ORIGINAL ARTICLE

Incidence and management of priapism in sickle cell disease: A study from local hospital at south west Saudi Arabia

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Abstract

Objectives: To review the incidence, clinical presentation and management of sickle cell disease patients presenting with priapism.

Material and methods: A prospective analysis of data base of total 212 male patients with sickle cell disease (43 patients) presented with priapism was performed. All data was transferred into a structured questionnaire which included patient's demographic features, treatment given, duration of hospital stay and outcomes. The SPSS version 19 was used for statistical analysis.

Results: Out of 212 male patients with sickle cell disease 43 (20.3%) male patients complaints of priapism. 27 patients came in emergency and they were admitted and given treatment and other 16 complain in OPD that they experience priapism. 5 patients (11.6%) know about priapism as a complication of sickle cell disease. All patients who came in emergency due to priapism were admitted and given initial treatment which consisted of intravenous fluid and aspiration/irrigation with normal saline. 5 patients were irrigated with dilute epinephrine (Adrenaline 1:1,000,000) Oral etilefrine therapy was prescribed for adults and children over 12 years of age. The self-administered intracavernous injection (SICI) technique was used in 7 patients who were very conscious about their sexual ability and impotence.

Conclusion: Priapism is common in patients with sickle cell disease mostly in age group 20 to 25 years. It carries a significant role in impotence. It can be prevented by conveying knowledge to patients with sickle cell disease about priapism, appropriate diagnosis and prompt treatment. It has been suggested that with appropriate treatment preservation of potency can be achieved.

Keywords: Sickle cell, priapism, sickle cell disease, impotence, Hemoglobin S, Hemoglobin F

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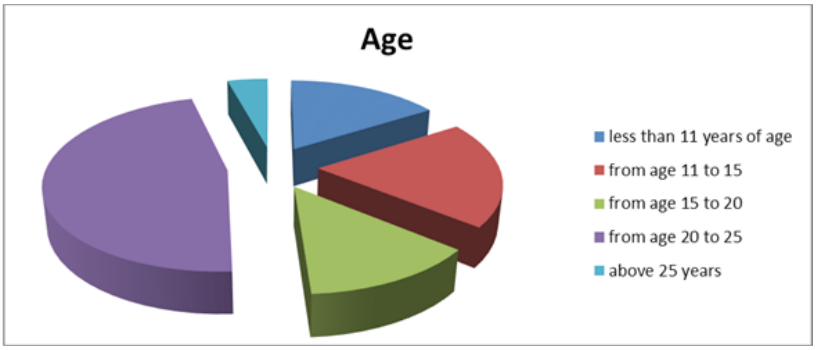
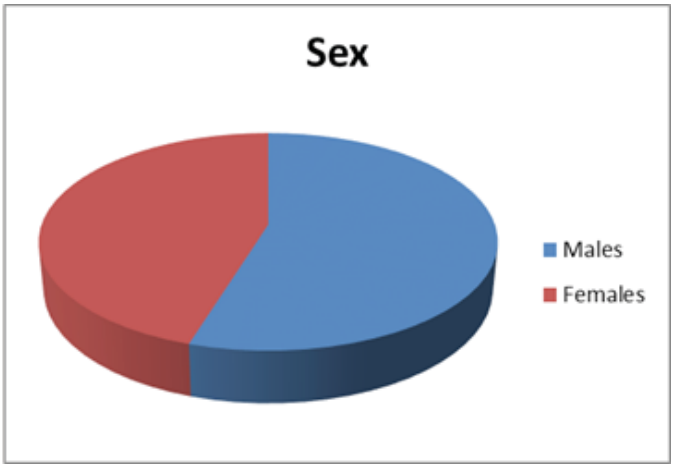
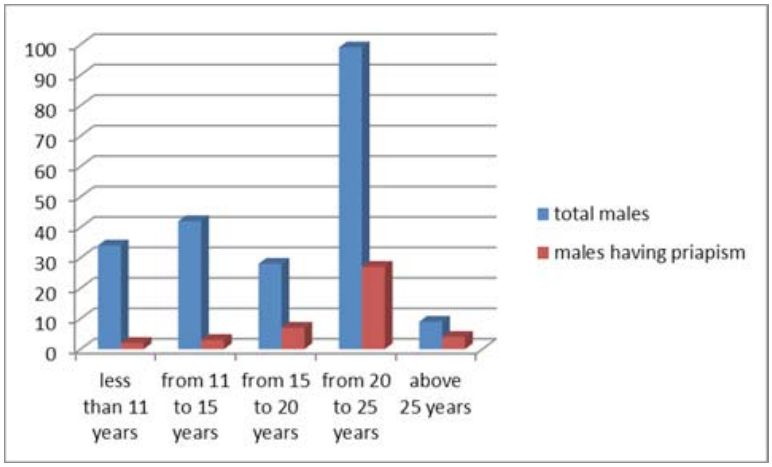
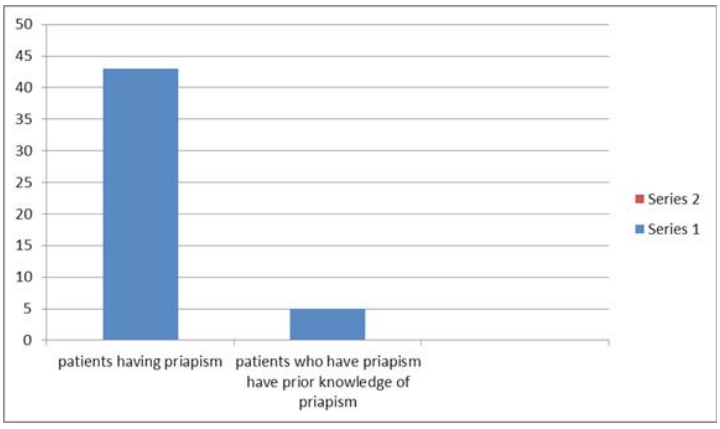
Introduction:

Sickle cell disease (SCD) is a group of inherited blood disorders caused by genetic mutations in the HB b - gene, resulting in abnormal hemoglobin commonly found among people of tropical countries and transmitted as autosomal recessive disorder. If a person inherit only one gene responsible for sickle hemoglobin from either of the parent, the condition is called carrier or trait. If one receives two defective genes, one from each parent, the condition is called SCD. Person with trait leads a normal life but the diseased person suffers from various complications throughout the life, such as anemia, bone & joint pain, joint swelling, recurrent infection, osteomyelitis, necrosis of bone, aplastic crises, abdominal pain, splenic sequestration crises, hepatosplenomegaly etc. SCD usually manifests early in childhood. For the first 6 months of life, infants are protected largely by elevated levels of Hb F. Approximately half the individuals with homozygous HbS disease experience vaso-occlusive crises.[1] Acute priapism (AP), a stubborn painful erection lasting more than 3 hours and that affects 42% of adults and 6% of children. Due to advance in medicine there is increase life expectancy of sickle cell patients, priapism has become one of the most common complication (vaso-occlusive) type and it requires prompt diagnosis and early management.[2]









Priapism is a condition of painful, purposeless, persistent penile erection, first described by Tripe[3] in 1845, and first associated with sickle-cell disease in 1934.[4] Classically, there are two types of priapism that are described as low-flow I.e (ischaemic) and high-flow i.e. (not ischaemic), flow refers to the oxygenation of blood that goes in the corpora. The ischemic type is very painful condition and laboratary finding shows hypoxia. Priapism in sickle-cell anaemia is a typical form of the ischemic that is lowflow type.

Priapism is a very common problem among male patients having sickle-cell disease.[5]

Methodology:

A prospective analysis of 386 patients (212 male patients) diagnosed assickle cell disease from 2012 to 2015 was accomplished at a local hospital at south west of Saudi Arabia. Male patients were confirmed with sickle cell disease on haemoglobin electrophoresis, were eligible for the study. All females were excluded from the study.

All data was transferred into a structured questionnaire which included patient's demographic features, treatment given, duration of hospital stay and outcomes. The SPSS version 19 was used for statistical analysis.

Results:

Total population size of our study was 386 sickle cell disease patients, among them 45.1% (174) were females and 54.9% (212) were males. Because our study is based on priapism so all the females were excluded from the study. The induction of the entry is noted according to their ages. Among 212 males less than 11 years of age were 16% (34), from age 11 to 15 years were 19.8% (42), from age 15 to 20 years were 13.2% (28), from age 20 to 25 years were 46.7% (99) and patients of more than 25 years were 4.24% (9) respectively with mean age was 19.28. We follow these patients for 3 years Among these male patients with sickle cell disease 43 patients (20.3%) complain of erection for more than 2 hours unrelated to sexual interest or stimulation and progressive penile pain, among these

43 males 27 came in emergency other 16 came in OPD. Among these 43 patients with priapism less than 11 years were 2 patients, less than 15 years were 3 patients, from 15 to 20 years were 7 patients , from age 20 to 25 were 27 patients and more than age 25 were 4 patients.

Out of 43 patients only 5 patients (11.6%) know about priapism as a complication of sickle cell disease.

All patients were diagnosed previously with sickle cell disease by hemoglobin electrophoresis. The patients who came in emergency with complain of erection for more than 2 hours unrelated to sexual interest or stimulation and progressive penile pain were admitted.

All patients who came in emergency due to priapism were admitted and given initial treatment.

Initial resuscitation consisted of intravenous fluid and aspiration/irrigation with normal saline. Aspiration and irrigation were carried out on the lateral mid-shaft of the penis at 3 o'clock or 9 o'clock using a 19 gauge butterfly needle. Aspiration is then done with a 10 or 20 ml syringe with intermittent injection of normal saline. At conclusion of the procedure, the puncture site is compressed for up to 2 min to reduce hematoma formation. Pain killers are also given as initial management.

