

milk preparations are inadequately diluted, will tend to produce hypernatraemia and uraemia. The child may be able to compensate for a time but the extra fluid losses that will ensue if vomiting and an osmotic diuresis occur, or if fluid losses exceed intake as in increased sweating during a fever or bout of hot weather, will rapidly lead to hypernatraemic dehydration.

The importance in recognizing this condition in distinction to the commoner isonatremic dehydration is the greater mortality and morbidity that accompanies it. Of children so affected, 8% die and 8% have residual brain damage (Macaulay and Watson, 1967). Morris-Jones, Houston, and Evans (1967) have shown that 71% of cases with plasma  $\text{Na}^+$  greater than 158 mEq/l. have convulsions often during the period of recovery. 87% have convulsions if, in addition, blood urea level exceeds 90 mg/100 ml. These may contribute to permanent brain damage and are less likely to occur if plasma osmolality is reduced slowly and rehydration is gradual.

The mother of our patient found that she could only satisfy the child's 'hunger' by greatly increasing the amount of milk powder per feed. The milk that she made up contained 86 mEq/l. sodium, osmolality 1103 mOsm/kg, the normal values being respectively 26 mEq/l. and 315 mOsm/kg. Such a high solute load will stimulate thirst but if the demands of the child are met with further highly concentrated feeds, more solute than fluid is received and a vicious cycle is set up. Hot weather may have been an important contributory factor in our patient.

It would perhaps be wise for the manufacturers' instructions on tins of powdered milk to contain a warning about the dangers of exceeding the recommended concentration.

### Summary

A 9-month-old infant, admitted to hospital in coma due to hyperosmolar dehydration, was slowly rehydrated over a period of 10 days. Neurological recovery, though slow and interrupted in the second week by the transient appearance of choreoathetoid movements, was complete after 5 weeks.

A regimen of gradual rehydration and slow reduction in plasma osmolality may minimize the considerable risk of fits and permanent brain damage attendant upon this condition.

This child's condition apparently resulted from the injudicious use of highly concentrated milk feeds during a bout of hot weather when increased insensible fluid losses were not replaced.

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## Congenital Hypothyroidism and Neonatal Jaundice

The association between congenital hypothyroidism and jaundice in the neonatal period has been recognized for some years (Åkerrén, 1954; Christensen, 1956) and is mentioned in reviews of neonatal jaundice and in textbooks. None the less it is the authors' experience that few paediatricians think of this diagnosis unless other obvious features of hypothyroidism are present in a jaundiced baby.

This paper reports 12 babies with significant hyperbilirubinaemia which was apparently caused by hypothyroidism. 4 of these babies were referred to one author for diagnosis of neonatal jaundice, 6 additional cases were found by searching the case notes of 56 babies and children with hypothyroidism seen at the Royal Children's Hospital, Melbourne, in the period 1960-70, and 2 were diagnosed by another paediatrician in Melbourne.

Only 18 of the 56 case histories reviewed contained a statement regarding neonatal jaundice. Neonatal jaundice had been the presenting feature in 4 babies (including 3 of those diagnosed by the

TABLE  
 Details of 12 Babies with Jaundice and Hypothyroidism

Case No.	Sex	Gestation (wk)	Birth-weight (g)	Major Presenting Symptoms	Other Clinical Features	Age at Presentation	Highest Serum Bilirubin Recorded (mg/100 ml)	Duration of Jaundice	Age at Diagnosis	Serum Protein-bound Iodine ( $\mu\text{g}/100\text{ ml}$ )
1	F	41	3400	Severe jaundice	Lethargy; constipation	9 dy	20	12 dy	10 dy	0.7
2	M	41	4420	Severe jaundice	Suggestive faeces	6 dy	17	3½ wk	3 wk	2.4
3	M	37	3060	Severe jaundice	None	4 dy	22	3 wk	4 wk	2.2
4	F	42	3550	Prolonged jaundice	None	5½ wk	10	6 wk	6 wk	1.4
5	F	40	3660	Prolonged jaundice	Umbilical hernia	4 wk	18	4 wk	4 wk	<1.0
6	M	40	3800	Prolonged jaundice	Typical cretin	7 wk	NK†	7 wk	7 wk	1.2
7	F	40	4825	Typical cretin	History of prolonged jaundice	7 wk	NK	3 wk	7 wk	1.6
8	M	40	4940	Hypothermia; fatal bronchopneumonia; typical cretin	History of prolonged jaundice	9 wk	NK	7 wk	9 wk	1.7
9	F	40	3520	Typical cretin	History of prolonged jaundice	3 mth	NK	5 wk	3 mth	0.8
10	M	40	3190	Goitre*; lethargy; pallor	History of prolonged jaundice	2 wk	NK	3 wk	3 mth	0.6
11	F	40	3940	Typical cretin; mental retardation	History of prolonged jaundice	10 mth	NK	5 wk	11 mth	1.3
12	F	40	4090	Typical cretin	History of prolonged jaundice	12 mth	NK	3 wk	12 mth	0.8

\*Goitre was present at birth. Thyroxine was given without investigation and delay in final diagnosis resulted.  
 †NK = serum bilirubin not measured.

authors), and a history of prolonged neonatal jaundice was recorded in 5 other patients. Jaundice was insignificant, or had an adequate explanation, in the remaining 9 babies.

Relevant facts regarding these 12 children are given in the Table. In those babies (Cases 1-6) whose presenting problem was jaundice, the serum bilirubin was indirect reacting. Maternofetal blood group incompatibility was excluded by blood grouping of mother and baby, by direct Coombs test, and by testing of maternal serum against a range of blood group antigens. Blood cultures were negative in all except Case 1 whose culture yielded *Staph. albus*. Viral studies, and tests for toxoplasmosis and syphilis, were performed in several cases.

