Research Article

Descriptive approach for sickle cell disease in Eastern of Algeria

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ABSTRACT

In Algeria as in all countries of the world, sickle cell disease has become increasingly common in children as in adults, which leads to a worsening of anemia and the development of secondary diseases connected with sickle cell disease known as major sickle cell syndrome (MSCS), which includes the cerebro vascular accidents CVA, heart disease and especially infectious diseases, So many secondary complications of sickle cell disease. It is in this context that this study is therefore an epidemiological and clinical approach to this disease. This Works occurred during the years 2010-2011-2012 and aims to identify children with this form of anemia that are at the beginning of the disease and not know that a late diagnosis. To know their way of life, the repercussions of this disease on their education, how come they have overcome the pain and especially the frequency of appearance of secondary diseases such as anemia. Sickle cell disease results in serious repercussions on children and high costs of care. It is therefore necessary to monitor children at an early age. A daily hydration and several times during the day is very important coverage during the cold and not heat exposure is a major priority in the prevention against sickle cell disease because it prevents dehydration asthma triggers by increasing blood viscosity .

Keywords: CVA, MSCS, School failure, Sickle cell anemia.

INTRODUCTION

Sickle Cell Disease is a common genetic disorder, autosomal recessive, which combines 03 major categories of events, chronic hemolytic anemia, vaso-occlusive Phenomena and fragility to infections. It is a disease which is by its frequency and seriousness a major public health problem worldwide.[1] Its distribution is different from one region to another in Africa to India, West Indies, America and the Mediterranean rim (classified as medium risk). Recent WHO estimates indicate that this disease affects 100 million people worldwide.[2] The WHO mortality is estimated at 80% before the age of 05 years caused mainly by the poor management of this costly disease[2,3] and often require: Repeated blood transfusions, Specialized Laboratory (phenotyping and serology), Transcranial Doppler (CT and MRI) in case of neurological complications.

in Algeria the homozygous sickle cell disease is 5%, the heterozygous 1.2% and 1.6% B Thalassemia[3] The risk of infection is important in our study population, with particular vulnerability to germs and complications, it is especially septicemia, meningitis, pulmonary infections and osteomyelitis.[3] The sickle cell anemia is a hemoglobinopathy with a very complex clinical polymorphism, late diagnosis and inadequate care in our study area make it a plague that raises the interest of local officials knowing that through the all its hemoglobinopathy complications can reach all areas of the body.

The aim of this work is to describe the disease, to define the frequency of sickle cell disease and its complications in the schools of the city of Sidi Amar.

MATERIALS AND METHODS

Location

The study took place in the city of Annaba city in the north-eastern Algeria. It is considered the fourth largest city in the country in terms of population. Formerly called Bonne for the French colonization and Hippone in antiquity, it is the fourth largest city in population after Algiers capital of Algeria, Oran and Constantine . The head of the wilaya of Annaba is 152 km north of Constantine, 246 km east of Jijel and 80 km west of the Tunisian border. Annaba is a coastal city with a population of 600,000.

Type of investigation and study population

This is a descriptive cross exhaustive study referred that took place during the years 2010, 2011, 2012, and during the first half of 2013. The study population consists of both sexes school students aged 6 to 12 years old and lives in Sidi Amar (Annaba), 53% are boys and 47% girls. The sample size is estimated at 8,401 students all examined by doctors of School Health, 341 are anemic and 30 with sickle cell anemia and possibly a major sickle cell syndrome (MSCS). The examination is a fundamental step in the diagnosis because it provides half of guidance when conducted carefully. In fact, it allows you to specify the personal and family characteristics of the student and to know the genetic predisposition.

Inclusion criteria

- Electrophoresis greater than 50%

- Specific sickle cell clinic

Exclusion criteria

- A 50% lower electrophoresis

- Patients who received a transfusion less than 2 months before the exam.

