RESEARCH LETTER

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Liver failure in a neonate with congenital adrenal hyporesponsiveness

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The association between hypopituitarism and neonatal hepatitis is well described [1], however, hypopituitarism leading to acute liver failure has not yet been reported. Our patient, a 5-week-old infant presented with acute liver failure. She was delivered by emergency caesarean section at 31 + 3 weeks because of intrauterine growth retardation and fetal distress with a birth weight of 975 g. At 4 weeks of age, she was noted to be jaundiced with prolonged INR (international normalised ratio), unresponsive to vitamin K. Detailed investigations for causes of neonatal liver failure including neonatal haemochromatosis were normal except for a low random cortisol level of 77 nmol/l. Glucocorticoid deficiency was confirmed with an abnormal short synacthen test (cortisol levels at 0 min, 30 min and 60 min were 56, 122 and 172 nmol/l respectively after synacthen injection). There was no biochemical evidence of mineralocorticoid deficiency. Basal thyroid function and a thyrotrophin (TRH) stimulation test were normal. Urinary steroid profile showed no evidence of a steroid biosynthetic defect. MRI scan of the brain was normal. No adrenal abnormality was seen on ultrasound scan and X-ray film. Replacement treatment with oral hydrocortisone (2.5 mg thrice daily) was commenced on day 43. INR gradually improved and was normal (1.07) by day 55 with a concomitant improvement in liver function tests (Table 1).

At 6 months follow-up she was anicteric with a 4 cm palpable liver. Serum transaminase levels were normal. Liver biopsy (at 6 months) showed mild portal tract

Table 1 Serial INR, AST and bilirubin before and after cortisol replacement

	INR	AST U/l	Bilirubin (µmol/l)
Admission	2.4	100	226 (192 conjugated)
Before cortisol replacement	2.6–3.3	73–113	178–224
8 days after cortisol replacement	1.31	101	112
1 month after cortisol replacement	1.07	115	58
Six months follow-up	0.94	47	3

fibrosis with focal porto-portal bridging fibrosis. In our case, isolated cortisol insufficiency was responsible for liver failure as suggested by the improvement after introduction of replacement therapy with hydrocortisone. This highlights the need to consider cortisol deficiency as a differential diagnosis in neonatal liver failure.

Reference

1. Spray CH, Mckiernan P, Waldron KE, Shaw N, Kirk J, Kelly D (2000) Investigation and outcome of neonatal hepatitis in infants with hypopituitarism. *Acta Paediatr* 89: 951–954

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