Lung function in pediatric patients with sickle cell anemia from Central Africa

Online data supplement

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BACKGROUND

The Democratic Republic of the Congo (DRC) has a very high incidence of SCA but only a minority of patients has access to neonatal screening and secondary care for SCA, due to limited healthcare resources.[1]

METHODS

Caregivers of patients with sickle cell anemia (SCA) were interviewed about a history of asthma (parent report of physician-diagnosed asthma ever or current prescription of asthma medications) and frequency of pain crises in the last 12 months (bone pain in the chest, extremities or other areas, that required at least 24 hours of analgesic therapy) Controls aged 6-12 years were enrolled in four schools of different areas of Kinshasa. Of these, two were private schools attended by children of intermediate or high socio-economic status (SES) and two were public schools, mainly attended by pupils of low SES. Overall SCA patients and controls lived in the same urban environment and had similar variety of SES within them, although the age range was narrower in controls (6-12 years vs 6-18), as we could enrol pupils only in primary schools.

Controls with current asthma (at least one episode of wheezing or whistling in the chest in the last 12 months) or known chronic conditions that could affect lung function or spirometry performance (e.g., congenital heart disease, mental retardation) were excluded. This information was obtained from the children and their teachers, interviewed by a local investigator.

Lung function data from controls has been previously published.[2]

Assessments

Age was recorded with one decimal accuracy. Height was measured to the nearest mm using a stadiometer with a movable right angle headpiece (Leicester stadiometer, *Seca, Hamburg, Germany*). Weight (*Seca digital scales, Hamburg, Germany*) was measured with light clothes and without shoes, and Body Mass Index (BMI) was calculated.

The principal investigator performed all the spirometry tests, using a Pony FX spirometer (*Cosmed, Italy*), which meets American Thoracic Society (ATS) requirements.[3] Children performed two up to seven forced expiratory maneuvers standing upright with nose clip *in situ*. Quality control was performed according to ATS standards adapted for children.[3,4]

Statistical analysis

Normality of distribution of continuous variables was assessed by the Shapiro-Wilk test. Group comparisons were performed using unpaired *t* test, Wilcoxon rank-sum test, χ^2 test or Fisher's exact test as appropriate. A p value < 0.05 was considered statistically significant.

Regression analyses were restricted to SCA patients. A multivariable logistic regression model for "restrictive spirometry pattern" (versus "normal") was built, including the following

covariates: sex, age, zBMI < -2, history of acute chest syndrome, history of asthma, at least 3 pain crises in the last year, SpO2 < 96%, being followed in a primary care clinic (versus secondary care centre). Covariates selection was carried out through a backward approach, by retaining in the final model variables with a *p* value < 0.2.

Comparison of data from 73 patients with SCA and 73 controls would provide 80% power at the 5% significance level to detect a prevalence of restrictive spirometry pattern of 20% in patients with SCA (higher than 12-14% reported in recent studies from high-income countries[5,6]) and 5% in controls. There was over recruitment in both patients and controls due to the fact that for the SCA patients, we included all individuals who were seen at the SCA Clinic during 10 working days, as parents had been previously communicated that their sons would have undergone lung function assessments. We felt that it was ethically correct to recruit all children, whose parents gave the informed consent. For controls we needed to enrol at least 150 boys and 150 girls to validate the GLI-reference values for DRC[7]. We had to enrol all the eligible pupils whose parents had given their consent to lung function assessment.

RESULTS

Frequency of clinical variables in patients with sickle cell anemia (SCA) are provided in table E1.

Table E1. Frequency of clinical variables potentially related to lung function in 112 patients with sickle cell anemia (SCA) aged 6-18 years from DR Congo.

Variable	% of total (N)
Current hydroxyurea treatment	14.2 (16)

History of asthma	7.1 (8)
History of acute chest syndrome	12.5 (14)
At least 3 pain crises in the past year	26.7 (30)
SpO2 at baseline <96%	25.9 (29)

Among pediatric patients with SCA from the Democratic Republic of the Congo (DRC) included in the final analysis, asthma prevalence (parent report of physician-diagnosed asthma ever or current prescription of asthma medications) was 7.1% (8/112). Six out of 8 patients with a history of asthma had a current prescription for short-acting beta agonists as needed, whereas no one was taking prophylactic inhaled corticosteroids. Of these patients, 2 had an obstructive spirometry pattern, 2 had a restrictive spirometry pattern, 1 had a mixed pattern and 3 had spirometry values within the limits of normality. Of two SCA patients with asthma but not current prescription of anti-asthma medications, one had an obstructive pattern and the other had a restrictive pattern.

References

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