Ultraviolet irradiation for hepatic rickets

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SUMMARY An infant with chronic cytomegalovirus hepatitis and a child with atypical Alagille’s syndrome had vitamin D deficiency rickets due to malabsorption. Both received ultraviolet irradiation. This treatment corrected biochemical abnormalities and healed the rickets. In the infant use of a sunlamp at home maintained normal 25 hydroxyvitamin D for over a year. Our study shows that ultraviolet irradiation is an effective treatment of hepatobiliary rickets.

Vitamin D deficiency rickets is a common complication of chronic hepatobiliary disease in children.1 2 We and others have shown that the cause of the vitamin D deficiency is malabsorption of vitamin D, not impairment of hepatic hydroxylation.3 We have shown that intravenous injection of physiological doses of vitamin D can cure rickets in children with hepatobiliary disease. The present study assessed the effectiveness of ultraviolet irradiation in two children with hepatobiliary rickets.

Case reports and methods

Case 1
An 11 month old girl was admitted to hospital with pathological fractures of long bones. She was known to have had chronic cytomegalovirus hepatitis since birth. A liver biopsy specimen at 3 months of age had shown giant cell transformation of hepatocytes as well as hepatocyte necrosis and focal fibrosis. Later biopsy confirmed the presence of cirrhosis. She was deaf, small, and jaundiced, and had hepatosplenomegaly, a rachitic rosary, and pain in the left arm and leg. She had been receiving 600 U of vitamin D daily by mouth.

Plasma total bilirubin concentration was 203 μmol/l and direct bilirubin was 178 μmol/l. Alkaline phosphatase activity was 2300 U/l, aspartate aminotransferase activity 300 U/l, and bile acid concentration 39 μmol/l. Plasma calcium concentration was 1·64 mmol/l, phosphate 0·81 mmol/l, 25 hydroxyvitamin D 4 nmol/l (reference range 20–80 nmol/l), and parathyroid hormone 110 pmol/l (reference range 3–25 pmol/l). She had generalised aminoaci-
duria and faecal fat was 40% of intake. Skeletal radiographs showed severe osteoporosis, florid rickets, and fractures of the left humerus and femur. The patient received ultraviolet irradiation daily in hospital for 32 days (for technique see below). She continued it daily at home for a year with a sunlamp.

Case 2
An 8 year old boy, known to have atypical Alagille’s syndrome, was admitted to hospital for investigation of rickets. He had a triangular face with prominent forehead, deep set eyes, mild hypertelorism, and a small chin. He had deep jaundice, anaemia, hepatosplenomegaly, bleeding gums, muscle weakness, ataxia, absent tendon reflexes, and genu valgum. Previous investigations had indicated vitamin K and E deficiencies. A liver biopsy specimen showed a paucity of interlobular bile ducts and cholestasis. Faecal fat was 66% of intake. The patient had been receiving 11 000 U vitamin D daily by mouth.

Concentration of plasma total bilirubin was 520 μmol/l and direct bilirubin 360 μmol/l. Alkaline phosphatase activity was 240–680 U/l and aspartate aminotransferase activity 200 U/l. Plasma calcium concentration was 2·25 mmol/l, phosphate 0·64 mmol/l, and 25 hydroxyvitamin D undetectable. Radiographs showed mild rickets. The patient received ultraviolet irradiation in hospital for 44 days.

Ultraviolet treatment
In hospital ultraviolet treatment was applied with a Burdick lamp UV 800 (Burdick Corporation). The manufacturer stated that this generates an ultraviolet spectrum of 220–320 nm, two thirds of it in the region of 280–320 nm. Lamp to skin distance was 75 cm. The full body, excluding the head and neck, was irradiated. The irradiation started at 20 seconds and rose to 10 minutes each anteriorly and posteriorly.

