# Review Of Consultations For Children With Sickle Cell Disease (CHR Kenitra Morocco)

Mouden Samira, Ahami Ahmad Omar, Mouilly Mustapha, Fayez Noureddine, Hajji Ahmad

Abstract: Also called sickle cell anemia Sickle cell anemia is a genetic disease of autosomal recessive linked to abnormal structure of the hemoglobin which leads to the formation of hemoglobin S (HbS). The descriptive and cross that we conducted at the pediatric ward of the regional hospital Gharb study Chrarda benihessen Kénitra we were enjoying a work force of 164 children with sickle cell disease over a period of twenty four months (from June 2010 to June 2012). On average seven to eight (7-8) new cases per year. The majority of these children admitted to the exhibit acute complications that are typically associated either to a chronic hemolytic anemia, vaso -occlusive crisis, acute chest syndrome, or severe infections, which aims to identify factors likely to play a role in the occurrence of the sickle cell crises. This can cause severe functional consequences (with renal, lung, bone, etc.). In order to understand the problems and difficulties faced by sickle cell and their families in their daily lives, we have established a protocol form of a questionnaire exploring various aspects related to eating habits and lifestyle of these children. and their families, as well as family socioeconomic status, and the context of environmental life The size of the control group study consists of 60 children aged 7-14 years. Clinical and analytical information is collected from records, medical records and doctors during consultations of these patients. The results show that 71% of these children are from rural areas against 20% of children of urban origin, while 9% live in suburban area however. The majority of parents have irregular income, low educational attainment. 68% of these children use septic false in parallel they use well water in consumption and domestic use. Clinical examination and blood cell abnormalities formula revealed prevalence of 76% for fever cases painful crises severe recurrent and unpredictable also observed, as well as "acute chest syndrome " ( pneumonia or pulmonary infarction), a bone or joint necrosis unpredictable 43% for leukocytosis . As we noted the presence of diarrhea in this sample. In conclusion, the present study showed an association between the frequency of sickle cell crises especially infectious presented by the children studied and the environmental quality of family life in which the child develops.

Index Terms: Keywords: child, sickle cell crises, Environment, socio-economic determinants region gharb Chrarda benihssen, pediatrics.

## 1 Introduction

Sickle cell disease is a genetic disorder of hemoglobin (Labie et al., 2003), related to the presence in high concentrations in the red blood cell abnormal hemoglobin called hemoglobin has SHbS [1] All which May Be circulating behind the chronic organ failure Requiring Multidisciplinary care. It is a serious disease Characterized by high morbidity and mortality [2][3] and which is by its frequency and seriousness a major public health problem on a global scale [4]. It a disease with clinical severity is highly variable[5] and whose symptoms are multiple. The three main events are the anémie hémolytique, vaso-occlusive pain crises that can affect different organs and less resistance to certain infections [6] that can develop in a rapid and must. It is therefore important to prevent outbreaks to reduce their risk of developing[7]. We propose to determine the origin of the high frequency of sickle cell crises in children with sickle cell disease followed in the pediatric ward of the regional hospital center gharbe chrarda Beni hssen Kenitra, which has significant sickle cell recruitment with a total of 164 children followed with an average of 7-8 new cases per year.

- Dept. of Health and Human Biology population. Unit of Clinical Neurosciences and notional health. Department of Biology. University Ibn Tofail .kenitra. <u>ahami\_40@yahoo.fr</u>
- Regional Hospital Centre Kenitra, Pediatri Service.

The majority of the children admitted to the pediatric ward have acute complications, vaso-occlusive and infectious complications associated with chronic hemolytic anemia. The aim of our work is to list the etiology and determinants of these complications and crises factors. In addition, we found that the high incidence of these sickle cell crises is related to socioeconomic and environmental characteristics of these children.

## 2. Part of the study

The Gharb plain or watershed Sebou-out there is a region in the northwest of Morocco, characterized by a very important agricultural and industrial potential. It is crossed by the Sebu, one of the most important wadis of the country. However, the diversity of polluting human activities in the region (agriculture, industry, traffic ...), generally located along its banks, and the lack of wastewater treatment plants affect the environmental quality of the region [8]. Other units, low and irregular household income generated by these activities influence the level and quality of life [9]. All these environmental factors, socio-economic and nutritional could affect the health of the population, especially children's health.

