Neonatal cholestasis and hypopituitarism

Sir,

We read with interest the paper by Kaufman *et al*¹ on neonatal cholestasis and hypopituitarism. We have been treating a similar patient whose problems were complicated by diabetes insipidus of pituitary origin. She was referred from another hospital at the age of 1 month for assessment of secondary hypothyroidism, cholestatic jaundice, temperature instability, and microcephaly. She was born at 41 weeks' gestation after a spontaneous, normal delivery. Her mother was aged 31 years and had two other healthy children.

Though the initial serum sodium concentration was normal, by the age of 2 weeks she became hypernatraemic. When transferred to our hospital she was drowsy, hypotonic, and icteric. She had microcephaly and roving eye movements, and visually evoked responses were absent.

In view of her secondary hypothyroidism, a full assessment of anterior pituitary function was performed: this indicated panhypopituitarism. At an early stage she had been noted to be passing large quantities of dilute urine and on the basis of this and the hypernatraemia, a diagnosis of diabetes insipidus was made. Treatment with intramuscular vasopressin was begun. The serum sodium value fell and the urine became more concentrated and of smaller volume. She was extremely sensitive, however, to small alterations in the dose of vasopressin. Replacement treatment with growth hormone, hydrocortisone, and thyroxine was started. No intracranial lesion was identified on computed tomogram.

The jaundice was cholestatic in nature (raised transaminases and alkaline phosphatase activities). No viral or metabolic cause could be identified and liver biopsy showed a giant cell hepatitis. After one month's treatment with hydrocortisone and growth hormone her jaundice resolved, but her transaminases and alkaline phosphatase activities took seven months to return to normal values.

Subsequent progress was poor. There was little head growth, developmental progress was profoundly retarded, and the diabetes insipidus proved difficult to control. She died at the age of 11 months. Necropsy examination of the pituitary gland showed a noticeably gliotic posterior portion. The anterior pituitary consisted of morphologically normal cells (the stalk was extremely small, suggesting a functional separation of the pituitary from the hypothalamus).

We feel our patient is of particular interest because of the associated diabetes insipidus. This suggests that the primary pathological process in this condition is hypothalamic. Unfortunately, as in the patient of Kaufman, no causative agent was identifiable.

Reference

- ¹ Kaufman FR, Costin G, Thomas DW, Sinatra FR, Roe TF, Neustein HB. Neonatal cholestasis and hypopituitarism. *Arch Dis Child* 1984;**59**:787-9.

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This correspondence is now closed, Ed.