The sunlamp used at home by case 1 was purchased from Hanscroft (Ajax), and the manufacturer stated that it generates 26% ultraviolet B (290–320 nm) and 70% ultraviolet A (320–400 nm). Lamp to skin distance was 60 cm. The daily ultraviolet dose was 10 minutes each anteriorly and posteriorly during the first months at home, but was reduced to 3 minutes anteriorly as a maintenance dose.
Results

In case 1 the ultraviolet irradiation made the skin erythematous at first and later tanned. The patient began to move the fractured limbs within a few weeks of starting treatment. The biochemical response to ultraviolet irradiation is shown in fig 1. All the measurements started to improve within a few days, and, with the exception of alkaline phosphatase, maximum improvement was achieved in one month despite repeated respiratory infections and continuing hepatic dysfunction. The severe rickets (fig 2a) improved within two weeks (fig 2b) and resolved in six months (fig 2c). One year after starting ultraviolet treatment the patient was maintaining normal concentrations of plasma 25 hydroxyvitamin D (40 mmol/l), calcium (2.42 mmol/l), and phosphate (1.5 mmol/l). The patient continued to have jaundice, hepatosplenomegaly, and abnormal liver function tests.

In case 2 the skin became erythematous and dry. During treatment normocalcaemia was maintained and plasma phosphate concentration rose to 1.52 mmol/l. The low plasma 25 hydroxyvitamin D concentration became normal (50 nmol/l) during the ultraviolet irradiation. After discharge from hospital the 25 hydroxyvitamin D concentrations gradually fell. Although no follow up study could be done, it is known that his liver disease worsened and he died from liver failure and uncontrollable haemorrhage two years later.

We measured plasma vitamin D and 25 hydroxyvitamin D concentrations simultaneously before and during ultraviolet irradiation in case 2: before ultraviolet treatment vitamin D was <2.5 nmol/l and 25 hydroxyvitamin D 5.7 nmol/l, and during treatment vitamin D was 22 nmol/l and 25 hydroxyvitamin D 25.5 nmol/l.

Discussion

This study shows that ultraviolet irradiation can cure
Ultraviolet irradiation for hepatic rickets

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SUMMARY

A review of 173 infants who were started on whey based formula showed that 40 (23%) had their feeds changed, usually to a casein based feed by 6 weeks. A double blind randomised trial, comparing casein based with whey based formula confirmed that about 20-25% of babies have their formula changed within six weeks irrespective of the nature of the feed on which the infant was started.

Are babies more satisfied by casein based formulas?

Two types of infant formulas based on cows’ milk are now marketed by baby food manufacturers and are officially approved as infant feeds; those which contain mainly casein as their protein and those which are mainly whey based.1 The latter tend to be more highly modified and resemble breast milk more closely in their chemical composition with lower electrolyte and total protein content and a smaller solute load. It is policy in most maternity units to offer babies whey based formulas but many babies are subsequently switched to casein based formulas.2

There appears to be a widespread belief among mothers and some health workers that casein based feeds are in some way more ‘satisfying’ and so a switch to them in crying, unsettled babies or in infants who have various digestive symptoms such as colic, wind, vomiting, or possetting is not unusual. There may even be a notion that switching to a formula perceived as ‘stronger’ in some ways indicates progress by the baby. It has recently been suggested that parents may be ‘voting with their mouths’ in switching from whey to casein based formulas and that perhaps maternity units should ‘drop their emphasis on whey dominant milks that

hepatobiliary rickets. Jaundice did not interfere with cutaneous generation of vitamin D. Both patients had developed vitamin D deficiency rickets even though they had been receiving supraphysiological doses of vitamin D by mouth. Comparison of plasma vitamin D and 25 hydroxyvitamin D concentrations indicated no accumulation of vitamin D. Thus there was no evidence that a defect in hepatic 25-hydroxylation was responsible for the low 25 hydroxyvitamin D concentrations.

Most patients with mild malabsorption of vitamin D can be treated with an increase in oral vitamin D. Some investigators prefer using 25 hydroxyvitamin D because it is more polar and slightly better absorbed. If malabsorption is severe and a child develops vitamin D deficiency while receiving pharmacological doses of vitamin D or 25 hydroxyvitamin D, parenteral vitamin D or ultraviolet treatment are the next alternatives. The choice depends on the availability of the treatment modality. Intravenous vitamin D formulations are investigational, and their administration is inconvenient.

Although ultraviolet irradiation can be achieved from exposure to sunlight, it is generally available only seasonally, and children with chronic liver disease are often too ill to spend much time outdoors. A sunlamp can be an effective substitute and can be purchased at a modest cost. The effective ultraviolet dose to raise 25 hydroxyvitamin D concentrations varies from lamp to lamp4 and depends on skin pigmentation.5 The optimal ultraviolet dose can be determined by regular monitoring of 25 hydroxyvitamin D concentrations.

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References


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