## 3. Subject and method

A cohort of 64 patients with sickle cell disease was examined in a descriptive, prospective study conducted from June 2010 to July 2012, we decided to study anthropology such, this approach offers the possibility of further exploration and detailed analysis of the problem studied. Thus interviews were planned using a predetermined protocol through a questionnaire. It written in French language was administered in dialectal Arabic. We chose to develop our own questionnaire includes closed questions to identify socio-economic and environmental conditions of existence of families under our assumptions may play a role in access to care. A total of 64 children with sickle cell disease were identified by consulting the records of consultation of pediatrics. Attempts to contact the parents concerned were considered before the start of the investigation and that, in order to:

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- To obtain their consent to participate in the survey.

- Identify the points of attachment of these children (specific address, workplaces father and mother, etc)

The choice of questions answered with the objective of collecting as much information in our study population and within families, they revolve around two main components

- The first part of the questionnaire was used to assess the socio-economic and demographic characteristics of households (household income, education level of parents, sex, age and educational attainment of children, household size) and environmental pollution (use of septic tanks, water type used in consumption and domestic use).

#### - Food household practices

- Clinical symptoms at the time of crises and frequencies per year (presence of fever, pain (abdominal, joint pain), vomiting, frequency of seizures per year).

#### **Physical examination**

Each patient was clinically examined by a pediatrician looking suggestive of sickle cell crisis signs: asthenia, abnormal paleness of the skin and mucosa, dyspnea, palpitation, dizziness. Fever, abdominal pain and vomiting joint, in some cases the presence of diarrhea

#### **Biological study.**

-Blood Collection

Blood was drawn by venipuncture after an overnight fast in a tube containing EDTA K2 (anticoagulant). -Blood count.

### 4. Results

Blood counts were determined on a counter type PLC (Coulter): The information collected is the number of white blood cells, platelets, red blood cells, hematocrit (Haute), hemoglobin (Hb), mean corpuscular volume (MCV), mean corpuscular hemoglobin concentration (MCHC) and mean corpuscular hemoglobin (MCH). While, these examinations were performed in the biology laboratory of El Idrissi hospital Kenitra. This descriptive study examined a total of 64 children as planned but four died. Their age is between 7 and 14 years, the average age of the latter is 11, among them 65% are male (39/60), all of whose children are in school (76.6%) are primary level (46/60) (23.4%) are college level, for parents, the vast majority of them are of primary education (fathers) while almost all mothers are illiterate regarding the family income 71.6% of families have irregular and low incomes (43/60) the major part of the children involved in the survey (71 %) Were from Rural Areas (43/60) All which all use the water wells in consumption and domestic use in parallel They Use septic false, 20 % of urban origin (12/60) and (9 %) in peri-urban area. The symptoms that motivated the consultation can be grouped into banal signs (pallor, fatigue, fever) and presenting symptoms (jaundice, joint pain, abdominal pain, presence of diarrhea, vomiting, repeated hospitalization (4-7 times per year) for Laboratory tests showed that the majority of children suffer from severe anemia (3-7 g/dl) . Thereafter we studied the distribution of the study population According To three variables namely the habitat area, the rate of temperature and rate of blood cell bank (the curriculum of physiological

variations of white blood cells is subdivided into three categories: normal, leukopenia and leukocytosis).

Table I. Distribution of the population according to three
variables

	Rurban (U) N=12			Rural (R) N=43			Suburban (PU) N=5		
	Ν	Leu	Нур	Ν	Leu	Нур	N	Leu	Нур
Fever (F)	4	0	3	2 2	0	19	0	0	4
Normal ( NF)	4	0	1	2	0	0	1	0	0

Leu: leukopenia Hyp: leukocytosis

The illustration of the distribution of the study population according to the state of temperature, white blood cell count and the habitat area is shown in Figure I.

