Major acute pain complications in Congolese children with sickle cell disease

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Abstract

Objective: Sickle cell disease is the most frequent genetic blood disorder in the world and pain complications are the most prominent clinical aspects in sickle cell population. This study aims to describe the morbidity feature of major acute pain complications in children with sickle cell disease and outcome.

Material and Methods: 166 sickle cell disease children (4.3%) with a mean age of 85.31 months old±60.40 admitted in the pediatric intensive care unit for major acute pain complications were enrolled in the study from January to December 2014. Major acute pain complications were defined as acute sickle cell related pain with an intensity assessed at 3 on the Wong-Baker faces pain rating scale.

Results: Over the period study, 118 patients were admitted for diffuse vaso occlusive crisis (70.09%). Localized vaso occlusive crisis occurred in 13.25%, acute chest syndrome in 09.64% and hand foot syndrome in 06. 02%. The most common trigger factors were bacterial infections (65.12%) followed by malaria (34.88%). Mortality of 12.05% was recorded while 27.71% were discharged and 60.24% transferred to other departments. The mortality rate was significantly higher in the range age over 5 years (p=0.004)

Key words: Sickle cell disease, Major acute pain, Pediatric Intensive care department, Congo.

Introduction

Sickle cell disease (SCD) is the most prevalent blood disorder in the world. The highest prevalence occurs in Sub-Saharan Africa where 75% of 300, 000 annual births are born in Africa (1). By the year 2015, there will be 50% increased in the number of affected births (2). SCD is a real public health issue in the Sub Saharan region that contributes to major the rate of mortality among children whose data varies according studies (3,4). Acute pain complications are major symptoms of SCD and principal of cause hospitalization of children with sickle cell disease. In Africa they account in the pediatric intensive department for 1.8 to 6% (5, 6).

SCD has a prevalence of 0.9% in the Congo Brazzaville and is also a public health issue. The department of public health in the Congo has introduced diverse interventions such as free consultations for children in pediatric outpatient department at Teaching hospital and Sickle Cell Center. However, mortality remains high and the pattern of major acute pain complications in the PICD is unknown in the Congo. This current study describes the pattern of major acute pain complications in SCD children and their outcome in the pediatric intensive care in the Teaching Hospital of Brazzaville in the Congo

Material and Methods

The study was conducted from January to December 2014 in the department of pediatric intensive care in the Teaching Hospital at Brazzaville.

Brazzaville is the biggest city of the Congo located in the south part of the Republic of Congo with a population of 4 millions. The Teaching Hospital is the biggest medical facility of the country where attend to almost all SCD patients living in Brazzaville. There is in Brazzaville only one pediatric intensive care department (PICD) that receives and manages all life threatening medical emergencies of children aged 1 month to 17 years old.

All medical records of children admitted during our study in the PICD were reviewed. Medical records with diagnosis of major acute pain complications were enrolled in the study. The diagnosis of SCD disease has been confirmed by hemoglobin electrophoresis on children that were identified initially from previous medical records.

Definition

Acute pain sickle cell episodes related are pain symptoms linked to the presence of abnormal hemoglobin (HbS). They can occur spontaneously or precipitated by infection, stress or dehydratation.

Acute pain episodes are defined as major when, the intensity is assessed at 3 on the Wong-Baker faces pain rating scale. Acute pain episodes can be limited or diffuses when they affect multiple sites [9, 10].

The diagnosis of infection was based by the following clinical findings: fever over 38.5C or hypothermia<36.5C, tachypnea> 40 breaths per minute, "toxic-looking" associated with symptoms specifics to the infection developed.

Bacterial infections were: pneumonia, meningitis, angiocholitis, Acute Pyelonephritis and sepsis. Parasitical infectious were malaria. Pneumonia was defined as the presence of abnormal breath sounds and pathological chest X-ray. Meningitis was defined as stiff neck and abnormal cerebral spinal fluid. Angiocholitis was diagnosed when patient was presenting jaundice, pain in the right upper quadrant and mild hepatomegaly. Acute Pyelonephritis was associated flank pain, nausea, vomiting with positive urinary strip and urine culture. Hence, sepsis was defined by tachycardia, polypnea leukocytosis or leucopenia, and positive C-reactive protein. Report of blood culture are usually not released within a week, therefore are not available when in the patients are in the PICD, that is why distribution of microorganimes are not provided in the present study.

