facilities. The skin in these patients shows normal sweat glands but their intervention by the post-ganglionic sympathetic fibres is defective. When external temperature is raised, as happens in hot summer months in Pondicherry, patients have inability to sweat, leading to alteration in thermoregulation and spells of high grade fever requiring lowering of external temperature by physical means like cold/ice sponging or air conditioning. This method proved successful in preventing heat exhaustion in the long term management of our patient.

## REFERENCES


1. Swanson AG. Congenital insensitivity to pain with anhidrosis: a unique syndrome in two male siblings. Arch Neurol 1963, 8: 299-306.
2. Pinsky L, Digeorge AM. Clinical familial sensory neuropathy with anhidrosis. J Pediatr 1966, 68: 1-13.
3. Vassella F, Emrich HN, Kraus-ruppeat R, Aufdermaur F, Tonzo. Congenital sensory neuropathy with anhidrosis. Arch Dis Child 1968, 43: 124-130.
4. Mazar A, Herold HZ, Vardy PA. Congenital sensory neuropathy with anhidrosis: Orthopedic complications and management. Clin Orthop 1976, 118: 184-187.
5. Gillespie JB, Peucca LG. Congenital generalized indifference to pain. Am J Dis Child 1960, 100: 124-128.
6. Swanson AG, Buchan GC, Alvrod EC. Anatomic changes in congenital insensitivity to pain. Arch Neurol 1965, 12: 12-18.
7. Murray TJ. Congenital sensory neuropathy. Brain 973, 96: 294-387.
8. Denny-Brown D. Hereditary sensory radicular neuropathy. J Neurol Neurosurg Psychiatry 1951, 14: 237-252.
9. Thrush DC. Congenital insensitivity to pain: A clinical, genetic and neurphysiological study of four children from the same family. Brain 1973, 96: 369-386.


# Priapism in Children with Sickle-Cell Disease


**E.O. Enwerem**
**E.M.L. Endeley**
**C. Holcombe**
**R.V. Patel**


Priapism is a painful and persistent penile erection. Majority of cases are primary (idiopathic), present in middle age and require surgical treatment(1). We wish to report two pediatric cases of secondary priapism due to hematological disorder of sickle-cell disease leading to slugging of blood within the corpora cavernosa which were treated successfully with conservative measures.

## Case Reports

*Case 1:* A 9-year-old male child was admitted with a history of painful and persistent erection of penis associated with yellow sclera and high grade of fever with rigors of 2 days duration. He was a known case of sickle-cell disease (HbSS) diagnosed at the age of 2 years. The patient was in severe pain, febrile, mildly icteric and had pallor. Hepatosplenomegaly with tender erect penis was observed. Peripheral smear showed *P. falciparum*, features of sickle-cell anemia and polymorphonuclear


*From the Department of Surgery, College of Medical Sciences and University of Maiduguri Teaching Hospital, Maiduguri, Nigeria, West Africa.*

*Reprint requests: Dr. R.V. Patel, 210, A.G. Office Staff Quarters, Near Saurashtra University, Rajkot 360 005.*



*Received for publication: September 25, 1991; Accepted: February 6, 1992*

leucocytosis. Hemoglobin was 7.4 g/dl, total bilirubin 75 $\mu$mol/L, conjugated bilirubin 21 $\mu$mol/L, creatinine 54 $\mu$mol/L and urea 4.9 m mol/L. Serum electrolytes were within normal limits and hemoglobin electrophoresis confirmed HbSS disease.

He was put on conservative measures which included bed rest, sedation, analgesics, hydration with intravenous M/6 sodium lactate, folic acid, B-complex, multivitamins, antimalarials and Vitamin C. He responded within 24 hours and was relieved of priapism within one week.

*Case 2:* An 8-year-old male child who was a known case of sickle-cell disease was referred to us for possible surgical treatment for priapism of 3 days duration from a peripheral district hospital. On examination, he had moderately severe painful persistent erection of penis with mild hepatosplenomegaly and pallor. All laboratory investigations including renal and hepatic functions were within normal limits except for hemoglobin (7.2 g/dl). Hemoglobin electrophoresis suggested HbSS pattern. He was put on conservative measures described in Case 1 except antimalarials. He responded to treatment promptly and was relieved of symptoms within 10 days.

## Discussion

Priapism has been reported to occur in 2 to 5% of patients with sickle-cell disease. In sickle-cell disease, priapism is a well known, but fortunately, rare complication especially below the age of eight years(2,3). Anemia, hypoxia or stress of infection often precipitate these attacks(4).

Initial therapy, should include pethidine or morphine and rehydration. If still there is no response, the opinion of a pediatric or urosurgeon should be sought, but surgery is not often recommended(4). However, aspiration and irrigation under anesthesia may prove to be of great value. Under general or spinal anesthetic, a wide bore needle (14 or 16 gauge) is inserted into the lateral surface of the base of the penis and the dark viscous blood aspirated, this is followed by repeated irrigation with either saline or 10% heparin and by aspiration until only fresh blood is obtained.

Although priapism is uncommon in sickle-cell disease in children and responds most often to nonsurgical traditional management (hydration, analgesics, oxygen), early creation of a percutaneous shunt fistula between the glans penis and the corpora cavernosa should be considered in those patients in crisis who develop signs of priapism and who fail to respond to conservative treatment(5,6). The prognosis in pediatric cases is good as priapism rarely results in impotence in young sickle-cell patients.

## REFERENCES

1. Gupta RL. Priapism. Indian J Surg 1987, 49: 199-200.
2. Attach EB. The Pathology fo sickle-cell disease *In:* Sickle-cell Disease 1st edn. Ed Fleming AF. Edinburgh, Churchill Livingstone, 1982, pp 42-56.
3. Fleming AF. Sickle-cell disease during and after puberty. *In:* Sickle-cell disease, 1st edn. Ed Fleming AF. Edinburgh, Churchill Livingstone, 1982, pp 73-89.
4. Effiong CE. Sickle-cell disease in childhood. *In:* Sickle-cell disease 1st edn. Ed Fleming AF. Edinburgh, Churchill Livingstone, 1982, pp 57-72.
5. Tarry WF, Duckett JW, Snyder HM. Urologic complications of Sickle-cell disease in pediatric population. J Urol 1987, 138: 592-594.
6. Noe HN, Jerkin JW, Jerkin GR. Surgical management of priapism in children with sickle-cell anemia. J Urol 1981, 126: 770-771.