbSS genotype show significant lower levels of IGF-1( 211.7 ± 93.2 vs. 315.3 ± 89.3 ng/ml) and IGFBP3 (3267.1 ± 828, 4 vs. 3761.7 ± 773.5 ng/ml) than children with HbSC

### 5. Relationship between values of IGF-1 and parameters of clinical severity




In the study group IGF-1 values were
- positively related with Hb
- negatively related with lactate dehydrogenase (LDH)

## Conclusions

- Metabolic and endocrine alterations are very common in children and adolescents with SCD
- A regular follow-up is necessary to identify subjects at risk for complications
- An appropriate treatment to improve the outcome disease and the quality of life of SCD patients is necessary

*References:*
*Barden EM, Kawchak DA, Ohene-Frempong K. et al. Body composition in children with sickle cell disease. Am J Clin Nutr 2002;76:218-225 Rees DC, Williams TN, Gladwin MT. Sickle-cell disease. Lancet 2010;376:2018-2031.*

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