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Correlation between Magnetic Resonance Imaging of Posterior Pituitary and Neurohypophyseal Function in Children with Diabetes Insipidus

MOHAMAD MAGHNIE, ANDREA VILLA, MAURIZIO ARICO, DANIELA LARIZZA, STEFANO PEZZOTTA, GIAMPIERO BELUFFI, EUGENIO GENOVESE, AND FRANCESCA SEVERI

Departments of Pediatrics, Radiology (A.V., E.G.), Neurosurgery (S.P.), and Pediatric Radiology (G.B.), University of Pavia, Istituto di Ricovero e Cura a Gerattere Scientifico Popiclinico S. Matteo, Pavia, Italy

ABSTRACT. The posterior pituitary lobe and stalk were studied by magnetic resonance imaging in 20 children with diabetes insipidus of different origins: primary familial autosomal dominant (n = 2) or idiopathic (n = 2), and secondary to craniopharyngioma (n = 6, resected in 5), to Langerhans cell histiocytosis (n = 5), to excessive water intake (dipsogenic; n = 3), to renal vasopressin insensitivity (n = 1), and to osmoreceptor dysfunction (n = 1).

Of the four children with primary diabetes insipidus, the posterior bright signal was recognizable in two with the familial autosomal dominant form and one with the idiopathic form; in the latter, the pituitary stalk was thin, while it was normal in the first two patients; no posterior hyperintense signal with enlarged and gadolinium-enhanced pituitary stalk was observed in the fourth. The posterior hyperintense signal was absent without evidence of ectopic posterior pituitary tissue regeneration in all five children with surgically removed craniopharyngioma and was doubtful in the child with unresected craniopharyngioma; the stalk was unrecognizable in all patients. In the five children with Langherans cell histiocytosis, the posterior bright signal was absent, while the stalk was normal in two and unexpectedly enlarged in three (uniformly in two and mainly at the level of median eminence and hypothalamus in one). All five patients with dipsogenic or nephrogenic diabetes insipidus or osmoreceptor dysfunction had normal images of posterior pituitary lobe and stalk. Normal posterior pituitary bright signal and stalk were found in all 25 healthy control children.

Plasma vasopressin was undetectable in all patients except in nephrogenic one, in the child with osmoreceptor dysfunction, and in two of three dipsogenic children, the third mimicking partial neurogenic diabetes insipidus.

We conclude that 1) the absence of a magnetic resonance posterior pituitary signal in patients with central diabetes insipidus is always associated with hypothalamic-neurohypophyseal axis lesion; 2) the absence of the posterior pituitary bright signal correlates closely with undetectable plasma vasopressin only in the presence of organic hypothalamic-neurohypophyseal tract lesion; 3) evidence of posterior pituitary hyperintensity in diabetes insipidus patients does not necessarily indicate that functional integrity of the hypothalamic-neurohypophyseal axis is preserved; 4) the release of stored vasopressin may be impaired in some cases of autosomal dominant diabetes insipidus as well as in some idiopathic forms; and 5) evidence of isolated enlarged stalk in children with acute onset of diabetes insipidus suggests that magnetic resonance may disclose a preclinical-oligosymptomatic phase of systemic disorders (Langherans cell histiocytosis dependent?) affecting the hypothalamic-neurohypophyseal tract. This could help to clarify both the natural history of anatomical and functional alteration during the course of diabetes insipidus and the origin of some idiopathic forms. Clear definition of isolated stalk alteration as a precocious manifestation of Langherans cell histiocytosis could lead to early specific treatment in such patients. (*J Clin Endocrinol Metab* **74:** 795–800, 1992)

CENTRAL neurogenic diabetes insipidus (DI) is a disorder characterized by chronic polyuria and polydipsia, corrected by vasopressin administration. It may be primary (idiopathic, sporadic, or familial) or, more often, acquired after injury to the hypothalamic-neurohypophyseal system (1, 2). Acquired DI is most frequently due to surgical removal of tumor in the hypothalamic-pituitary area. Organic lesion in this region may also be due to inflammatory diseases, such as Langherans cell histiocytosis (LCH), sarcoidosis, tuberculosis, or autoimmune neurohypophysitis (1–6). Polydipsic conditions mimicking vasopressin (AVP) deficiency, such as dipsogenic DI and nephrogenic DI or the osmoreceptor dysfunction disease, are far less frequent (1, 3). The standard reliable way to diagnose central DI has been based on the AVP response to water deprivation, followed by administration of 1-desamino-8-D-arginine vasopressin (DDAVP), as well as by assessment of the renal response to hypertonic saline infusion.

Radiological computed tomography (CT) scan findings of neurohypophyseal system damage due to large lesions or, far less often, to small tumors of the pituitary stalk,

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Address all correspondence and requests for reprints to: Dr. M. Maghnie, Department of Pediatrics, University of Pavia, IRCCS Policlinico S. Matteo, I-27100 Pavia, Italy.