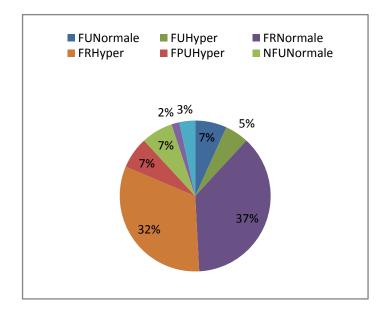


Figure I. The distribution of the study population according to the state of temperature, white blood cell and the habitat area

FUNormale: fever urban area rate normal white blood cell. FUHyper fever: leukocytosis Urban Area. FRNormale fever: rural, normal. FRHyper fever: rural, leukocytosis. FPUNormale fever: periurban, normal. NFUNormale: nonfièvre, urban, normal. NFUHyper: no fever, urban, leukocytosis. NFRNormale: no fever, rural, normal.

Sickle cell disease is a chronic disease that accompanies so sickle cell children in all stages of development. His Individual and family psychosocial impact Must Be taken into account in the care devices [10]. The people here share common Characteristics making it fairly homogeneous: pathology and its consequences. Were the Children, for The Most of Them share from Rural Areas with lower primary education. It is known that thesis children with sickle cell disease are penalized by school Usually Increased Absenteeism with a



Corresponding underachievement. This is the case of some children in our series Who Had to leave school early and / or Who-have Accumulated years behind In Their education [11]. The socio-economic analysis in this study Showed that income is irregular and low in the majority of these families. Also illiterate fathers and mothers. These Social and Economic Characteristics of the home environment light reflection frame life less favorable to the growth in the devices of care, while recommendations on the management of sickle cell crisis published in the British Journal. Of Hematology advocate a therapeutic education for the parents of children Affected and is designed to help prevent prevention Recognize gold some early complication of the disease [10]. However, some children are poorly supported and usually arrives at hospital disaster because of the remoteness carry home to the hospital, the parents' ignorance or Their Inability to bear costs victims talk care. Studies in compliance, the Precarious Economic conditions of families is described as a difficulty factor, both, in terms of Regularity of that treatment consultations, Given the Weakness of health coverage and Moyne de transport. Obviously the regular Followed thesis children causes' huge fairs That Heavily weigh on family budgets and limited resources (the case of needy families in our area) is Likely to Seriously Impede the management of sickle cell disease [12]. In addition, children require a thesis special diet [13] and nutritional supplementation with Proper hydration micronutrient supplementation are the Proposing as **Clinically Indicated:** 

- Folic acid daily has supplémentassions (5 mg/ day) is recommended Because Of Increased hemolysis related Needs;
- A daily zinc supplementation (10 mg zinc/cell) May Be Proposed Pendant prépubertair
- An significant hydration is Necessary

The fragment infection is a feature of sickle cell [14], the susceptibility to infectious agents is the result of a shift proved the immunological dynamics of sickle cell [15], Thesis in children Studied infectious syndrome is Accentuated Because Of Their opponent Conditions favoring Their exposure to sources of infection, what is Confirmed by the high prevalence of leukocytosis Associated with febrile state (32%) children presented by thesis sickle Especially When The original rural Comparing our Those results with --other of crisis work That fever is found That the first trigger of vaso-occlusive crisis in addition. thesis children use well water for food and household use do not meet the standards and THUS That May Be Contaminated with pathogens. These microorganisms Can Be's because the sickle cell as septicemia, urinary tract infections and pulmonary ostéomyélite (High Authority for Health 2005) of severe infections in patients. In general, the susceptibility to infections is Known sickle. It is functional asplenia Attributed To, All which is the result All of splenic infarction repeated [16] THUS, there are some fault and antibody synthesis substances Involved in antimicrobial defense. Sickle cell disease is Characterized by the Lack of a Specific treatment; for reduce the risk for complications of Involve daily vigilance and adequate reactions In Some Circumstances Health education for families is a requirement of management and crisis prevention of success[17], Theoretically easy, since we know the Contributing factoring and triggers: efforts, dehydration, cold, fever, infections, and the stress. Preventive Measures are Not Easily accepted by

children To Whom It is recommended to stay calm and drink enough. It is significant for patients to Understand the issues of tedious daily treatments, discerned warning signals to the driver in emergency units (extreme pallor, splenomegaly, high fever ...), also reassured about goal BE Their Ability to disease and That Their marriages, therefore, eases anxiety to Suffer Their again, Itself Generates painful phenomena.