Data Collection

Data were collected through individual interviews with students and their parents based on a questionnaire approved and used by services of hematology and another questionnaire adapted to the conditions of Algerian society, the first part of the questionnaire was designed to capture mainly demographic, socio-economic and socio-cultural information, the second part was to gather information on the genetic predisposition of sick students, the third party supports clinical and laboratory examinations and laboratory tests that the child has suffered for a diagnosis of sickle cell disease and certain of these secondary complications .

The biological data

Laboratory tests based on several tested with blood frotti which was the first examination among students who had anemia and it may determine for the presence of sickle cell red blood cells so as fossil, The second test is the complete blood count or CBC which allows us to count the number of elements in each of the three categories of blood cells are red blood cells, white blood cells and platelets , The third test was the test of sickling or EMMEL test which aims to seeking the presence of sickle cell in the case where the blood frotti is negative, diagnostic certainty is that the hemoglobin electrophoresis is performed.

Data processing and statistical analysis

Data analysis was performed using the following software: SPSS v6, Excel 2007, stat plus 2007.

RESULTS AND DISCUSSION

Description of the sample

In this study the age range of students was between 6-12 years in 14 primary schools and six secondary schools, the male is slightly predominant: 53% compared to female sex ratio is estimated at 1 1, the majority of students come from households where parents are a professional social level low or medium, 40.8% of students reside in a narrow apartment 48% suffer from a lack of transportation, 83.3% did not have hot water and 91.8% are without heating. The level of parental education was very different from one family to another. The rate of illiterate or with primary education parent represents 24.53% of the sample. Respondents in higher education over Baccalaureate represent 37.11% (Table 1). 45.91% of parents were asked which 73.68% are fathers. Dads

unemployed represent 48.42%, mothers 59.09%, 40.78% are workers, 13.15% are MSP (managers and skilled professionals) and 10.52% are retired (Table 2).

Table1: Sample distribution by level of education

Parameters	Treatment groups			
	Higher	Secondary	Middle	Primary I + Illitrate
Number of respondents	118	76	46	78
%	37.11%	23.90%	14.47%	24.53%
Women %	33.88%	24.79%	14.05%	27.27%
Men %	47.37%	21.05%	15.79%	15.79%

Table 2: Distribution of population by socio-professional categories

Parameters	Treatment groups		
	Men	Women	All
Security workers	2.63%	0 %	0.62%
Craftsman	5.26%	0.82%	2.2%
Retailers	10.52%	0.82%	3.14%
MSPs	13.15%	9.91%	10.69%
Workers	4O.78	25.61%	
Teachers	1.31%	0.31%	29.23%
Retirees	10.52%	3.71%	5.34%
Unemployed	15.78%	59.09 %	48.42%

Of the 16 sites, we noted a peak in 03 establishments primary schools (Bahi Amar 1-10%, Aicha 1-12%, Aicha 2-11%), and an average secondary school Aisset IDIR school idir 11% (Figure 1).
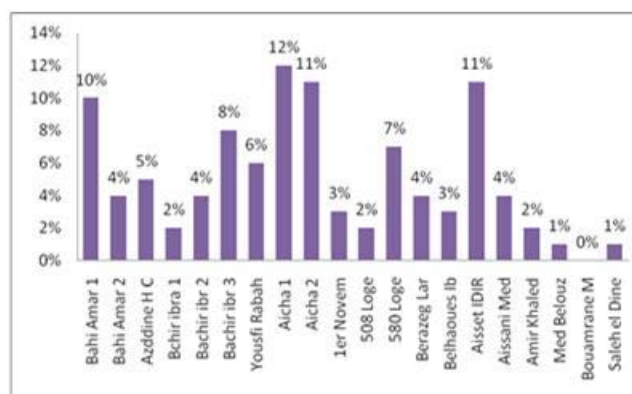
Figure 1: Graphical representation of the percentage of school children with sickle cell disease.

In all children with sickle cell disease, we noted behind in school which is very significant it is 63%. These students redo the year and the end of a very difficult (Figure 2A).

