DOI: https://doi.org/10.53350/pjmhs2023173821

ORIGINAL ARTICLE

Skeletal Problems in Sickle Cell Anemia Children

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ABSTRACT

Background and objective: Red blood cells in sickle cell anemia (SCA) take on an unnatural, stiff, sickle-shaped appearance. The musculoskeletal problems of sickle cell anemia (SCA) stem from the fact that blood vessels get blocked, which causes tissue ischemia, infarction, and gradual damage to end organs. The researchers in this study set out to identify common musculoskeletal problems in children diagnosed with SCA.

Methods: In this retrospective study, 140 children with sickle cell anemia were included. Musculoskeletal problems, subject age, and gender make up the variables that are examined. In order to compare means, we utilized the Mann Whitney U-test, and to check for significant associations between categorical variables, we utilized Fisher's exact. For statistical significance, a P-value lower than 0.05 was considered.

Results: Among all, 86 (61.4%) were males and 54 (38.6%) were females. 3 (2.1%) cases had age < 1years, 45 (32.1%) cases had age 1-5 years, 60 (42.9%) cases had age 6-10 years and 32 (22.9%) had age 11-15 years. Pain bone crisis 55 (39.3%) was the most common cause of musculoskeletal disorder. Frequency of mortality was 10 (7.1%). Physiotherapy referrals were 13 (9.3%).

Conclusion: Due to the fact that sickle cell disease affects more individuals than expected, screening is not widespread, and clinically proven, multidisciplinary treatment is scarce, many people are poorly managed. Musculoskeletal complaints can be treated with physiotherapy, although its relationship to the underlying condition is rarely acknowledged.

Keywords: Physiotherapy, Musculoskeletal complications, Sickle cell anaemia,

INTRODUCTION

The sickle cell anemia gene, which can be passed down from one generation of blood cells to the next, is characterized by the presence of sickle hemoglobin (Hb S). In rare instances, this gene can be passed down from both parents. Sickle cell anemia is the most common and dangerous form of the blood disorder. Hemoglobin S results from a mutation that swaps out adenine for thymine at the sixth codon of the beta-chain gene, moving it from GAG to GTG. During coding, this causes the Hb beta chain to have valine instead of glutamate at position 6. The resulting hemoglobin has the physical properties of a polymer when exposed to deoxygen. Its molecular stability and solubility can also change. These traits are responsible for the severe clinical signs of sickling syndromes[1,2]

Ischemia and arterial occlusion create a wide range of symptoms in those who are affected. Common symptoms include recurrent pain and worsening infarction. When hemoglobin S is present, red blood cells can change shape during deoxygenation, shifting from a biconcave disk to a crescent or sickle. When red blood cells are reoxygenated, they first assume their normal shape. But after many rounds of "sickling and unsickling," the erythrocyte hemolyzes due to irreparable damage[3]. Hemolysis causes anemia, which is a hallmark of sickle cell disease. Damage to tissues, both immediate and long-term, can result from a reduction in blood flow due to the aberrant shape of red blood cells[4]. Possible side effects include the following: acute chest syndrome, priapism, CVAs, renal and splenic malfunction, and painful episodes affecting bones and soft tissues. Magnetic resonance imaging (MRI) is an important tool for the diagnosis of anomalies and multisystem illnesses caused by sickle cell disease. Understanding the full scope of imaging data is crucial for accurately diagnosing the condition and starting the right treatment.[5].

In sickle cell anemia (SCA), the red blood cells take on an abnormal, rigid, sickle-shaped appearance; this disorder is inherited from one generation to the next. Sickle cell disease, which causes cells to become less elastic, is connected with several dangers[6]

Sickle cell diseases, which originated in the tropics and subtropics, have now spread to every part of the world due to individuals migrating to the temperate zone[7] The incidence rate of SCA in Nigeria ranges from 0.4% to 3%, affecting around 20 out of 1,000 neonates. The majority of sickle cell diseases (85%) and the majority of sickle cell newborns (70%) are found in Africa[8] The sickle cell trait is present in at least 5.2% of the world's population. As a result of tissue loss, infarction, gradual injury to end organs, and aberrant bone marrow growth brought on by blocked blood arteries, musculoskeletal disorders can be observed in SCA[9] Bone marrow hyperplasia causes the medulla to swell and the cortical layer to shrink. The typical trabecular pattern becomes coarser and cortico-medullary distinction disappears in both long and flat bones as a result of this.Musculoskeletal issues are common as the disease advances. Some examples of these conditions include growth defects, pathological fractures, osteomyelitis, septic arthritis, femoral head aseptic necrosis, soft tissue abnormalities (such as dactylits, anomalies in growth, myonecrosis, leg ulcers, and pyomyositis), and pathological fractures.It is unacceptable that patients with SCD do not receive comprehensive care, even if there have been few studies that highlight the significance of PT in this population[10,11] Actually, research in Nigeria has only focused on a subset of musculoskeletal SCA symptoms. Consequently, this study aims to quantify the prevalence of musculoskeletal problems, the types of therapies offered (including physiotherapy), and the mortality rate in SCA patients.

