Short Reports

Archives of Disease in Childhood, 1970, 45, 581.

Osteogenesis Imperfecta Tarda Treated with Sodium Fluoride

Osteogenesis imperfecta is a disorder of connective tissue, existing in a severe form (congenita) and a milder form (tarda) both being inherited as an autosomal dominant (Mckusick, 1966). Recently Aeschlimann, Grunt, and Crigler (1966) and Bilginturan and Ozsoylu (1966) reported three infants with osteogenesis imperfecta congenita who improved, as regards survival and frequency of spontaneous fractures, while on sodium fluoride for about 12 months.

The effects of sodium fluoride on the clinical course of a child with osteogenesis imperfecta tarda are described.

Case Report

This girl was born at term after an uneventful pregnancy and labour, with a birthweight of 3·18 kg. Blue sclerotics were noted after birth and skeletal x-ray survey on the first day of life revealed fractures of the 5th, 6th, and 7th ribs on the left side—features typical of osteogenesis imperfecta. During the subsequent 6 months she progressed very slowly, requiring almost continuous treatment for recurrent pulmonary infections. She received the usual vitamin supplements. Her progress regarding frequency of spontaneous fractures up to the age of 20 months was as follows: Birth—ribs 5, 6, and 7; 7 months—clavicle (right); 11 months—femur (right); 13 months—radius, ulna (left); 20 months—clavicle (left), ulna (right), tibia and fibula (left).

At the age of 20 months she weighed 8·0 kg., was unable to walk, but could manage to sit up unaided. She appeared of average intelligence. Hearing was normal. Investigations—serum calcium, inorganic phosphate, haematological values and urine amino acid chromatography were normal; alkaline phosphatase slightly raised. She was now (20 months) started on sodium fluoride, 2 mg. twice daily, increased to 3 mg. twice daily after 3 months. Her motor development greatly improved after three months of this treatment, she could now walk, climb stairs, and though she had falls, no fractures were noted. At 28 months she was knocked down by a bicycle; she had a few superficial bruises but a full skeletal x-ray showed no fractures. She remained on sodium fluoride for 14 months (i.e. between 20–34 months of age). Two months after it was discontinued she fractured her right fibula after a minor fall, and one month later, after a jump from a chair, she collapsed her 7th and 8th thoracic vertebrae. Haematological values, serum calcium, inorganic phosphorus and alkaline phosphatase estimated every 3 months remained normal. Examination of teeth did not show fluorosis.

Family history. The father has osteogenesis imperfecta tarda and had had a number of fractures of long bones between 5–14 years of age. One other child, a boy aged 6 years, is well.

Discussion

Since osteogenesis imperfecta tarda is characterized by exacerbations and remissions of varying periods, often with great spontaneous improvement at puberty, it is difficult to claim certainty of benefit from treatment. In the infant with osteogenesis imperfecta congenita reported by Aeschlimann et al. (1966) metabolic studies showed that fluoride influenced the Ca/P ratio in bone, and Zipkin, Likins, and McClure (1959) have shown that in rats with rickets fluoride increased phosphorus retention, hence there can be no doubt that bone composition can be influenced by sodium fluoride. Regrettably, calcium and fluoride balances were not possible in the present patient. Aeschlimann et al. (1966) and Bilginturan and Ozsoylu (1966) reported radiological improvement in the bone density and architecture in their patients while on fluoride. Similarly, improvement was thought to be observable in our patient, but x-rays were not taken under strictly comparable conditions, so that no valid claims could be made. Significantly, our patient's motor attainments advanced and she gained 3·68 kg. and grew 8·5 cm. during the 14 months she was taking sodium fluoride, considerably more than in any similar period before treatment.

Summary

Administration of sodium fluoride 4–6 mg. per day appeared to reduce the number of fractures and to improve the growth rate in a girl of 20 months with osteogenesis imperfecta.

I am most grateful to Professor C. E. Dent for his helpful comments.

References

Short Reports

Recurrent Haemophilus Septicaemia and Immunoglobulin Deficiency

Infection with *Haemophilus influenzae* type b is often subclinical but may give rise to septicaemic illnesses (Farrand, 1969). One infection appears to give long-lasting immunity (Fothergill and Wright, 1933). Hypo-γ-globulinaemia or splenectomy can predispose to a single attack of haemophilus meningitis, or to repeated respiratory infections due to non-capsulated organisms. Recurrent pneumococcal septicaemia has often been described in immune-deficiency syndromes, but in a survey of the published papers no report of a second episode of haemophilus septicaemia was found (Good et al., 1960; Eraklis et al., 1967).

