Hospitalization of children with sickle cell disease in a region with increasing immigration rates

Sickle cell disease (SCD) has become a paradigm of immigration hematology in Europe. Accurate up-to date information is needed to determine SCD prevalence, define real burden of disease and develop appropriate clinical networks of care, especially in regions lacking screening programs. We used two independent sources of data (Regional Register of Rare Disorders and Regional Register of Hospital Discharge Records) to determine extent of SCD and pattern of hospitalization of pediatric patients in the Veneto Region of North-East Italy. A steady increase of case notifications and hospitalizations has been observed in the past five years. Ninety-five percent of patients are immigrants with HbS/HbS SCD. Specialized regional registers can be used to define disease extent and guide targeted interventions in regions still lacking comprehensive care screening programs.

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Inherited hemoglobin disorders are an increasing global health problem.[1-2] Every year an average of 300,000 children are born with sickle syndromes[3] and prevalence of sickle cell newborns ranges from 0.1/1,000 in non-endemic countries to 20/1,000 in several parts of Africa.[3] In fact, 19-27% of the African population bears the sickle cell gene.[1]

Sickle cell disease (SCD) is Italy's most frequent hemoglobinopathy. It used to be a disease common among white Sicilians.[4] Sickle cell gene frequency is 2-13% in the Sicilian population.[5] Recent immigration from Africa, South America and the Balkans,[6] mainly in the Northern Regions, has changed the profile of SCD (7-8) with an increase of new SCD diagnosis in the North (20% in 1994 *vs.* 13% in 2003), coupled with a rise in HbS/HbS genotype (28% *vs.* 21%). In the Veneto Region there are 320,793 immigrants making up 6.8% of the entire population. Twenty-eight percent of immigrants are African of which 25% are children.[6] The birth rate among immigrants is the highest in the country (25.8/1,000) and the number of births from to immigrant couples reaches 30%.[6]

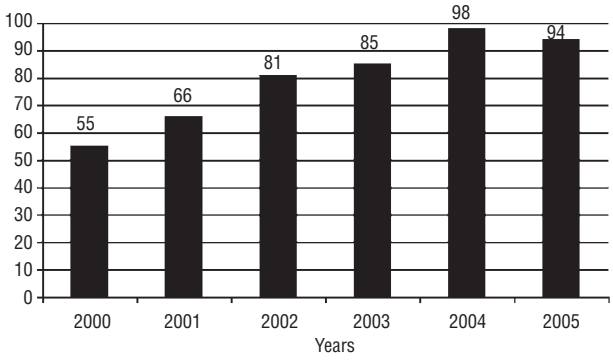

Figure 1. Inpatient admissions due to sickle cell disease in the Veneto Region per year from 2000 to 2005.

Evidence of an increasing number of children with SCD has come from clinical practice at hospitals and pediatric offices throughout the Region,[9] but there are no accurate figures for disease prevalence and characteristics.

We evaluated the extent of SCD and the profile of its clinical manifestations in children aged < 15 years in the Veneto Region, using two independent sources of information: the Regional Register of Rare Disorders (RRRD) and the Regional Register of Hospital Discharge Records (HDR).

RRRD was created in 2002 and is based on a computerized network connecting the Regional Referral Centers for Rare Disorders. The Hospital-Discharge Register records discharges from regional public and private hospitals reimbursed by the National Health Service. HDRs collected from 2000 to 2005 with codes for HbS/HbS, HbS/HbC, HbS/HbD, HbS/HbE (282.60-282.61/2/3/9) according to the International Classification of Diseases, 9th Revision, Clinical Modification (ICD-9-CM) were included. HbS/β thalassemia patients, coded with thalassemias, were identified through the RRRD and their HDRs added. To safeguard privacy, each patient was given an identification number to trace their hospital data. HDR were analyzed for patients' social conditions, number of hospitalizations, average length of stay (LOS) and ICD-9-CM codes of SCD-related complications.