MATERIALS AND METHODS

In this retrospective study, 140 children with sickle cell anemia were included. The painful and passive limitations of hip motion were described by this study as avascular necrosis of the head of the femur. Radiographs taken of the hip from the front and back confirmed this. However, fistulous tracts, low-grade inflammation, sequestrum, and new bone apposition can develop over the course of months to years due to chronic osteomyelitis (COM), a recurring, persistent infection. Radiographs showed a fractured left fifth metacarpal bone and a wedge collapse of the spinal column with Gibbus.

Individual patient records were reviewed by investigators after retrieval from the hospital's records department. Researchers used cellulose acetate electrophoresis at pH 8.6 to identify patients with hemoglobin genotype SS, and they ranged in age from 6

months to 15 years. Inclusion criteria for SCA in children did not include the presence of any congenital musculoskeletal abnormalities, inadequate data, or an ambiguous diagnosis. The research team behind this study set out to find out how many kids in this age group experienced musculoskeletal issues from SCA, as well as the distribution of these complications by gender.

The tools used for data analysis were Graphpad Prism 5 and SPSS version 24 (Chicago, IL, USA). The first step was to tally the frequencies of all the variables. Age and gender distributions, as well as the different musculoskeletal problems, were among the variables examined. For categorical variables, we utilized Fisher's exact test for significant correlation, and for continuous variables, we compared means using Mann Whitney U-test. Tables were used to display the data. The provided P values were all two-sided, and we considered them significant if they were less than 0.05.

RESULTS

Among all, 86 (61.4%) were males and 54 (38.6%) were females.(figure 1)

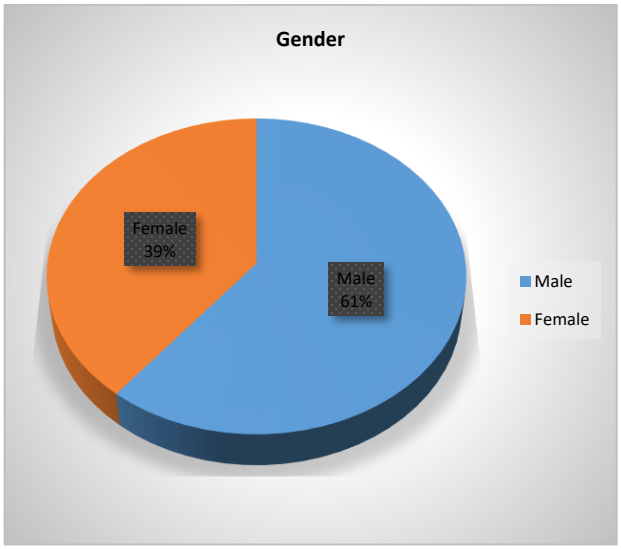

Figure-1: Gender distribution

In our study, 3 (2.1%) cases had age < 1years, 45 (32.1%) cases had age 1-5 years, 60 (42.9%) cases had age 6-10 years and 32 (22.9%) had age 11-15 years.(figure 2)

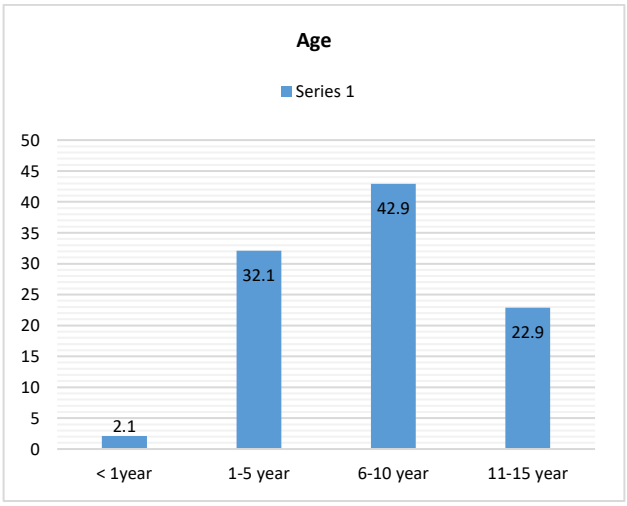

Figure-2: Age of the presented children

Pain bone crisis 55 (39.3%) was the most common cause of musculoskeletal disorder followed by osteomyelitis, avascular necrosis, low back pain, Chronic leg ulcers, vertebral collapse and pathological fracture.(table 1)

