Dental fluorosis associated with hereditary diabetes insipidus

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There is little information in the medical or dental literature regarding oral findings of hereditary pituitary diabetes insipidus. This article presents a family in which the mother and her four offspring are affected by this disorder. Oral examinations revealed severe to mild fluorosis of the teeth, directly related to the stage at which hormonal therapy was introduced.

Diabetes insipidus may result from pathologic changes in any part of the close functional unit made up of the posterior lobe of the pituitary, the supraoptic and paraventricular nuclei of the hypothalamus, and the hypothalamic-hypophysial fibers. Such changes result in deficiency of the antidiuretic hormone (vasopressin), which acts directly on the distal tubules and collecting ducts to facilitate reabsorption of water. This is manifested clinically by the excretion of a large volume of dilute but otherwise normal urine and by an unrelenting thirst. The daily volume of urine may be 4 to 10 liters or more.¹

In a small percentage of instances, diabetes insipidus may occur as a hereditary disorder of which two types can be distinguished. The nephrogenic or vasopressin-resistant type occurs soon after birth.² Since polyuria is not considered an abnormal symptom in infancy, it may pass unnoticed until signs of severe dehydration are noted. Mental and physical retardation are occasionally associated with this disorder, presumably as a result of repeated episodes of severe dehydration.³ This form of diabetes insipidus appears mainly in the male members of the affected families,⁴ although several cases of affected females have also been reported.⁵ The main feature of this type of diabetes insipidus is that the concentrating defect is not affected by the administration of large doses of vasopressin. It is assumed that the end organ (that is, the distal convoluted tubules) and the collecting ducts are unresponsive to hormonal action.
Fig. 1. Mother, 35 years old. Horizontal white-brown discoloration of the complete dentition and mild enamel hypoplasia of the right upper canine.

Fig. 2. Girl R., 14 years old. A, Severe mottled enamel with brownish discoloration of the upper incisors, enamel hypoplasia, and a chalky white appearance of the lower anterior teeth. Photograph was taken during crown preparations of the upper anterior teeth. B, Radiograph showing normal radiopacity of enamel and dentin and normal supporting bone structure.

Familial pituitary diabetes insipidus is a very rare condition which also appears in early life, but it is usually less severe than the vasopressin-resistant type. Dehydration and retarded growth are uncommon. The disorder is transmitted as a sex-linked recessive or an autosomal dominant trait and affects members of both sexes. In this form of the disease, atrophy of the posterior lobe of the pituitary and hypoplasia of the paraventricular and supraoptic nuclei in the hypothalamus have been found at autopsy. In this condition the treatment of choice is the administration of vasopressin in the form of Pitressin tannate in oil. Intramuscular injections of 0.5 to 1 ml. administered every 1 to 3 days can adequately control the polyuria and polydipsia.

Oral manifestations connected with hereditary diabetes insipidus have not been reported in the medical or dental literature. There follows a report of oral findings observed in a family affected by hereditary diabetes insipidus.
Two daughters of the family were referred to the Department of Pedodontics of the Hadassah School of Dental Medicine in Jerusalem by an outpatient clinic of a Jerusalem suburb because of severe discoloration of their anterior teeth. The letter of referral mentioned that the children of this family were affected by hereditary diabetes insipidus.

Medical history revealed no consanguinity of the parents. Both parents were born and raised in Kurdistan. After their immigration to Israel in their early twenties, four children (three girls and one boy) were born and reared in the vicinity of Jerusalem. The gestation periods, deliveries, and postnatal periods of these four children were normal. Miscarriages did not occur. The father of the children had no medical record. The mother reported the onset of her diabetes insipidus in early childhood. She could not remember that anyone else in her family or her parents' family suffered from this disorder. Five years ago the mother and her four children were hospitalized at the Hadassah Medical School in Jerusalem for evaluation of their disorders. The final diagnosis was established as hereditary pituitary diabetes insipidus.

The two younger children (a 10-year-old girl and an 8-year-old boy) have received Pitressin tannate injections every 2 to 3 days since the ages of 5 and 3, respectively. They were heavy water drinkers until the onset of therapy. They now consume large amounts of water only when the influence of Pitressin injection diminishes. The mother and her two elder daughters, aged 14 and 13, who do not receive hormonal therapy, consume 10 to 15 liters of water daily and have been heavy water drinkers since early infancy. Except for this disorder, the children have been in good health and are well developed for their ages; the appearances of body height, skin, and hair were normal.