The weight and size of students were below the standards recommended by WHO, the average size was 134 cm for females and 140 cm for males (Figure 2B), the average

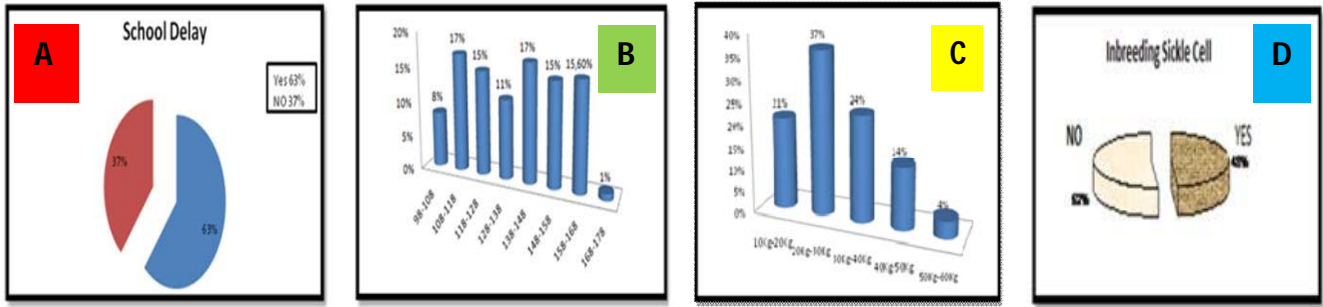weight was 29.92 kg girls against boys 31.46 kg (Figure 2C).

Figure 2: Graphical representation of school delay (A), depending on size (B), by weight (C) and inbreeding among school children with sickle cell disease (D).
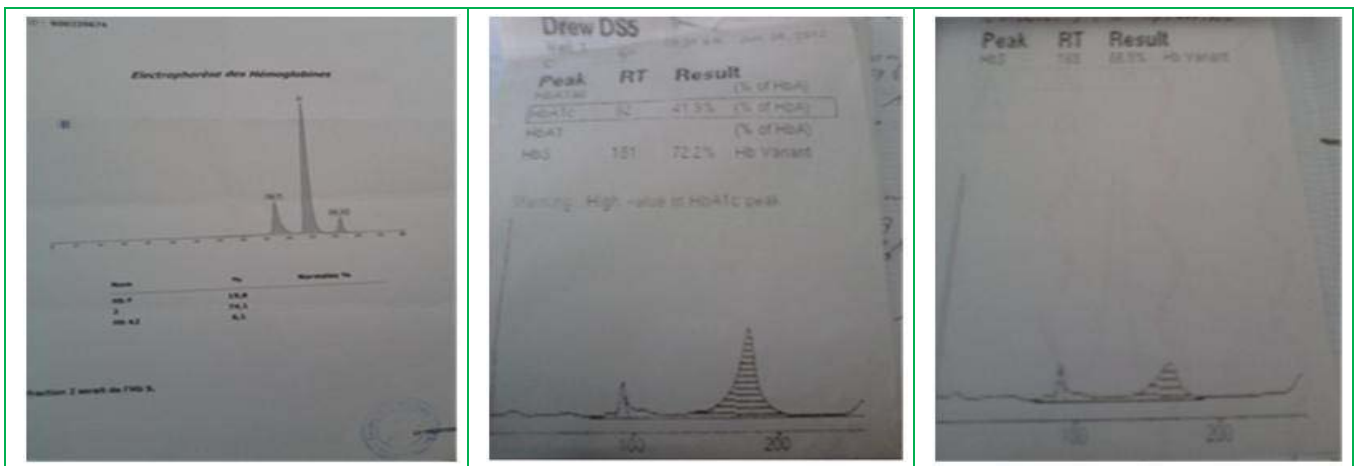