One hundred and thirty-five SCD pediatric patients were identified, 51 had been registered at the RRRD, 20 are male (39.2%) and 31 female (60.8%). Mean age at

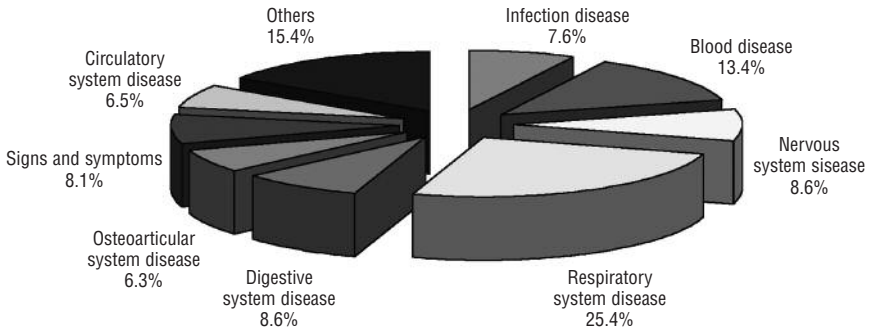

Figure 2. Diagnosis of discharge according to diagnostic groups in patients discharged with SCD from 2000 to 2005

entry into the register was 3.8 years (range 0-15), 98% were immigrants (3 Brazilian, 2 Albanian, 46 African), 31.3% were born abroad, 66.6% were born in Italy from immigrant families, one child was Italian. Thirty-three had been admitted at least once and their HDRs were included in the hospitalization analyses, 18/51 (mainly infants, two with prenatal diagnosis) had never been admitted.

One hundred and seventeen had been discharged with diagnosis of SCD, 60 males (51.3%) and 57 females (48.7%). Mean age at first admission was 4.4 years (range 0-15), 84% had foreign parents, 91.6% from Africa (Nigeria, Ghana, Congo). Eighty-nine percent had a permanent address in the Veneto Region, 4.2% in other Italian Regions, 6.8% in foreign countries but were living in Italy.

Admissions totalled 921 (52% inpatients, 48% DH). A progressive increase in the number of inpatient admissions was observed (Figure 1). A subgroup of 9 patients (7.7%) accounts for 133 admissions (14.4%), confirming extreme phenotypic diversity of SCD. Mean inpatient admissions/patient were 4.2 (range 1-27). Mean admissions/patient/year were 1.8 (range 1.6-2).

One hundred and twenty-three admissions (13.3%) were due to sickle cell crisis, 44 to pneumonia, 15 to aseptic necrosis of bone and 5 to severe infections (sepsis/meningitis). Mean LOS was 6.42 days (range 1-118), while LOS for sickle cell crisis was 7.4 days (range 1-118), higher figures than previously reported (4.4-4.9 days in the United States), equalling adult mean LOS.[10]

Diagnosis of discharge, excluding SCD crisis, is shown in Figure 2. Interestingly, 8.1% are in the general Signs and Symptoms group (Figure 2). Sixty-four out of 117 patients had at least one DH admission, 1,744 DH days were observed, with a mean of 27 DH days/patient and 10 DH days/patient/year. No death was observed.

Data from Regional Registers help define local SCD patterns in countries still lacking comprehensive care screening programs, but experiencing increasing regional trends. In our region, age at diagnosis is high, correlating with the lack of newborn screening. This is warning to physicians of the risk of severe acute complications due to late implementation of prophylactic measures. A high rate of admission, DH use and long LOS suggest personnel and parent training are needed.

Raffaella Colombatti, Laura Visonà Dalla Pozza,° Monica Mazzucato, Laura Sainati,* Marta Pierobon,* and Paola Facchin°*

**Clinic of Pediatric Hematology and Oncology, Department of Pediatrics, University of Padova; °Epidemiology and Community Medicine Unit, Department of Pediatrics, University of Padova, Italy*

Correspondence: Raffaella Colombatti, Clinic of Pediatric Hematology Oncology, Department of Pediatrics, University of Padova, Italy. E-mail: rcolombatti@gmail.com

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