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.

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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.

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leucocytosis. Hemoglobin was 7.4 g/dl, total bilirubin 75 μmol/L, conjugated bilirubin 21 μmol/L, creatinine 54 μmol/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 urologist 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.

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