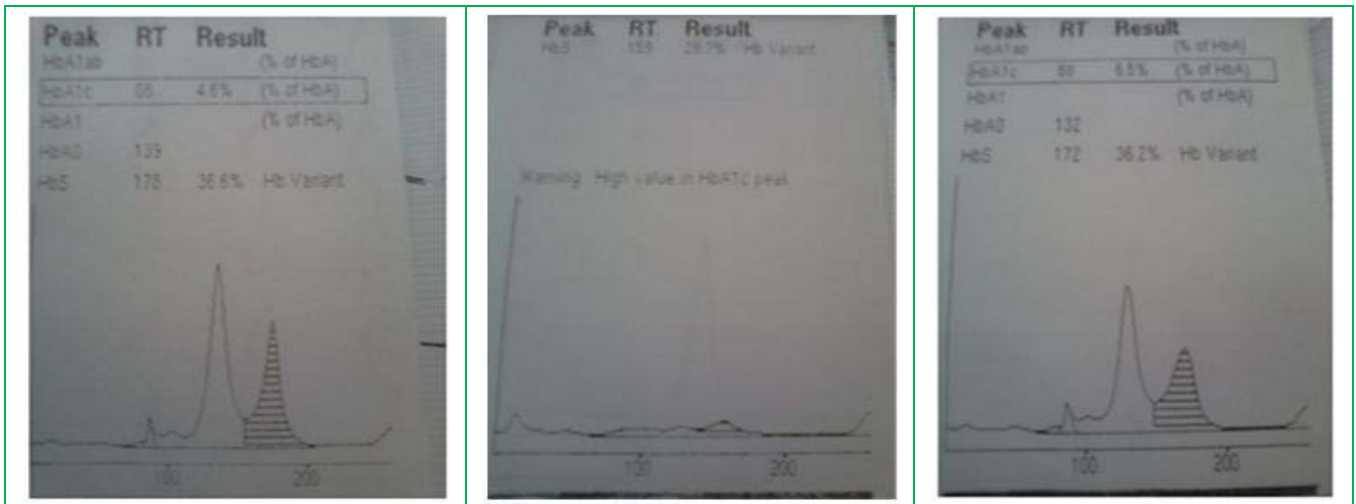**Figure 3:** Example electrophoresis of the first batch with Hbs more than 50%.

Figure 4: Example electrophoresis of the second batch with Hbs lower than 50%

Genetic predisposition

For the genetic part we found that inbreeding is present in 48% of cases especially among families of children who live in rural areas, on the other hand, our survey shows that 76% of children from families with an average of 3 children and at least two brothers are sick (figure 2D).

Results of laboratory tests

Based on the results, we shared our sample into two lots. In the first batch the results are as follows: there is a NFS disturbed namely a decrease in GR = 2.69 × 109 elements, a HGB rate = 6.8 g / dl, a MCHC = 32.3 g / dl and an MCV = 91 fl, for a normocytic normochromic anemia. The blood smear detects the presence sickle erythrocytes (or sickle) with anisopoikilocytose, Hb electrophoresis objective a HbS rate = 87.6%, The sickling test is strongly positive which confirms the presence of sickle cell As part of a family survey, electrophoresis of the parents shows that they are both heterozygous sickle with: Hb = 58%, HbS = 38.5%, HbA_2 = 3% and traces of HbF. These data were used to make the diagnosis of sickle cell type homozygous

sickle cell syndrome. In the second group the results are different, there is a NFS which has a rate of GR = 3 × 109 elements, HGB = 9.6 g / dl, MCHC = 26 g / dl and MCV = 66 fl. for a microcytic hypochromic anemia, a blood smear that shows both sickle and anisopoikilocytose. A positive sickling test and hemoglobin electrophoresis showing: a HbS rate = 88.8%, HbA $_2$ = 6.4% and HbF = 4.8%.