Case History

This 10-month-old boy was found in a collapsed state, with clinical signs of meningitis. CSF contained 4800 polymorphonuclear cells per cu. mm., with a protein of 256 mg./100 ml. and glucose of 25 mg./100 ml. The stained film showed Gram-negative bacilli, and on culture *H. influenzae* type b was isolated in large numbers. Blood cultures also yielded *H. influenzae* type b. WBC 15,600 per cu. mm. (53% neutrophils, 40% lymphocytes, 7% monocytes). The patient was treated with ampicillin and responded satisfactorily. He was discharged home fully recovered after two weeks, and ampicillin was withdrawn after one further week.

The boy had been in good health before his attack of meningitis, and had not previously suffered any infection of note. He remained very well for four months, until when aged 14 months he developed otitis media. This relapsed after a 5-day course of penicillin and was treated with ampicillin for 7 days with full recovery. No bacteriology was undertaken, but as *H. influenzae* type b is a common cause of otitis media in early childhood, and characteristically shows resistance to penicillin and response to ampicillin, it seems very likely that this episode of infection also was caused by *H. influenzae*.

One month later, aged 15 months, the boy became febrile again, and was admitted to hospital with a temperature of 40°C. He was reluctant to use the right arm, and there was some local tenderness over the inner margin of the right humerus but no sign of local inflammation and no definite abnormality on x-ray examination. CSF normal; WBC 16,900 (84% neutrophils); ESR 20 mm./hr. Blood cultures were taken. No precise diagnosis was possible at this stage, and the possibility of an early focus of osteomyelitis in the humerus was noted. Large doses of intramuscular benzyl-penicillin were given, and after 24 hours the temperature returned to normal.

He was allowed to continue with oral penicillin at home, but after three days was readmitted with an inflammatory swelling over the distal end of the right humerus. ESR had risen to 73 mm. per hour, and further blood cultures were taken. Osteomyelitis of the right humerus was diagnosed, and treatment started with ampicillin and lincomycin.

*H. influenzae* type b was isolated from the initial blood

<table>
<thead>
<tr>
<th>Age</th>
<th>IgA (mg./100 ml.)</th>
<th>IgG (mg./100 ml.)</th>
<th>IgM (mg./100 ml.)</th>
<th>Haemophilus influenzae type b Haemagglutination Titre</th>
</tr>
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<tbody>
<tr>
<td>Case: 1st specimen</td>
<td></td>
<td></td>
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<tr>
<td>(day 12 of osteomyelitis)</td>
<td></td>
<td></td>
<td></td>
<td>Nil</td>
</tr>
<tr>
<td>Case: 2nd specimen</td>
<td></td>
<td></td>
<td></td>
<td>Nil</td>
</tr>
<tr>
<td>(day 23 of osteomyelitis)</td>
<td></td>
<td></td>
<td></td>
<td>Nil</td>
</tr>
<tr>
<td>Father</td>
<td>15 mth.</td>
<td>Undetectable (72 ± 35)</td>
<td>130 (792 ± 216)</td>
<td>220 (47 ± 18)</td>
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<td>300 (288 ± 121)</td>
<td>1500 (1200 ± 300)</td>
<td>83 (80 ± 29)</td>
</tr>
<tr>
<td></td>
<td>Adult</td>
<td>110 (288 ± 121)</td>
<td>1000 (1200 ± 300)</td>
<td>83 (80 ± 29)</td>
</tr>
<tr>
<td></td>
<td>7 yr.</td>
<td>140 (178 ± 58)</td>
<td>135 (960 ± 264)</td>
<td>83 (53 ± 20)</td>
</tr>
<tr>
<td></td>
<td>5 yr.</td>
<td>30 (135 ± 40)</td>
<td>500 (960 ± 240)</td>
<td>83 (46 ± 15)</td>
</tr>
<tr>
<td></td>
<td>3 yr.</td>
<td>20 (136 ± 40)</td>
<td>500 (960 ± 240)</td>
<td>83 (46 ± 15)</td>
</tr>
</tbody>
</table>

Normal levels of immunoglobulin are shown in parentheses (Stichm and Fudenberg, 1966).
Osteogenesis imperfecta tarda treated with sodium fluoride.

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