LETTERS TO THE EDITOR

Crigler-Najjar syndrome type I: management with a phototherapy crib mattress

EDITOR,—Crigler-Najjar syndrome type I is a rare autosomal recessive disorder characterised by absence of uridine diphosphate glucuronic acid transferase activity. Affected newborns develop severe unconjugated hyperbilirubinaemia and are at risk of death or permanent neurological damage from kernicterus. Phototherapy is the basis of treatment until the child grows to a more favourable size for liver transplantation, the only cure presently available.1

A girl, born at term to non-consanguinous Italian parents, presented at 8 days of age with a total bilirubin concentration of 419 μmol/l and no conjugated bilirubin. Investigations were normal including complete blood count, Coombs test, liver function tests, urine and blood cultures. Family history was negative for jaundice. After 4 days of phototherapy the baby was discharged and then readmitted two days later because of unconjugated hyperbilirubinaemia. Thyroid function tests, pyruvate kinase, and glucose-6-phosphate dehydrogenase activities were normal. A percutaneous liver biopsy specimen showed mild non-specific changes including steatosis of some hepatocytes and canicularial cholestasis. A diagnosis of Crigler-Najjar type I was based on the analysis of bile obtained by duodenal hyperbilirubinemia and absence of unconjugated bilirubin only (Dr Roy Chowdhury, personal communication).

The baby was treated with phenobarbitone (5 mg daily) and with single and double banks of phototherapy lights (Sylvania F20T12/CW cool white and Philips TL20W/037 special blue) and discharged with a portable Omehda ‘Bili Blanket’ at 7 weeks of age. The bilirubin concentration increased by approximately 5 μmol/l per day so additional phototherapy using two banks of bililight was given twice a week for nine hours each in a medical daycare unit. She was readmitted on three occasions for continuous double photothera-py when her bilirubin concentration rose above 250 μmol/l, associated with viral illnesses or vaccination. The phenobarbitone did not induce bilirubin conjugation.

A fan cooled phototherapy lamp was designed to replace a standard crib mattress (130 cm × 70 cm) (figure). The ‘treatment’ surface consisted of a 9.5 mm thick transparent acrylic lid. The lamp held ten 120 cm fluorescent tubes (Philips FT40T12/BB). The five ballasts (Philips Mark III, R2540), TPC, sound rating A) to power the lamps were housed in a separate fan cooled enclosure, all necessary interconnections being made by a multicore cable and connector. Radiation levels at 45 cm, measured at the treatment surface exceeded 25 μW/cm² (Bio-tek phototherapy radiometer, model 74345). A transparent layer of plastic air bubble sheeting (Astro Polyfoam, Toronto) was placed in line on the treatment surface with a white crib sheet, lowering the light intensity to approximately 15 μW/cm²/μmol and reducing the glare from the lamps to a more comfortable level. The baby slept with her head on a small pillow and was covered with a standard cotton sheet and blanket.

The baby was treated initially with eight hours of phototherapy at night using standard ‘daylight’ fluorescent lamps but had significant skin tanning over a three week period. After the baby was discharged with the Philips F40T12/BB lamps the bilirubin concentration decreased and was maintained around 200–250 μmol/l with eight hours of nightly phototherapy. This allowed more occasions for blood transfusions while the admissions were required and the baby’s development was appropriate at 16 months of age.

Side effects associated with phototherapy include headache, nausea, vertigo, painful sensations in the eyes and photophobia,2 and photosensitive changes in the skin.3 The eyes were covered during treatment with a black eye shield and later protected by her pillow. Ophthalmological examination was normal at 15 months of age.

There has been one other report of a phototherapy bed used to treat a 10 year old girl who remained well without neurological sequelae until lost to follow up at age 17 (J M Littlewood, personal communication).4

As the child gets older a new phototherapy mattress with more lights will be made to fit a standard twin bed frame. This way we hope to control the hyperbilirubinemia for as long as possible with the development of new phototherapy techniques such as gene transfer5 thus avoiding the need for liver transplantation.

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