The objective family survey for this lot shows that both parents are: one heterozygous for thalassemia Hb electrophoresis high HbA$_2$ = 5%, HbF = 1% and Hb = 94% and the other is heterozygous sickle cell to electrophoresis with: rate HbA = 57.5%, HbS = 41%, HbA$_2$ = 1.5% and traces of HbF. Personal ATCD β0thalasso types of sickle cell-sickle cell syndrome, these children have regular CVO requiring many hospitalizations, These children have a regular biological followed by: Periodic-SNF, Determination of iron and ferritin, Determination of TGO,TGP and TP, Determination of micro albuminuria and blood creatinine.

Diagnostic certainty "hemoglobin electrophoresis"

The hemoglobin electrophoresis has resulted in a separation of the different hemoglobins by their electrical charge and molecular weight, and highlight the presence of a fraction of hemoglobin different migration normal hemoglobin, so the diagnosis and to differentiate between homozygous forms more than 50% (figure 3), heterozygous Hbs lower forms 50% (figure 4) and we got two lots on the following routes.

Aspect of pain in sickle cell

The painful crisis is a major clinical symptoms of sickle cell disease. It is a symptom complex where the body and the psyche are intertwined. He child puts forward a somatic damage that comes with sometimes confusing even psychopathological psychic manifestations[4].

In this work we try to explain the painful phenomenon, including its specificity by psychosomatic entanglement and social dimension with the family environment as well as social, we speak here mainly of severe sickle cell pain and how it is perceived by these children. Thus we found that the pain of sickle cell crisis is localized most often in the bones. It is related to a vaso-occlusive phenomenon of microcirculation due to sickle red blood cells, decreased tissue oxygenation results in micro-infarction in the bones but also the muscles, intestines and potentially throughout the body[5,6,7]. The pain is sudden, unpredictable, it is not seen, so no external sign, By cons we noticed that the breathing and heart rate increase as well as muscle tension and sometimes the child is sweating. Some children are restless, prostrate or completely frozen paralyzed attitudes or take painkillers And to assess the chronic pain we opted for the use of concise Questionnaire pain (QCD), which is the French translation of the Brief Pain Inventory (BPI) approved by WHO.[8] This questionnaire has been extracted from the book The Brewer[9] holds the following basic tools validated and adapted to the Algerian society.

- A scale for measuring the intensity of the pain that is the visual analogue scale (VAS).
- A scale for measuring the pain intensity and which is digital scale (DS).
- A diagram showing the topography of the painful areas.

These self-assessment scales and instruments are questionnaires completed by the child without peer pressure. These simple scales have been validated for measuring the intensity of the pain, they do not appreciate other dimensions of pain, and do not allow for accurate diagnosis of the underlying mechanisms, but they are useful to better detect patients needing symptomatic treatment.

Visual Analog Scale (EVA)

Was used in this work the mechanical form of the scale consists of a strip on which is drawn a horizontal or vertical line of 100 mm on the "patient side" oriented from left to right (1). Both ends of the line are respectively defined by terms such as "no pain" and "worst pain imaginable." The child responds by moving the cursor to the ruler. The pain intensity is measured by the distance between the cursor and the end "no pain" to "face contact" (figure 5).
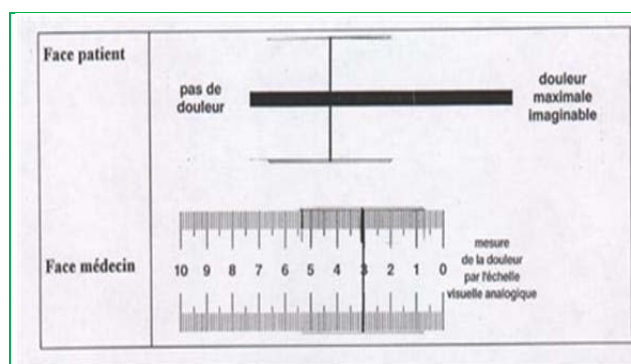

Figure 5: Visual analog scale (EVA)

Digital Scale (DS)

