Respiratory impairement in children with sickle cell anemia (SCA): differences between the UK and Nigeria

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Background

Acute and chronic respiratory complications are common in subjects with Sickle Cell Anemia (SCA) and can cause longitudinal decline of lung function since childhood[1]. Little is known about lung function in African children with SCA. In this multicenter cross-sectional study we compared spirometry lung function in black African children with SCA living in Nigeria and in the UK.

Methods

Black African subjects with SCA (Haemoglobin phenotype: HbSS), aged 5-18 years, from Evelina London Children's Hospital, UK, and from the Barau Dikko Teaching Hospital, Kaduna, Nigeria, underwent spirometry and anthropometry. Z-scores were derived from the GLI2012-black equation[2] for spirometry and from the WHO 2007 growth standards[3] for anthropometry. Children were recruited on the occasion of a follow-up appointment in the outpatient sickle cell clinic. Healthy Nigerian control children were also included in urban and rural schools in Kaduna State, Nigeria. A portable spirometer (Easy on-PC, NDD, Switzerland) which meets the ATS/ERS requirements[4] was used. The Principal investigator performed 97% of the tests. Patients with SCA-related morbidity in the previous two weeks or poor quality test[4,5], were excluded. Spirometry patterns were classified as normal, obstructive (zFVC≥-1.64 + zFEV$_1$/FVC<-1.64), restrictive (zFVC<-1.64 + zFEV$_1$/FVC≥-1.64) or mixed (zFVC<-1.64 + zFEV$_1$/FVC<-1.64). Group differences were assessed by t tests. Linear regression models were developed to explore the association between spirometry outcomes and age.

References

[1]MacLean JE et al. Am J Respir Crit Care Med. 2008 Nov 15;178(10):1055-9.
[2]Quanjer PH et al. Eur Respir J. 2012 Dec;40(6):1324-43.
[3]http://www.who.int/growthref/en/ [accessed 2017 August 28th]
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Results

The main results are presented in the table and in Figure 1. The GLI-black equation fitted 341 Nigerian controls [mean(SD) z-score -0.34(1) for FEV$_1$ and -0.28(0.97) for FVC]. Among children with SCA, the prevalence of obstructive spirometry pattern (Figure 1) was 12% (9/75) in UK and 5.2% (8/154) in Nigeria (bottom right Fig. 1) . Restrictive spirometry pattern had a frequency of 10.7% (8/75) in UK patients and 29.9% (46/154) in Nigerian ones (top left Fig. 1). 9.3% (7/75) of subjects with SCA in UK and 3.2% (5/154) in Nigeria had a mixed restrictive/obstructive spirometry pattern (bottom left Fig. 1).

	SCA UK	SCA NIGERIA	Mean diff. UK-NIG (95% CI)
N (%male)	75 (54%)	154 (54%)	
Age	11.7 (2.8)	11.4 (3.2)	0.3 (-0.5; 1.1)
Height z-score	0.19 (1.25)	-1.77 (1.21)	1.96 (1.62; 2.3)
BMI z-score	0.11 (1.17)	-1.38 (1.34)	1.50 (1.13; 1.87)
FEV$_1$ z-score	-1.08 (1.03)	-1.38 (0.96)	0.29 (0.02; 0.56)
FVC z-score	-0.70 (1.01)	-1.21 (0.96)	0.51 (0.24; 0.78)
FEV1/FVC z-score	-0.89 (0.90)	-0.47 (0.98)	-0.41 (-0.64; -0.17)

Table 1. Anthropometry and spirometry z-scores in subjects with sickle cell anemia (SCA) from the UK and Nigeria. Results are presented as mean (SD), unless otherwise specified.

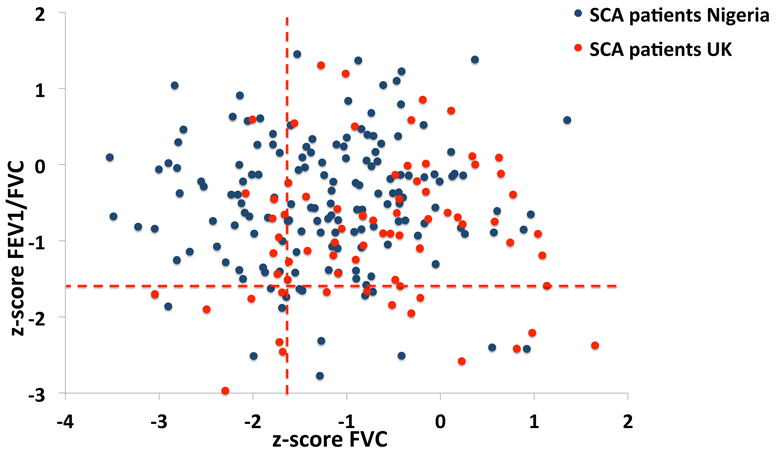

Figure 1. Scatter plot of FEV1/FVC z-score vs FVC z-score in children with SCA from Nigeria (blue dots) and UK (red dots). The dashed lines represent -1.64 z-score.

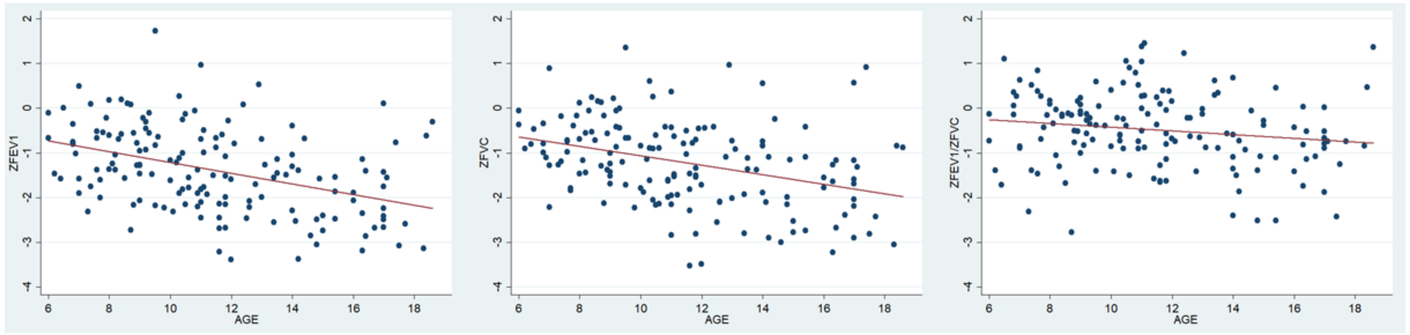

Figure 2. Trend of spirometry outcomes with age in Nigerian subjects with SCA (n. 154). FEV$_1$ and FVC z-scores tend to decrease with age [linear regression coefficient and R^2 were respectively -0.12 (95% CI −0.13; −0.10) and 0.73 for zFEV$_1$; -0.11 (95% CI -0.12; -0.9) and 0.66 for zFVC)].

Conclusions

Children with SCA from Nigeria have lower FEV1 and FVC z-scores and more frequent restrictive spirometry pattern than those from the UK. Nutritional status and inequalities in health care provisions might account for some of differences.

This research was partially funded by Sickle Cell Cohort Research Foundation (www.scorecharity.com) and by A.L.P.I. (Associazione Allergie e Pneumopatie Infantili)

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