## Conclusion

Our study had the advantage of free medical and socioenvironmental problems remain exposed sickle cell children in our region, despite the significant improvement in terms of care and quality of life of these patients. So serious reflection should be undertaken as part of a strategy to build and unify protocols supported crisis drépanocytaires and to involve parents in the care home in their awareness sense of environmental health and preventive actions to minimize complications.

## References

- [1] HAS, Syndromes major dre'panocytaires child ETDE adolescents. January 2010.
- [2] Embury SH, Steinberg MH.Clinical considerations.In:Embury SH, Hebbel RP, Mohandas N, Steinberg MH, eds.Sickle Cell Disease. Basic Principles and Clinical Practice. RavenPress 1994: 505-73.
- [3] Assimadi K. Sickle. School sports and crafts. Family and Development, 1990; 57: 15-6.
- [4] Zohoun I. Drépanocytose et santé publique. Famille et Développement, 1991
- [5] Pollack CV. cell disease. HematolOncolEmerg 1993; 11 :365-78.
- [6] Labie D, Elion J. Genetics and pathophysiology of sickle cell disease in LaDrépanocytose editions John LibbeyEurotext, 2003 (dir Girot R., Begue P., F. Galacteros.): 1 12.
- [7] The nouhoum TraoreLa sickle cell child Internal holder hospitals at Gabriel Toure Hospital: Bamako (Mali) on Jun 19, 2013.
- [8] Azzawi FZ Ahami AOT Khadmaoui A., 2008, the relationship between socio-economic, environmental and malnutrition: Case of children aged 6 to 8 years in the Gharb plain (Northwest Moroccan), Antropo 17
- [9] Bennasser L., 1997 Diagnosis of the state of the environment in the Gharb plain: followed by the macropollution and its impact on the hydrochemical and biological quality of low Sebu. PhD thesis, Univ. Ibn Tufail, Kenitra, 177p
- [10] Laine A. Sickle. Perspectives on an orphan disease. Paris: Karthala; 2004.
- [11] R. Hamza (1), S. Fattoum (2), Mr. Péchevis (3), J. Cook (3), R. Girot (4) contribution to the sociological analysis of the impact of sickle cell disease in families of northern Tunisian public Health, 1999, volume 11, No. 3, p. 297-31
- [12] Rees DC, Olujohungbe AD, Parker NE, Stephens AD, Telfer P, Wright J. Guidelines for the management of the acute painful crisis in sickle cell disease. Br J Haematol 2003;120(5):744
- [13] HAS / S / management of sickle cell disease in children and adolescents Guidelines Department / September 2005.
- [14] van der Dijs FP, Fokkema MR, Dijck-Brouwer DA, Niessink B, van der Wal TI, Schnog J-J, et al. Optimization

of folic acid, vitamin  $B_{12}$ , and vitamin  $B_6$  supplements in pediatric patients with sickle cell disease. Am J Hematol 2002;69(4):239-46

- [15] Vilelam.m.s., sampaiom.s.c., costa f.f., brandalises.r.Reduce phagocytic activity mediated by C3b Fc monocyte receptors from children with sickle-cell disease.Brazilian J Med Biol Res 1989;
- [16] Cetiner S., Akoglut.f., Klinc there., Akoglu e. Kumi m. Immunological studies in sickle cell disease: comparison of homozygote mild and severe variants.ClinImmunol Immunopatho1989; 53: 32-9.
- [17] Hofmann M, de Montalembert M, Beaucquier Maccotta-B of VillartayP, Golse B. Post traumatic stress syndrome in children Affected by sickle cell disease and Their Parents. Am J Hematol.2007; 82: 171-2.