Aches and positive smear were defining malaria

Data abstracted from records were variables related to children: gender, age, length of time between beginning of symptoms and hospitalization, pre hospital consultation, length of hospitalization, diagnosis and outcome.

Patients

During the study period, 3,620 children were admitted in the PICD. Among them 188 (5.6%) children were diagnosed for acute SCD complications. Among the 188 children, 166 (4.2) were hospitalized for major acute pain episodes. Major acute pain complications was the first cause of hospitalization (88.30%) followed by hyperhemolytic crisis.

Statistical analysis

Data were entered into Microsoft Excel and analyzed SPSS (Chicago-USA). Categorial variable were described as proportion (percentage), continuous variables were described as medians (inter-quartile range) Differences between proportions were calculated using Fisher's exact test The P-values or less than 0.05 were considered to be statistically significant.

Results

One hundred sixty six were enrolled in the study. They were 86 boys (51.81%) and 80 girls (48.19%) with a sex ratio of 1.07. Children were aged between 24 and 192 months old (mean age 85.31 ±6.40 months old). Almost two-third (72.89%) were below 5 years of age. In this study 94 children (56.63.7%) were coming from their homes, whereas 48 children (28.92%) were referred by primary and secondary medical facilities.

The time interval between the beginnings of the symptoms and the admission to the PICD ranged from 2 to 12 days (mean 3±1.1 days), whereas the duration of hospital admissions in PICD ranged from 1 to 16 days (mean 3±1.5).Table 1.

Table 1: Characteristics of 166 children with SCD admitted in PICD for major acute pain complications

Characteristics	
Gender n (%)	
Female	92 (48,9)
Male	96(51,1)
Sex ratio	1.04
Age (month)	
Mean ±SD	69.26±50.4
Min-Max	6-192
Range of age (months) n (%)	
1-24	57(30.32%)
25-60	64(34.04)
61-120	41(21.81)
>120	26(13.83)
Duration of illness (days)	
Mean ± SD	3±1.5
Min-Max	2-12
Coming from n(%)	
Teaching hospital departments	24(12,8)
Secondary health facilities	28(14,9)
Primary health facilities	20(10,6)
Home	116(61,7)
Lifetime hospitalization (days) n%	
1-2	88(46,8)
3-8	68(36,2)
9-16	32(17)

Table 2 summarizes the distribution of major acute pain syndrome. Diffuse vaso occlusive was the most common major acute pain episode observed in 71.09% patients followed by limited vaso occlusive (13,25%) and acute chest syndrome (09.64%).

Major acute pain syndrome were associated with infectious factors in 90.96% (n=151), without factors noticed in 09.04% (n= 15). Infections were due to bacterial infections (65, 12%) and malaria (34.88%).

Among those bacterial infections, pneumonia was observed in 61 patients (67.78%), pyelonephritis in 12 (13.34%), meningitis in 8 (08.89%), septicemia in 5 (05.56%) and angiocholitis in 4 (04.44%).

Table 2: SCD Severe acute complications and trigger factors

Sever acute complications	N	%
Severe acute hyperhemolyis crisis N	88	46.81
Severe mixte crisis	52	27.66
Severe vaso occlusive crisis	36	19.15
ACS	4	11.11
Stroke	12	6,38
Total	188	

Table 3 shows that 46 (27.71%) patients were discharged, while 100 patients (60.24%) were transferred to other departments and 20 (12.05%) died. A comparison of outcome by range of age showed statistically relationship. Mortality rate was the highest when the children were older than 120 months (p=0.04).

Table 3. Outcome of the 188 children admitted in the PICD for SCD severe acute complications. ***P=0.004**

Outcome N(%)	
Discharged	46 (24.47)
transferred	110 (58.51)
death	32(17.02)
Age when died* (months)	
1-24	2(12,5)
25-60	0(00)
61-120	12(37.5)
>120	16(50)

Discussion

Demographics range of non major acute pain syndrome SCD related is fully identified in Brazzaville since it has been the subject of several publications (11-13). However, pattern of major acute pain complications SCD related have received little attention even though they have been reported to increase the mortality of children with SCD (11,13,14). The present study is the first in the Congo to attempt to report the feature major acute pain complications SCD related in children admitted in the PICD, and their outcome.