In this scale of measurement we asked the students sick to tell us below a score of 0 to 10 which best describes the importance of the pain they feel. A score of 0 is "no pain." Note 10 is the "worst pain" (figure 6).
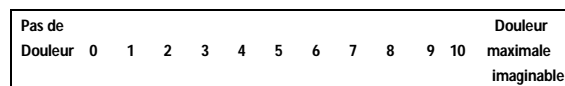

Figure 6: Digital scale (DS)

Schematic topographic

For children first second and third school year saw their infancy we asked them to indicate on a diagram where they feel pain by filling the area with a pencil and put in the drawing an "S" or believe X for pain near the surface of their body or a "P" or a double cross XX deeper pain in the body, and an "I" or a triple cross XXX where they feel the pain more intense. The results are highly significant for the intensity of the pain that is very different from one

child to another Thus the results of the first and second test are very close to each other with 81% of sick children "39% of boys and 42% girls" were all chosen by more than 5 degrees on both scales EVA IN which reflects very clearly the intensity of the pain that these children experience during the crisis (figure 7).
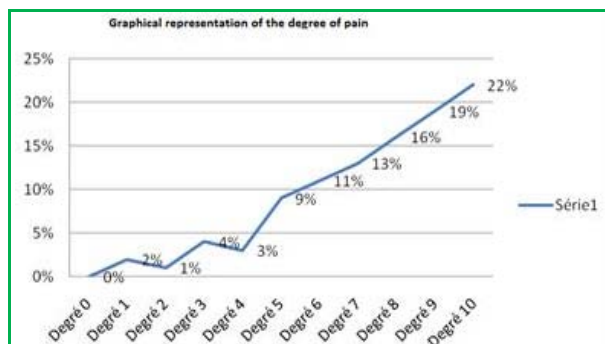

Figure 7: Graphical representation of the degree of pain

For the third test given to affected children of 1st, 2nd and 3rd Year All figures are obtained double cross on all the joints of the hind limbs while those of the hip and shoulder area "65%" and that can only be explained by the different osteo-articular and bone infarcts that the child suffered during the crisis, and triple crosses more about "25%" of the figures obtained from the children of three years and are already quoted on the forehead and all head the survey shows that the child had either a TIA or transient ischemic accident "19%" is stroke with severe convulsions and loss of consciousness in "6%" of cases (Table 3) .

Table 3: Percentage of answering children due to of the third test of pain

Treatment groups	Pain sensation		
	A Cross	Two Cross	Three Crosses
Boys	4%	39%	13%
Girls	6%	26%	12%

Threads

Magnitude of the problèmes

This study estimated the prevalence of the disease in schools is on the order of 0.28% compared to the rates of diseases encountered, in this community the past 10 years, if it is low the clinical polymorphism, the expensive cost burden on families and the state, serious complications caused, the lack of blood make it a scourge.

The age is 6 to 12 years, we have seen a slight male predominance seen autosomal sickle cell anemia. Failure to thrive weight is common and academic delay from the failure to retire.

The family survey also revealed that families have an average of three children, two are sick and the socio-economic level is modest and intellectual level is low which makes it difficult patient physician relationship.

The motif discovery is most often found the anemia explained by the natural course of the disease is asymptomatic until the age of 4 months and a dominant first 6months anemia, 5 years and then installation infections and vaso-occlusive crises 5 years - 12 years, giving way to organic complications and SDM. Splenomegaly regressed to the age of 6-7 years so rare in this sample. Our study showed that patients on average are polytransfused five transfusions per year with no phenotype whole blood (alloimmunization). The vaso-occlusive crises are common in this age where the pain is the most feared by the sickle cell test require adequate care and appropriate psychotherapy. Lack of genetic counseling (complexity of the disease, the intellectual level). Inability of parents to understand the disease and to support their children properly.

In view of these results, sickle cell anemia should be considered a public health priority, it is imperative to establish a local program management and monitoring of these patients in the wilaya of Annaba.

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