No relevant tests had been performed in those children (Cases 7-12) who presented at a later age with a history of prolonged jaundice.

### Comments

In Cases 1, 2, and 3, the major presenting feature was jaundice of excessive severity early in the neonatal period. Other features of hypothyroidism were present in Cases 1 and 2, but these were lacking in Case 3 and numerous investigations were performed before the serum protein-bound iodine was measured to exclude the diagnosis of hypothyroidism.

Cases 4 and 5 illustrate presentation with abnormally prolonged neonatal jaundice. Again, the general well-being and activity of these babies led to a period of delay before the correct diagnosis was established.

In Cases 6 to 12, flagrant features of hypothyroidism were present at the time of presentation, and the diagnosis was then rapidly established. However, prolonged neonatal jaundice had been present in each baby and the diagnosis could have been made much earlier if more attention had been paid to this feature. Diagnosis was delayed up to 12 months (Case 12). The danger of such a delay is emphasized by Case 8. This baby was diagnosed only when he presented moribund with bronchopneumonia and hypothermia. His death might have been avoided had the earlier symptoms of jaundice and lethargy been interpreted correctly.

These 12 cases are presented to indicate that significant neonatal hyperbilirubinaemia or significantly prolonged neonatal jaundice are important manifestations of congenital hypothyroidism. The incidence of neonatal jaundice due to hypothyroidism cannot be measured in this type of study, but it is clear that it is sufficiently frequent to warrant consideration of hypothyroidism when the indirect reacting bilirubin level is raised without an adequate cause, and especially when this hyperbilirubinaemia persists. Though early hyperbilirubinaemia as a

result of maternofetal blood group incompatibility is relatively common, persistent hyperbilirubinaemia is much less frequent. Because the other features of hypothyroidism may be minimal at this age, it would be ideal to perform thyroid function studies in all babies in whom unexplained hyperbilirubinaemia persists more than 2 or 3 weeks. These tests should certainly be done whenever slight inactivity, slowness with feeding, or constipation is present in addition to jaundice.

### Summary

Twelve patients are described in whom hypothyroidism was associated with significant neonatal jaundice which persisted for as long as 7 weeks in some babies. The fact that these babies were seen at one paediatric centre during an 11-year period indicates that this association is not uncommon. Recognition that hyperbilirubinaemia can be the only obvious symptom of hypothyroidism in the neonatal period is diagnostically important.

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## Infantile Cortical Hyperostosis with Lytic Lesions in the Skull

A case of infantile cortical hyperostosis is described in which lytic changes in the skull vault were a prominent feature and which posed a diagnostic problem.

### Case History

A female child, the second child of healthy unrelated parents, was born at term, birthweight 3617 g. For a chest infection in the neonatal period antibiotics were given. After a period of good progress she fell ill at the age of 4 weeks with a cold followed by bronchitis, for

which she was given a course of antibiotics at home. One week later swelling developed around the left eye and a few days later spread to both periorbital regions. Aged 9 weeks, she was seen in hospital because the swelling had persisted. No other abnormality was noted. Skull *x*-rays at this stage were unremarkable, but Hb was only 7.9 g/100 ml and there was a leucocytosis (white blood cells 21,700/mm<sup>3</sup>). Despite this anaemia and a further cold she remained well but feeding became a little slow.

At the age of 3 months because of the persisting periorbital swelling she was transferred to this hospital with a diagnosis of possible neuroblastoma. She was a pale baby on the 75th centile for weight. Both upper eyelids were swollen but there was no proptosis and the fundi were normal. There was no general oedema and no clinical evidence of cardiovascular or renal disease.

Investigations confirmed her anaemia, Hb 7.3 g/100 ml with anisocytosis, poikilocytosis, and a low serum iron of 21 µg/100 ml. Reticulocytes were 2.8%. Hb electrophoresis was normal. WBC 11,800/mm<sup>3</sup> with a normal differential. The platelet count was increased at 955,000/mm<sup>3</sup> ESR 80 mm/1 hour. Bone-marrow examination showed a slight shift to the left with morphologically healthy megakaryocytes. No blood loss was detected in the stools. The alkaline phosphatase was 25 KA units/100 ml. 24-hour urinary VMA was 0.25 mg. Other biochemical investigations were normal. Skull *x*-rays (Fig. 1) showed patchy lytic lesions in the frontal bones bilaterally and early hyperostosis of the mandible. An IVP showed no evidence of a neuroblastoma and the rest of the skeletal system appeared normal.

Shortly after admission her anaemia was corrected by transfusion since when she has remained clinically well, without further anaemia, and gained weight satisfactorily. No further therapy has been given.

Her face became progressively more deformed, and between 4 and 6 months of age she developed a heavy jowled appearance with squaring of the lower face. At the age of 7 months further skull *x*-rays (Fig. 2) showed a mandible which had the characteristic features of infantile cortical hyperostosis but the lytic lesions previously seen in the frontal bone had disappeared. At this time the ESR had fallen to 10 mm per hour, the platelets were normal, and Hb was being maintained at a normal level. When seen at 10 months of age there had been some clinical improvement in the appearance of the lower face.

### Discussion

The present case is of particular interest both because of the radiological features and because of the light it may throw on the pathogenesis of Caffey's disease. Radiologically evident bone erosions are not a well-known feature of the condition and seem to have been described previously only recently by Neuhauser (1970). In the absence of other osseous lesions, differentiation of the skull