**Oral examinations of the parents and their four offspring**

The father, 37 years old, had a normal dentition that was free of discoloration or any kind of hypoplasia.

The mother, 35 years old, had a complete dentition with white-brown horizontal discoloration of all the teeth (Fig. 1). In addition, discoloration in form of white areas was seen scattered irregularly on the labial and buccal surfaces of the premolars and molars. There was hypoplastic notching of the distobuccal surface of the upper right canine, and the incisal edges of the upper incisors showed a brown discoloration. There was no attrition of the dentition, and the enamel appeared hard. There were occlusal fillings in eight molars.

Girl R., 14 years old, had a complete dentition (Fig. 2). There was a severe brown discoloration on the labial side of the upper incisors and canines, and the lower anterior...
teeth had a chalky white appearance. All of the posterior teeth had several scattered white areas. Enamel hypoplasia was present in the maxillary incisors, canines, and first molars and, to a smaller extent, in the lower incisors, canines, and molars. Apart from initial occlusal caries of the upper first molars and the distal surface of the lower left second premolar, the teeth were free of caries.

Girl E., 13 years old, had findings similar to those observed in her older sister R. (Fig. 3). The chalky white appearance of the lower incisors and the hypoplasia were less marked. Oclusal caries was present in three first permanent molars and the lower left second deciduous molar.

Girl A., 10 years old, had white speckling of all permanent teeth (Fig. 4). Mild enamel hypoplasia was present in the maxillary incisors and in the upper and lower canines and premolars. One of the lower first deciduous molars had a small white spot on the buccal surface. The teeth were free of caries.

Boy E., 8 years old, had a mixed dentition (Fig. 5). There were a few white areas in the enamel of the permanent teeth. Small white spots were observed on the buccal side of the upper and lower first and second deciduous molars. There was slight hypoplasia of the upper first permanent molars. Interproximal carious lesions were found in the left upper and lower segments.

The dentitions of all four children were free of attrition. The gingivae and mucous membranes were normal in appearance. Radiographs of the five family members revealed normal tooth development and supporting bone structure. Enamel and dentin were of normal thickness and of normal radiopacity.

**DISCUSSION**

The water in the Jerusalem suburb where the four children lived during infancy and childhood had a fluoride content of about 0.5 ppm. It has been reported that a high water intake in warmer climates can produce moderate dental fluorosis, even though the water has a small fluoride concentration.
Different investigations showed an average daily water intake of 926 ml. (water drunk plus amount added to food) in healthy children 3 to 5 years old. However, the fluoride content of this water consumption was not mentioned. Since a relatively rapid increase in the average fluoride concentration of the enamel is found during the early years of tooth development, it seems that the affected family members showed, in different degrees, a propensity for retention and fixing of fluoride in calcified tooth tissues. The high water consumption with a low fluoride content during the early years of tooth development seems to have resulted in an accumulation of the fluoride concentration in the tooth substance. It is thought that in the two older girls the daily low-fluoride water consumption 3 to 4 liters or more the above-mentioned amount over a prolonged period during the early development of their dentition has resulted in severe fluorosis of the teeth.

In Girl A, who received hormonal therapy from age 5, the fluorosis is less severe, and it is mildest in the youngest child, who was treated from the age of 3. The mother also had fluorosis of the teeth. The fluoride content of the water she drank during her childhood in Kurdistan is unknown.

Staining due to tetracyclines produces a completely different clinical picture. The medical history of the four children did not reveal any tetracycline therapy. The brown discoloration of teeth sometimes seen in amelogenesis imperfecta is not limited to the incisal halves of the upper anterior teeth. The normal thickness and radiopacity of the enamel are also contrary to usual findings in this condition.

CONCLUSION

Extremely heavy intake of water with a fluoride content of about 0.5 ppm during the early years of tooth development has produced severe to mild fluorosis of the teeth in the family members affected by hereditary pituitary diabetes insipidus.
SUMMARY

Among the characteristics of hereditary pituitary diabetes insipidus are polydipsia and polyuria from early infancy.

Drinking of large amounts of water, even with lower than accepted fluoride content, can produce fluorosis of the teeth. A mother and her four children affected by this disorder presented different degrees of fluorosis directly related to the stage at which hormonal therapy was introduced.

REFERENCES


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