Case reports

Congenital rickets with maternal pre-eclampsia[1]

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Congenital rickets has been described in babies born to mothers with vitamin D deficiency. An unusual case of a very small-for-dates infant born to a mother with normal vitamin D status and severe pre-eclampsia is reported. The baby, who showed evidence of rickets from the first day of life, subsequently developed late-onset rachitic respiratory distress and died.

Case report

A female baby, weighing 0.684 kg at 29 weeks gestational age by certain dates, was delivered by lower segment caesarian section for intrauterine growth retardation and severe pre-eclampsia. It was the fourth pregnancy of her mother, a 28-year-old Caucasian. There had been severe pre-eclampsia in her first pregnancy which resulted in a 2 kg male baby at 36 weeks gestation. The second and third pregnancies ended in miscarriages. During and before this pregnancy the mother had a normal diet and no symptoms suggestive of malabsorption, and four weeks after delivery her plasma calcium, inorganic phosphate and alkaline phosphatase were normal. Her plasma 25-hydroxycholecalciferol level was 6.8 ng/ml (normal adult range 3–30 ng/ml).

The baby was well at birth and had no evidence of hyaline membrane disease. She was fed both maternal and banked expressed breast milk through a nasogastric tube from the second day with a maximum volume of 200 ml/kg/day. At two weeks of age she developed respiratory distress which quickly responded to oxygen therapy in a head-box. Her chest X-ray showed bilateral patchy collapse and consolidation. Retrospective review of her X-rays (days 1 and 8) showed generalized poor skeletal mineralization, and fraying of the metaphyses of the lower end of the radius and ulna (Figure 1). Plasma calcium was 2.45 mmol/l, alkaline phosphatase 595 U/l, more than seven times the upper limit of normal for adults in our laboratory (21–84 U/l) (Kovar *et al.* 1982); a full infection screen was negative. A diagnosis of rachitic respiratory distress was made and she was started on 0.1 μg of 1 α-hydroxycholecalciferol per day orally. She remained well in 25–30% oxygen. She had to be ventilated during the third week of life for deteriorating arterial blood gases and evidence of collapse and consolidation on her chest X-ray. She required only low peak pressure and rate and came off the ventilator at the end of the fourth week. Feeding was resumed through a nasojejunal tube.

Four days later she developed abdominal distension with increased gastric aspirates. Necrotizing enterocolitis was suspected and she was transferred to a regional unit for intravenous alimentation. There she was fed intravenously and gained weight slowly. She was ventilated soon after admission for increasing respiratory distress. Attempts at weaning her off the ventilator failed. Alkaline phosphatase levels dropped by the end of the seventh week, with normal plasma calcium and low inorganic phosphate in spite of phosphate supplements and increasing doses of 1 α-hydroxycholecalciferol. Phosphate excretion studies showed maximum renal tubular reabsorption of phosphate; parathormone level was 0.39 μg/l (normal range 0.1–0.73 μg/l). Her chest X-rays showed evidence

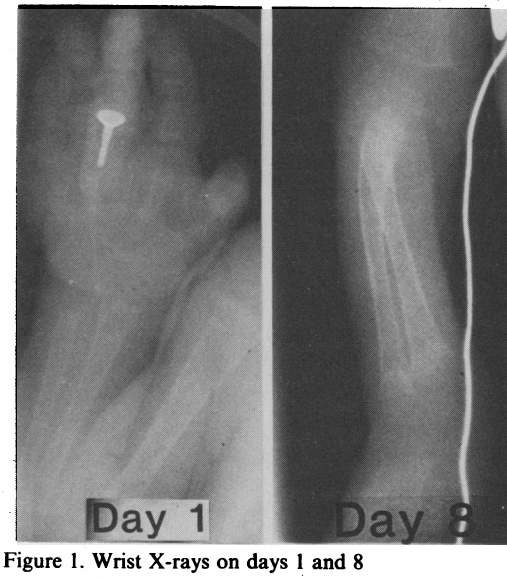

Figure 1. Wrist X-rays on days 1 and 8

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of bronchopulmonary dysplasia. She died of respiratory failure at the age of 65 days weighing 1.26 kg. The post-mortem report confirmed the clinical diagnosis. Retrospective analysis of plasma samples showed a 25-hydroxy-cholecalciferol level on day 2 of 9.0 ng/ml; plasma inorganic phosphate, calcium and alkaline phosphatase on days 2, 5 and 10 and subsequent biochemical data are shown in Figure 2.

Discussion

This baby was extremely small-for-dates and had no respiratory distress at birth. Rickets was diagnosed at the beginning of the third week with the onset of respiratory distress. Radiological changes were present from the first day of life on retrospective review. Begum *et al.* (1968), Moncrieff & Fadahunsi (1974) and Sann *et al.* (1977) reported congenital rickets in four babies due to either undiagnosed maternal mal-absorption or deficient intake of vitamin D by the mother.

In rickets impaired respiratory movement is brought about by softening and fracturing of the ribs as well as weakness of the respiratory muscles. This type of respiratory distress was reported by Glasgow & Thomas (1977) and Bosley *et al.* (1980) in 10 preterm babies. The lightest baby weighed 0.71 kg at 31 weeks gestation, while the earliest age of onset of respiratory distress was five weeks postnatally. The mothers of 5 of the babies had severe pre-eclampsia. One baby died.

This baby's normal plasma 25-hydroxy-cholecalciferol on day two and the normal maternal plasma calcium, inorganic phosphate, alkaline phosphatase and 25-hydroxy-cholecalciferol postnatally make maternal vitamin D deficiency an extremely unlikely cause of rickets in this case (Hillman & Haddad 1974). Khattab & Forfar (1971) showed that the transfer of calcium and phosphate across the placenta is impaired in conditions associated with placental insufficiency. It is suggested that severe maternal pre-eclampsia was responsible for the development of congenital rickets in this case.

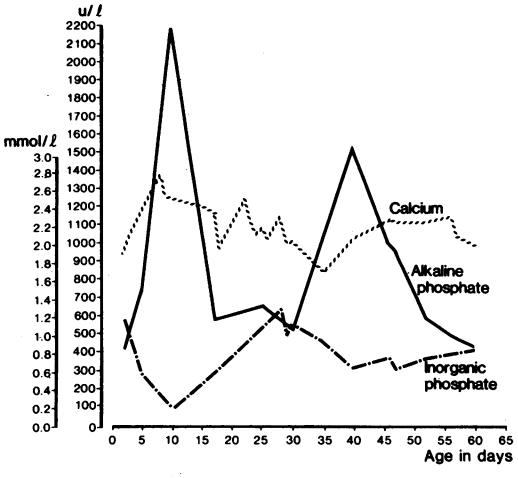

Figure 2. Serial calcium, inorganic phosphate and alkaline phosphatase from day 2

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Misleading aortographic finding in acute aortic dissection explained[1]

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Aortography remains the mainstay of the classification of aortic dissections and in the decision as to whether treatment should be operative or non-operative (De Bakey *et al.* 1965, Miller *et al.* 1979). Cineangiography represents a more recent refinement of what is essentially the same technique but may make interpretation easier (Gutierrez *et al.* 1980). Although this technique remains the 'gold standard' by which newer methods are judged, it is not without problems. Shuford *et al.* (1969) have discussed these problems and describe how errors in interpretation may arise. More recently the use of computer tomography has been described

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