Enuresis and Nocturia in Children with Sickle Cell Disease

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Patients with sickle cell disease (SCD) are known to excrete large quantities of urine of low specific gravity.[1] [2] They also have a much higher fluid intake on the basis of age and weight than normal controls.[3] The presence of nocturia and enuresis among these children is not, therefore, an unexpected event as has been reported recently.[4] [5]

In this paper we present our experiences with the frequency of enuresis and nocturia in children with sickle cell disease.

Materials and Methods

The parents of patients with sickle cell disease, diagnosed in the pediatric hematology clinics of Hacettepe Children's Hospital, were personally interviewed either in Ankara (by İ.B.) or in Tarsus (by T.E.). The families living outside of these two cities were sent questionnaires. A total of 26 patients whose parents responded to questionnaires (8) or attended interviews (18) were subjected to this study. Their ages varied from four to 19 years and all were of Eti-Turk extraction. Diagnosis of sickle cell anemia was confirmed by hemoglobin electrophoresis. Sickle cell preparations were made according to the standard sodium metabisulfite technique.[6] The specific gravity of random urine specimens was measured at room temperature using an urinometer.

The control group, made up of 78 children aged four to 16 years, was also interviewed in a similar manner regarding enuresis. These children were selected at random from the pediatric out-patient clinics with the exclusion of those with diseases causing polyuria (i.e. chronic renal diseases, diabetes insipidus). In this study enuresis is defined as bed-wetting occurring habitually at least once a week after four years of age and nocturia as micturition more than once per night after bedtime.

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Results

Table 1 summarizes the findings in SCD and control groups. Nine (34.6 per cent) of the 26 patients with sickle cell disease had enuresis, while only 13 (16.7 per cent) of the 78 control children manifested bed-wetting. Among the nine with enuresis, bed-wetting occurred nightly in three, two to three nights a week in another three and once a week in the remaining three. Nocturia was present in nine (34.6 per cent) who did not show enuresis. Questioning of the parents of the 26 children with SCD revealed that 25 of the mothers and/or the fathers had nocturia. Four of the fathers had a childhood history of enuresis. Again, nine siblings of the patients gave a history of bed-wetting which disappeared at a later age. There were seven children with active (current) enuresis among the brothers or sisters (over four years of age) of the nine enuretic patients with SCD. Bed-wetting was present in six siblings of the 13 children with enuresis from the control series (Table 1).

TABLE 1

INCIDENCE OF ENURESIS AMONG PATIENTS WITH SICKLE CELL DISEASE

	Sickle Cell Anemia Patients	Controls
Number Interviewed	26	78
Number of Enuretic Patients	9 (34.6 %)	13 (16.6 %)
Number of Enuretic Patients with Enuretic Siblings	7 (77.7 %)	6 (46.1 %)

Urine examination of the 26 children with SCD showed that the specific gravity was not over 1.015 in any of them. With the exception of a five-year-old boy who concentrated up to 1.014, none of the nine enuretic children had a specific gravity over 1.008.

Discussion

It is well known that carriers of hemoglobin S, either homozygous or in combination with other abnormal hemoglobins, are

unable to produce concentrated urine during periods of water deprivation. This hyposthenuria usually starts around the fourth or fifth year of age.[5] Etteldorf et al [7] found that the glomerular filtration rate is markedly increased in these patients. Inability to concentrate urine in patients with SCD has been explained by the presence of early tubular lesions causing fibrous scarring.[8] In the older patients, glomerular congestion and enlargement result in progressive ischemia and fibrosis leading to glomerular obliteration.[8] Saxena and colleagues [3] proposed that the cause of dehydration is based on primary kidney damage with obligatory water loss which leads to excessive thirst, polydipsia and polyuria.

Recently, the frequency of enuresis in children with SCD was investigated by two different centers: Suster and Oski [4] found enuresis to be 68.9 per cent among 29 patients with SCD, while their controls had an incidence of 16 per cent; the study of Noll et al [5] revealed that hyposthenuria with enuresis and/or nocturia among 21 children with SCD was 100 per cent and among 11 children with the sickle trait was 55 per cent. The frequency of similar phenomena in their 22 controls with normal hemoglobin was 18 per cent.

During this investigation, we were unable to measure the urinary specific gravity of all the family members of patients with SCD because they live in Southern Turkey; however, questionnaires revealed a high incidence of enuresis among their siblings which might be related to their being possible carriers of hemoglobin S. The frequency of nocturia was also quite striking among these patients, their parents and their siblings, again related to obligatory water loss. No attempt was made to detect emotional factors which could contribute to some extent to increased incidence of enuresis in this chronic condition. After seeing the disappearance of enuresis at a later age in nine siblings of enuretic patients, we agree completely with Noll et al [5] on its management. Extensive laboratory investigation and prolonged psychiatric therapy should be avoided except for absolutely indicated cases and both parents and patients should be given appropriate explanations. We believe that screening tests for hemoglobin S in Eti-Turk children with nocturia or enuresis should be performed.

Summary

The parents of 26 patients over four years of age with SCD were interviewed directly or through questionnaires to determine

the incidence of enuresis and/or nocturia. Of this group, 34.6 per cent had enuresis while only 16.7 per cent of the control group registered this complaint. The incidence of hyposthenuria with enuresis and/or nocturia among these 26 patients was 69.2 per cent. Their parents and siblings also presented a high incidence of nocturia and enuresis.

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