SHORT REPORT

Management of sickle cell priapism with etilefrine

A D Gbadoé, Y Atakouma, K Kusiaku, J K Assimadi

Abstract

Intracavernous injections of etilefrine were effective in seven children with acute sickle cell priapism, and stuttering priapism resolved in five children after one to seven months of oral etilefrine. Compared with our previous reports in adults, etilefrine appears to be more effective in childhood.

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Priapism is a painful vaso-occlusive complication of sickle cell anaemia (SCA). Recently, the actuarial probability of young males with SCA experiencing priapism by 20 years of age was estimated at 89%.¹ For some years, treatment of priapism has included the use of α adrenergic agonists.²⁻⁴ Most published information concerns adults. We aimed to assess the results of this treatment in 11 children with SCA consecutively treated in Lomé-Tokoin's teaching hospital.

Patients and methods

In 1996, we established a prospective management protocol for outpatient treatment of sickle cell priapism, approved by the ethics committee of the hospital. Eleven children with SCA, aged 2.5-15 years, and admitted from 1996 to 1999, were treated for acute priapism (AP, $n = 6$), stuttering priapism (SP, $n = 4$), or AP complicating SP ($n = 1$). Informed consent was obtained from parents. Treatment of AP consisted of intracavernous injections of 5 mg (0.5 ml) of undiluted etilefrine when priapism had been present for less than six hours, or intracavernous injections preceded by drainage of blood in more prolonged priapism. To prevent

relapse, we used 0.25 mg/kg twice a day of oral etilefrine for one month.

SP was treated with oral daily administration of 0.5 mg/kg etilefrine for one month. This was given in the evening to patients with nocturnal priapism; 0.25 mg/kg twice a day was given for priapism which occurred both during the day and night. These patients also self administered (or parent administered) intracavernous injections to reverse painful episodes lasting more than one hour. In those whose attacks had not ceased after one month, or who relapsed after remission, treatment was continued until one month after eventual resolution. Safety of treatment was assessed by measuring blood pressure and by direct questioning about erectile function or any other apparent side effects.

Results

The age at onset of SP varied from 11 years to 15 years (mean 13 years). The duration of an attack of SP ranged from five minutes to three hours. The duration of evolution of SP ranged from one to nine months.

Five patients with AP were given intracavernous etilefrine with a delay ranging from four to 28 hours. This procedure was successful in producing immediate detumescence in all children after a single injection. Neither hydration nor analgesia were necessary. Intracavernous injections were not needed in the five cases of SP. All 11 patients received oral etilefrine: six to prevent relapse of AP (patients 1-6) and five for the curative treatment of SP (patients 7-11, table 1). No patients treated for prevention of AP relapsed. After one month of treatment, attacks decreased in one patient with SP, while four others remitted. Three of the latter relapsed two weeks to two months later, but prolonged remission was achieved in all after one to seven months of treatment. No serious immediate or long term side effects (particularly hypertension or erectile dysfunction) were observed, either with intracavernous injections or oral etilefrine. One patient experienced agitation which ceased after dosage reduction.

Discussion

In our series, priapism attacks seemed less severe in intensity in children than in adults (for example, no need for intracavernous injections in all children, whereas eight of our 15 adults who presented with SP used intracavernous injections, personal communication). The delay to treatment of AP was relatively short (less than 28 hours) compared to the

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Table 1 Results of treatment of acute or stuttering priapism with oral etilefrine

Patient no.	Outcome	Treatment duration	Side effects	Follow up
1	No relapse	1 mth	None	1 y
2	No relapse	1 mth	Agitation	2 y
3	No relapse	1 mth	None	3 y
4	No relapse	1 mth	None	2 y
5	No relapse	1 mth	None	4 y
6	No relapse	1 mth	None	3 mth
7	Remission, relapse, cure in 4 mth	5 mth	None	4 y
8	Remission, relapse, cure in 6 mth	7 mth	None	3 y
9	Decrease in attacks, then cure in 3 mth	4 mth	None	1 y
10	Cure in 1 mth	1 mth	None	1 y
11	Remission, relapse, cure in 4 mth	5 mth	None	2.5 y

delays of longer than 30 hours observed in nine patients of 16 previously reported.³ We consider that this may explain the success in all those children who presented with AP, as intracavernous etilefrine is not usually effective beyond the thirtieth hour of priapism,³ because of necrosis of erectile tissue. These long delays in seeking medical help may be related partly to the beliefs held by many of our patients. A recent survey revealed that 31.5% of patients with SCA in Lomé, thought that priapism was caused by a spell cast over a man who had committed adultery.⁵ Thus adults with priapism often attend hospital only after previously seeking treatment from traditional healers.

Children responded better to oral etilefrine than adults. In fact, remission was obtained in almost all children after one month of treatment, and prolonged remission was obtained in all five patients after one to six months, whereas half of the adults whom we have described previously did not respond to oral etilefrine.⁶ Home management of SP is effective for almost all adult patients using such means as a cold penile bath, physical exercise, distraction, or other activities. Some adults reported these methods as effective⁶; they were not carried out

by the children, but we now advise suggesting them to children and their parents.

Preventing recurrent priapism with oral etilefrine seemed beneficial in our paediatric patients. In the series of paediatric SCA patients of Mantadakis and colleagues,⁴ where follow up oral therapy was not so structured as in our series, pseudoephedrine appeared to decrease the number of recurrent episodes. Nevertheless, we recommend multicentre, randomised, placebo controlled trials to assess the real benefit of oral α adrenergic agonists.

- 1 Mantadakis E, Cavender JD, Rogers ZR, *et al.* Prevalence of priapism in children and adolescents with sickle cell anemia. *J Pediatr Hematol Oncol* 1999;21:518–22.
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- 4 Mantadakis E, Ewalt DH, Cavender JD, *et al.* Outpatient penile aspiration and epinephrine irrigation for young patients with sickle cell anemia and prolonged priapism. *Blood* 2000;95:78–82.
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