Patients are encouraged to drink plenty of water and urinate.

5 patients were irrigated with dilute epinephrine (Adrenaline) which was very effective in producing immediate and sustained detumescence because they were having very painful erection. Adrenaline 1:1,000,000: dose 10 mL was used to irrigate.

Oral therapy was adviced for adults and children over 12 years of age. Etilefrine was given daily two times a day (morning and evening) (15 mg) for prevention of episode of priapism.

We use self-administered intracavernous injection (SICI) methodin 7 patients who were very conscious about their sexual ability and impotence. We taught our patients how to inject themselves with 6 mg of undiluted etilefrine (0.6 mL of 10 mg/mL solution) with a 1 mL syringe and 30-gauge needle in the right or left sideofpenis. We advised S1CI to patients if a episode lasts more than 1 hour or if the patient awakes with a painful pulsatile erection.

Discussion:

Recent endeavours to explicate the exact incidence of priapism in this group of patients have shown a much higher prevalence than antecedently apperceived. Mantadakis et al.[6] identified that up to 28% of patients aged 5–20 years will undergo an episode of priapism. Our present survey, which included a wider age group, corroborated similar findings, with 20.3% of patients experiencing their first episode by the age of 25 years. There was a designation that the incidence stabilised by the third decade of life. The possible inequitableness introduced by the different replication rates in the UK and Nigeria is acknowledged, but interestingly the prevalence of priapism was homogeneous in the two groups. The high life-time probability of priapism is consequential and provides incipient insights into the magnitude of the quandary among patients with sickle-cell disease.

Stuttering priapism in particular is a quandary which afore these patients were simply prone to abide.over half the patients in the currentstudy had consult about this problem. There is published-proof which shows that patients having sickle cell disease lack awareness about priapism that it is a complication of sickle-cell disease. Our study shows only 11.6% of patients who had experienced priapism were aware that priapism was a complication of the disease.

Report indicates patients have now learnt to deal with problem by variety of self help treatments. Recent studies suggest self help treatment ,like simple advice about analgesia, hydration and exercise can now be offered to patients as self-help strategies to curtail an attack of priapism. So we advice all our patients about taking self help treatment.Warm bath and voiding also have

a high benefit so we advice our patients to take warm bath.[6]

There is always a debate about management for acute priapism. There are two types of management surgical management and conservative management. Conservative management always have a upper hand on surgical management because it preserves potency. In a comprehensive review by Hamre et al.[7] the preservation of potency appeared to be more successful with conservative, i.e. non surgical treatment. The patients who were treated conservatively maintained erectile potency. In practice, there is no role of surgical intervention and it is often reserved for the most severe and prolonged cases, and it is possible that subsequent erectile dysfunction is related as much to the duration of acute priapism as to the surgical intervention. Early conservative management by penile aspiration and irrigation with α-adrenergic agonists has recently been shown to be successful, even in an outpatient setting, with no incidence of erectile dysfunction at a median follow-up of 40 months.[8]

Acute priapism is that complication of sickle cell disease that can be prevented. Limiting the duration of an episode of priapism is very important. In modern era of medicine priapism is reduce by successful medical intervention such as hydroxyurea which induce the bone marrow to increase the production of fetal haemoglobin, have been tried, with anecdotal reports of success.[9] We use (SICI) in 7 patients for preservation of potency. Intercavernosal injections of etilifrine which are self administered by patients are reported to have some success in prevention of priapism and preservation of potency.[10]

Current guidelines for the management of ischemic priapism recommend initial corporeal aspiration and injection of sympathomimetics.[11] In the cases of ischemic priapism of SCD origin, hydration, oxygenation, and systemic alkalinization can be done. When conservative management fails or where presentation occurs later, the surgical options are to be considered.

Men with SCD should be educated about need of seeking treatment immediately for any episode of priapism that lasts longer than 2 hours.

Use of epinephrine is very useful and effective in producing immediate and sustained detumescence[12] as we use in 5 patients as they were having very painful erection.

Conclusion:
Priapism is common in patients with sickle cell disease mostly in age group 20 to 25 years. It carries a significant role in impotence. It can be prevented by conveying knowledge to patients with sickle cell disease about priapism, appropriate diagnosis and prompt treatment. It has been suggested that with appropriate treatment preservation of potency can be achieved.

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Role and contribution of authors:
Dr Muhammad Yahya, Assistant Professor at Hamdard Universtiy Hospital conceived the idea, collected the data and wrote the initial write-up.

Dr Urwah Inam, collected data and also helped in collecting the references and discussion wrirting

Dr Masood Raza Khan, consultnat general surgeon at Kind Fahad hospital Madina, criticaly review the article and gave them final touchup.

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