control of choreiform movements in Sydenham's chorea(3) prompted us to try the drug in our case. The child was put on sodium valproate in the dose of 20 mg/kg/day in two divided doses and other sedatives were withdrawn. Two weeks after the initiation of therapy there was a remarkable control of abnormal movements and the child could roll over from prone to supine position. After 4 weeks of therapy when she was last seen, there was further control of choreoathetoid movements and child was able to sit, hold objects in hand and imitate. She could also bear weight on her legs. Sodium valproate not only controlled the abnormal movements in the present case, but also helped the child to exhibit her milestones.

Discussion

Sodium valproate is an effective anticonvulsant, which has been recently used in the treatment of Sydenham's chorea(3,4). The present study indicates that this drug could be used for the control of severe choreoathetoid movements resulting from kernicterus as well. We suggest that a larger controlled study should be carried out to validate our finding. The precise mechanism by which valproate controls involuntary movements is not known. However, sodium valproate is known to raise the level of gamma-aminobutyric acid (GABA), particularly in the striatum nigra(5). This increase may exert its effect through modification of the GABAergic synaptic transmission and hence control the abnormal movements. This effect on the basal ganglia may be totally different from the anticonvulsant effect of valproate(3).

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Perinatal Hypophosphatasia

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Rathbun(1) coined the term 'hypophosphatasia' for a heritable metabolic

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bone disease characterized by subnormal alkaline phosphatase activity, increased plasma levels and urinary excretion of alkaline phosphatase substrates (phosphoethanolamine, inorganic pyrophosphate and pyridoxal-5'-phosphatase) and defective bone mineralization manifesting as rickets in infants and children(1,2). It is a rare disease with an incidence of about 1 in 1,00,000 live births(3), occurs in all races and its clinical expression is extremely variable. In the present communication, we report a case of perinatal hypophosphatasia exhibiting an unusual survival despite belonging to the most severe form of the disease.

Case Report

A term (41 weeks 6 days) female baby was delivered by cesarean section (indication–cephalopelvic disproportion with previous cesarean section) to a 25-year-old 5th gravida (P_{1+3}) mother with no history of consanguinity. The antenatal period was uneventful. The baby was born without any birth asphyxia (1 minute Apgar score 8/10), weighed 2900 g at birth and had a crown heel length and head circumference of 46 cm and 32.5 cm, respectively. The abnormalities noted at birth were anterior bowing of both thighs with folds of skin on the medial aspect; large anterior (3×2.5 cm) and posterior (1×1.5 cm) fontanelles and widely separated sutures. The baby developed generalized seizures and groaning at 48 h of life without any other features of intracranial hemorrhage like poor activity, hypo or hypertonia, bulging anterior fontanelle, apnea, cyanosis or respiratory distress. The investigations revealed : Hb 14 g/dl, TLC 8,000/mm^3, polymorphs 22%, lymphocytes 78%, normocytic normochromic RBCs and adequate platelets. Blood sugar, serum calcium, phosphorus and alkaline phosphatase levels were 69 mg/dl, 8.5 mg/dl, 5.2 mg/dl and 3 King Armstrong units (normal range 5-15), respectively. Urinary calcium was 2.25 mg/24 h (normal range 0.1-0.3 mg/24 h), while phosphoethanolamine excretion in urine could not be estimated. VDRL tests of the baby and the parents were non-reactive. Skiagram showed bowing of femora and metaphyseal fraying of the long bones (*Fig. 1*). The metaphyseal fraying was more marked at the lower ends of radius and ulna but there was no deformity in the bones of the upper limbs (*Fig. 2*). Skull showed poor mineralization with widely open fontanelles and sutures without any fracture lines. Serum calcium, phosphorus

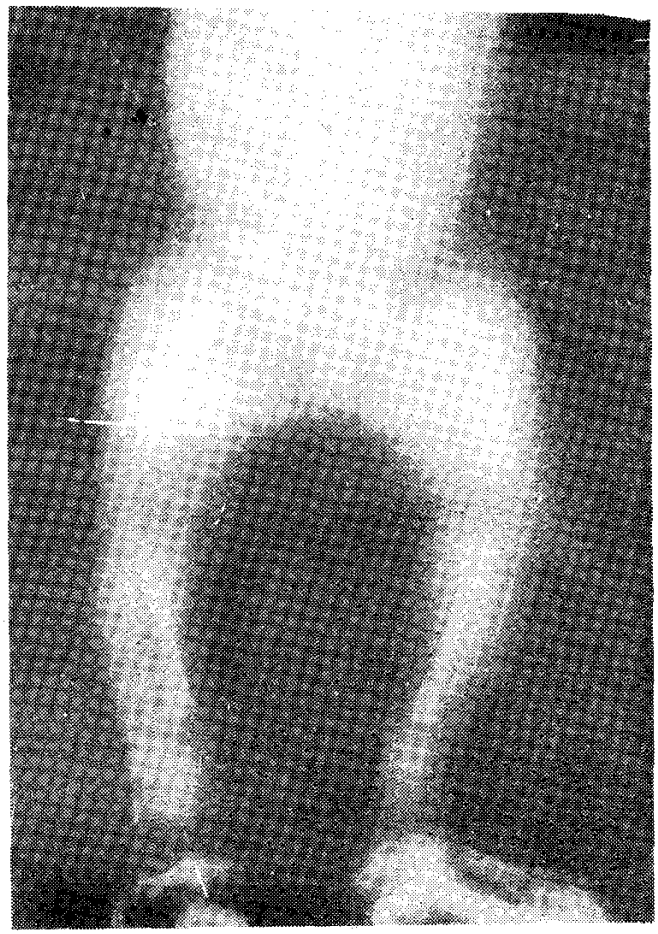

Fig. 1. Skiagram of the lower limbs showing bowing of femora and metaphyseal fraying of long bones.

