Preterm infants with athetoid cerebral palsy: kernicterus?

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Patient 1 was a 2078 g boy delivered in a regional hospital at 33 weeks gestation. His postnatal course was uncomplicated. His total bilirubin level was 13.1 mg/dl on the third day of life. Thereafter, measurement of total bilirubin was discontinued. Phototherapy was not performed. He was admitted to hospital at 48 days of age because of icterus. His total bilirubin level was 19.7 mg/dl. The hyperbilirubinaemia was rapidly improved with phototherapy.

Discussion
Neurological, neurophysiological, and neuroimaging features of these two patients are compatible with athetoid cerebral palsy due to chronic bilirubin encephalopathy despite the lack of clinical signs or symptoms of kernicterus during the neonatal period. Previous studies have shown that about 15% of patients with proven kernicterus fail to exhibit any definite neurological signs. The absence of signs of acute bilirubin encephalopathy does not exclude the possibility of athetoid cerebral palsy due to bilirubin. It is noteworthy that severe hyperbilirubinaemia was not present in patient 2. Previous studies have shown the possibility of chronic bilirubin encephalopathy without severe hyperbilirubinaemia. MRI may be useful for the assessment of chronic bilirubin encephalopathy. Both of our patients had characteristic abnormal high intensity areas in bilateral globi pallidi on T2 weighted images, although these are not specific to kernicterus. Brainstem auditory evoked response is also useful for the assessment of bilirubin encephalopathy.

Magnetic resonance imaging (MRI) of both patients showed abnormal high intensity areas in the bilateral globi pallidi (fig 1). Although the brainstem auditory evoked response showed elevated thresholds and abnormal interwave separation, the children could understand simple verbal directions.

Figure 1  Magnetic resonance imaging findings. (A) T2 weighted image of patient 1 at 5 months of corrected age. Abnormal high intensity areas were seen in the bilateral globi pallidi. (B) T2 weighted image of patient 2 at 9 months of corrected age. Abnormal high intensity areas were seen in the bilateral globi pallidi. (C) Magnification of basal ganglia. Arrows indicate abnormal high intensity areas in the globi pallidi.