

received by the hospital, or had not been taken within the recommended time of 6 to 14 days. Of 1000 sequential samples received from the Glasgow area during 4 weeks in 1977, in 4.2% of cases the child was 6 days old when the specimen was assayed, and must therefore have been less than 6 days old when the sample was taken. 93.8% of samples were taken at an 'acceptable' age of 7 to 21 days old; but in the remaining 2% of cases, the child was over 3 weeks old before the test was done; in 0.8% of cases, representing 100 samples per year in Glasgow, or 700 in Scotland, the child was more than 2 months—in some cases, as much as 7 months old.

Obviously in these cases the value of the test is much reduced, with the possibility that permanent brain damage would already have occurred before it was performed. This has two consequences—firstly, doctors who are working with sick young children should be aware that they may not have had inborn metabolic errors excluded by the National Screening Programme; secondly, if neonatal screening is to be extended to other conditions of personal or economic importance, such as hypothyroidism or lead poisoning, a truly fail-safe and universal screening programme would be needed, if it is to justify its cost.

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## Early onset of homozygous $\beta^0$ thalassaemia associated with neonatal jaundice

Sir,

I read with interest the paper by Furbetta *et al.* on this subject (*Archives*, 1978, 53, 250). Although they stated that 'all other common causes of neonatal jaundice had been excluded', the ABO set-up (blood group of baby A and the mother O) which is the most common cause of jaundice in the newborn, was present. In ABO incompatibility, the direct Coombs's test is usually negative. Therefore this possibility should be ruled out before ascribing the neonatal jaundice to thalassaemia.

I should also like to add that among our 90 patients with thalassaemia major (until 1970) pallor and distended abdomen were noticed at birth in two, at 17 days of age in one, at 20 days in one, at 30 days in two, and at about 40 days in two; the observation of splenomegaly as early as

17 days of age in one, might indicate that Cooley's anaemia in this area becomes symptomatic very early as stressed by the authors.

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Professor Cao comments:

The letter from Prof. S. Ozsoylu raises the possibility that the neonatal jaundice could have been due to ABO incompatibility.

However there was evidence of a moderate haemolytic process with Hb level of 13 g/dl in the 5th day, and the jaundice was first noted on the 3rd day, while in ABO incompatibility there is usually little or no evidence of a haemolytic process, and hyperbilirubinaemia appears usually within 24 hours of birth. Moreover, microspherocytosis, a prominent feature of ABO haemolytic disease, was absent.

The direct Coombs's test on the infant's erythrocytes was negative, while in our laboratory, as in others, a weakly positive reaction in ABO haemolytic disease is common. Finally when there is ABO incompatibility it is our custom to test the eluate from the erythrocytes of the affected newborn with A (or B) adult cell. This reaction was negative, and not strongly positive as found usually in ABO haemolytic disease. ABO haemolytic disease cannot be ruled out with certainty in this case but the evidence makes this diagnosis highly improbable.

Prof. Ozsoylu comments that in the population under his care Cooley's anaemia often presented early. In the last year we diagnosed 80 new cases of Cooley's anaemia due to homozygous  $\beta^0$ -thalassaemia. An early presentation with criteria such as symptomatic anaemia (Hb < 7–8 g/dl) at 75–90 days was observed in 30% of cases, but the severe anaemia (Hb < 6 g/dl) at 60 days as seen in our patient was rather uncommon. Splenomegaly in cases with earlier presentation is an unusual feature in this area. The age and clinical findings at presentation in Cooley's anaemia varies in different ethnic groups. This may be due to the genetic heterogeneity of the disease and different environmental conditions.

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## Successful restoration of immunity in the DiGeorge syndrome with fetal thymic epithelial transplant

Sir,

I read with interest the paper by Thong *et al.* on this subject (*Archives*, 1978, 53, 580). The authors stated that correction of the hypocalcaemia was achieved by the use of