Table-1: Frequency of musculoskeletal disorder

Musculoskeletal disorders	Frequency (140)	Percentage
Pain bone crisis	55	39.3
Osteomyelitis	25	17.9
avascular necrosis	20	14.3
low back pain	14	10
Chronic leg ulcers	12	8.6
vertebral collapse	8	5.7
pathological fracture	6	4.3

Frequency of mortality was 10 (7.1%). Physiotherapy referrals were 13 (9.3%).(table 2)

Table-2: Mortality and frequency of referrals

Variables	Frequency	Percentage
Mortality		
Yes	10	7.1
No	130	92.9
Physiotherapy referrals		
Yes	13	9.3
No	127	90.7

DISCUSSION

Our SCA patients had a prevalence of musculoskeletal problems of 40%. In Yaoundé, its predominance differed from Dipty15 in India and Bahebeck[12]. The prevalences in India were 7.8% and 41.6%, according to Bahebeck[13] in Dipty et al.[14]. Differences in sample size and location may account for this discrepancy. Acute osteomyelitis was the most prevalent musculoskeletal consequence, occurring in 12.8% of cases. This rate differed with the reports of Ejindu et al.[15], who found it to be 18%, and Bahebeck et al.[12], who looked at Yaoundé. Patients with sickle cell disease have an elevated risk of osteomyelitis, which has been known for a long time. Hyposplenism, reduced complement activation, and infarcted or necrotic bone are some of the suggested reasons.[16]

In order to cultivate S. aerus, blood samples were taken from children who suffered from acute osteomylitis and SCA. The outcomes deviated slightly from the typical pattern. From a buruli skin ulcer, Mycobacterium ulcerans can travel throughout the body, causing osteomyelitis in sickle cell disease and tuberculosis, among other complications[17,18] Neither TB nor buruli ulcers were present in our patient group. Most of our patients had access to radiographs, which allowed us to catch musculoskeletal problems early. As an illustration of the crucial relevance of radiographs in the diagnosis of musculoskeletal disorders in SCA, a couple of our patients with unusually widespread lower limb sensations resembling a vaso-occlusive episode had osteomyelitis, as shown on radiographs. The observation that radiographs of avascular necroses of bones can initially reveal no abnormalities before exhibiting the characteristic indications of sclerosis, subchondrial collapse, and flattening was supported by Ware et al.[19]. It may be difficult to distinguish between acute osteomyelitis and bone infarction based on radiological and clinical indicators. In some cases, magnetic resonance imaging (MR) could be useful. Cerebral abnormalities, nearby fluid accumulation in soft tissues, and an enlarged bone marrow are all signs that might point to an infection on magnetic resonance imaging (MR). Since we lacked the appropriate equipment, we opted not to include MR imaging in our series. Our suspicion that musculoskeletal problems in SCA children became worse with age was validated by our investigation. This could be due to an increase in hypoxemia or infarction, the coagulation system's progression from infancy to adulthood (also known as developmental homeostasis), or some other factor[20]

The study found that bacterial infections were the leading cause of death among SCA patients, accounting for 7.1% of the total patient mortality rate[21] life expectancy of sickle cell patients has increased dramatically due to better diagnosis and treatment, which is why current studies have revealed a reduced mortality rate among SCA patients. A century ago, the life expectancy of SCA patients was far lower than it is now, according to new data[22] is a The high rates of sickness and early mortality caused by sickle cell disease, however, impose a heavy financial burden on society. Carriers (those with the sickle trait) did not cause clinical pathology in the majority of cases, according to Oni et al., hence no one was admitted to the study as a carrier of the disease[22]

CONCLUSION

Due to the fact that sickle cell disease affects more individuals than expected, screening is not widespread, and clinically proven, multidisciplinary treatment is scarce, many people are poorly managed. Musculoskeletal complaints can be treated with physiotherapy, although its relationship to the underlying condition is rarely acknowledged.

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