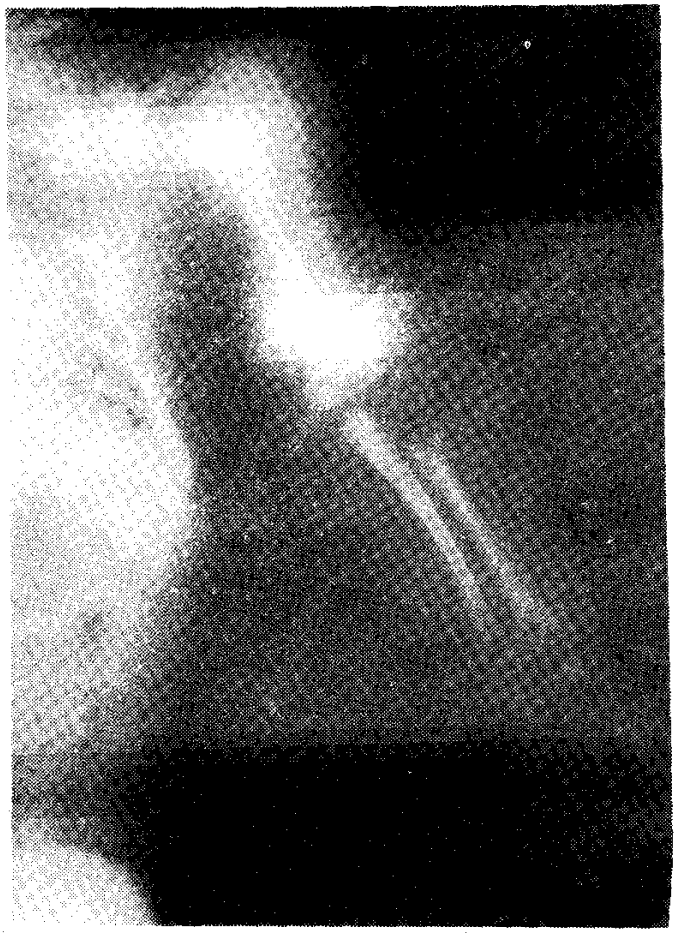

Fig. 2. Skiagram of upper limb showing metaphyseal fraying at the ends of radius and ulna.

and alkaline phosphatase levels of the mother and other sibling and skiagram of the pelvis of mother were within normal limits. Ultrasonography done 7 days prior to delivery demonstrated bilateral bowing of femora.

Treatment was started with intravenous fluids and anticonvulsants. Though there was no recurrence of convulsions, the baby developed features of septicemia on the 4th day of life. Repeat hemogram showed Hb 14 g/dl, TLC 23,000/mm^3, polymorphs 89%, lymphocytes 11% with vacuolisation and toxic granules in polymorphs and adequate platelets. Blood culture was sterile and CSF examination was normal. The baby was treated with parenteral antibiotics (ampicillin and cefotaxime) for 2 weeks. The patient gradually improved and was discharged on breast feeding. At the last follow up at 3 months of age, the baby was gaining weight normally and had been convulsion free but the bony deformities all persisted.

Discussion

The first case of hypophosphatasia was described in 1948 in an infant who died from severe rickets, weight loss, seizures and subnormal alkaline phosphatase activity in serum, bone, lungs, kidney and bowel(1). The presence of skeletal manifestations, subnormal alkaline phosphatase level, increased urinary calcium and seizures in our case suggest the diagnosis of hypophosphatasia. The patients of this condition are generally classified into one of the four forms (perinatal/lethal, infantile, childhood and adult) depending upon the age of skeletal manifestations but there is considerable heterogeneity within each of these forms. Detection of bony abnormalities antenatally by ultrasonography in our case and manifestations in early neonatal period indicates the perinatal form of the disease. The presence of hypercalciurea with normal calcium level observed in the present case corroborates the findings of previous authors(4). Demonstration of increased phosphoethanolamine in urine supports the diagnosis but is not specific for hypophosphatasia(2). The normal blood chemistry and skiagram of pelvis of the mother and subnormal level of alkaline phosphatase and hypercalciurea in the baby differentiates this condition from Vitamin D deficiency rickets manifesting during neonatal period. Another clinical condition which simulates hypophosphatasia radiologically is metaphyseal dysostosis but the latter is distinguished by normal

blood chemistry and absence of changes in the skull(5).

Although various measures like prednisolone, oral buffered phosphates and enzyme replacement therapy have been tried in this disease, there is no well established therapy(4,6). Death in the perinatal form occurs either *in utero* or few hours or at the most a few days after birth due to marked inability to ventilate the lungs properly in such patients(7). The unusual survival in the present case may have been due to the absence of severe involvement of the thoracic cage in the rachitic process.

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Neonatal Adrenal Hemorrhage

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The relatively large size and vascularity of the neonatal adrenal gland makes them vulnerable to traumatic and asphyxial injuries, with incidence of adrenal hemorrhage at necropsies being 1.7 per 1000(1). Though the exact cause of neonatal adrenal hemorrhage (NAH) is unknown, postulated etiologies include maternal diabetes, obstetric trauma, asphyxia, thrombocytopenia and coagulation defects(2,3). Our recent encounter of four patients with NAH who were evaluated by ultrasound(USG) prompted this report. USG which is a safe, portable, noninvasive, accurate, useful tool for the diagnosis of NAH, was the only imaging modality employed for diagnosis and follow-up of our cases.

Case Reports

From June to November 1990, four male neonates with mean birth weight of 3450 g presented with a flank mass in the

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