There is only one PICD in the Brazzaville region, which is the biggest city of the country. That explains the high proportion of children (28.92%) referred by primary and secondary health institutions. Our results should be interpreted bearing in mind the study setting. Indeed, data were collected in the PICD that admits only children aged 1 month to 17 years old, therefore had an inbuilt bias toward children aged 18 years. However, as we said previously, the PICD is unique; this study probably included all the majority of SCD children living in Brazzaville and then gives a good picture distribution of SCD severe acute complications in Brazzaville.

One hundred eighty eight children (5.6 %) were admitted to the PICD for complications of sickle cell disease which is remarkable lower than Abhulimhen's report but higher than Anyanwu's findings (5, 15).

Our population study is young with a median age of 85.31 months (7 years). Among them, nearly two third of the population study were aged under 60 months (5 years). Similar age distribution has been described by different authors (5, 15-17). The first cause of admission in PICD with similar mean age is major acute pain complication (5, 15-17).

Feature of SCD's complications vary according SCD children range of age (15). The authors agree on the predominance of hyperhemolytic crisis before the age of 5 vaso occlusive crises after the age of 5 (11-13). Nutritional deficiencies, acute chronic sequestration more frequent before the age of 5 to which is added malaria high infestation degree in Sub-Saharan Africa region are many aggravation factors of acute hyperhemolytic crisis that justify the clinical pattern distribution (14).

Non implementation of penicillin prophylaxis, pneumococcal and typhoid fever vaccines lead Infections to be first factors associated with sickle cell morbidity in children (10-18). Despite the fact that lake of effective laboratory facilities restricted the accurate diagnosis of bacterial acute febrile illness, distribution of the type of severe infection, in this current study was found to be similar to those described elsewhere (11,13,19-21). Pneumonia has been recognized as the main severe infection among sickle cell disease children. In fact, this clinical finding is not specific to the sickle cell disease population, since respiratory infections at the national level are the second cause of hospitalization in health facilities in the Congo (22).

Bacterial infections share similar findings between PICD and conventional pediatric departments in children with SCD. Meningitis accounted 08.89% in our study, 7.3% in Senegal and 6% in Nigeria (5,18). Septicemia rated 05.56% in our study and 4.1% in France (23). The prevalence of pyelonephritis (13.34%) was barely higher to the one reported in Brazzaville in conventional pediatric department, 10% à Brazzaville (11). However, osteomyelitis rate was higher than conventional pediatric departments (12,14,17).

Malaria is after bacterial infections the most common trigger acute sickle cell crisis in our region and second cause of hospitalization in sickle cell and not sickle cell pediatric population (5,6). Both population sickle cell and not sickle cell share comparable rate, even though we reported higher rate in our study (23). However, cerebral malaria is exceptional in the sickle cell population (21, 23,24).

ACS is an acute non infectious pulmonary complication seen in patients with sickle cell disease. It is one of the major causes of morbidity and mortality among children (25, 26). ACS's symptoms overlap with those of pneumonia and then can be sometimes very difficult to diagnose. That explains the quite diverse distribution reported (8,11,13,18).

Only one fourth of the patients (27.71%) were discharged while 20 (12.05%) were death. The outcome shows more severe outcome of the disease if we compare with Nigeria's findings (5,6,15,17).

Sickle cell disease in the western part of Africa, where the Nigeria is located, is known to have less severe morbidity and better outcome. Indeed, phenotypic modulator factors such high HbF levels, co-inheritance of α thalassemia and predominance of Senegal and Benin haplotypes are factors that may explain this difference (27).

The highest incidence of death is usually reported the in the first 5 years of life because children are at increased risk of life threatening infections due to absent or non functional spleens and a decreased immune response (28).

Paradoxically, in our study the lowest mortality rate is shown in that rate of age. That particularity also raised by Diagne and al deserves to be confirmed substantiated more thorough prospective study (18).

Conclusion

Major acute pain complications are predominantly marked by diffuse vaso occlusive crisis and triggered by infections. This enhances the relevance of early intervention as neonatal screening, penicillin prophylaxis, immunization but also malaria prophylaxis

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