FIG. 2.—Physiological adrenal stimulation test. Plasma cortisol ($\mu\text{g}/100\text{ ml}$) is plotted in the dexamethasone-suppressed patient given five increasing doses of intravenous tetracosactrin. Black columns represents the mean of 2 tests before transfusion therapy was started, the shaded columns represent 18 months after starting, and the open columns 2 years after starting regular transfusion therapy.

Discussion

The complications in the above case are almost certainly not unique. The hearing abnormalities are present in many cases both transfused and not transfused, but are more evident in those not transfused (J. W. P. Hazell, N. McIntosh, and C. B. Modell, unpublished). The improvement in height has been well recorded previously (Brook *et al.*, 1969). The endocrine impairment is interesting. In high transfused patients it is probably due to iron deposition in the endocrine glands similar to that seen in haemochromatosis, but it is also known that cases of thalassaemia intermedia develop endocrinopathy (Bannerman *et al.*, 1967). In these cases it has been presumed due to increased gastrointestinal iron absorption with deposition in endocrine organs. If serum ferritin levels do indeed represent total body iron (Jacobs *et al.*, 1972) then the iron load in this case was originally low and endocrine function has improved as iron load has increased. It seems likely that hypoxaemia in the untransfused patient owing to a combination of anaemia and the shift to the left of the oxygen dissociation curve has depressed endocrine function. These have been reversed with transfusion. There is some evidence in mice that hypoxia in addition to iron deposition is necessary to cause cardiotoxicity (Necheles, Beard, and Allen, 1969). The alternative possibility is that iron absorbed gastrointestinally is more toxic to the endocrine glands, but this is difficult to prove. Puberty is always delayed in patients with thalassaemia whether transfused or not (Canale *et al.*, 1974). It is impossible in this case

to prove that transfusion initiated puberty, but the growth velocity and puberty grading improved so dramatically that it seems likely that this was the case.

Summary

A case of nontransfusion-dependent thalassaemia major is presented showing growth retardation, recurrent fractures, late onset of puberty, deafness, and endocrine impairment of pancreas and adrenal gland. Regular blood transfusion to maintain mean haemoglobin at about 11 g/dl improved all these problems. It is suggested that this improvement was related to improved tissue oxygenation.

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Phalangeal microgeodic syndrome of infancy

The phalangeal microgeodic syndrome of infancy was first described as an entity by Maroteaux in 1970, but seems to be poorly documented in the subsequent published reports. We record a further case.

Case report

A girl was referred for x-ray on 30 October 1974 at the age of 18 months with a 2-month history of swollen, red, and shiny fingers (Fig. 1). Clinically they resembled chilblains and were warm to touch but the condition seemed to cause no discomfort, and movements were restricted only by the swelling. There was no systemic upset. According to the mother tiny blisters had preceded the swelling, and the patient had had mumps some weeks before.

X-ray of the hands (Fig. 2) showed sharply punched-out cortical erosions accompanied by periosteal reaction in the areas corresponding with the external lesions. The joint surfaces were unaffected. The rest of the skeleton was normal. Investigations elicited only one positive finding, an erythrocyte sedimentation rate of 50 mm/h at first attendance, dropping to 23 mm/h one week later. BCG had been given at birth, and the Heaf tuberculin test was positive, eliminating sarcoidosis. Blood calcium, uric acid, blood count, and blood urea were all within normal limits. Coombs's test and test for LE cells were negative. Mental development was normal.

FIG. 1.—Showing chilblain-like lesions in fingers.

FIG. 2.—Showing bone erosions and periosteal reaction.

No treatment was given and the external appearances had improved by 28 January 1975, though the bone defects had become more defined radiologically. These slowly regressed and 6 months later both bone x-rays and soft tissues appeared normal.

Discussion

At a meeting of the European Society of Paediatric Radiology in Warsaw in 1969, Pierre Maroteaux reported and subsequently published (1970) 5 cases of the phalangeal microgeodic syndrome collected from various sources. The radiological description 'microgeodic' conveys the likeness to 'a nodular stone containing a cavity', such being the definition of a geode in the *Shorter Oxford English Dictionary*, and of the original French word in *Petit Larousse*.

Maroteaux re-examined 3 similar cases from published reports, 2 sisters by Bøggild (1944), and one by Newns and Hardwick (1939), which had been regarded as sarcoidosis. All but one of the cases were between 15 and 24 months, the exception being a child of 5 years, a sib of a younger case. Biopsy in the acute stage showed inflammatory changes and biopsy in a healed case showed only slight lamellar irregularity. One patient underwent arteriography with negative results. In one instance an eosinophilia of 10% was noted. Bøggild's 2 cases suffered recurrences in winter, but one of Maroteaux's patients presented in August and no case was especially sensitive to cold. This histology did not support the diagnosis of sarcoid and the abnormality was confined to the hands. Both sexes were equally affected. All cases regressed spontaneously, in some instances over a period of years. He concluded that the aetiology was unknown.

This syndrome is mentioned by Taybi (1975) in *Radiology of Syndromes*, and both the x-ray appearances and the clinical behaviour of our case corresponds with Maroteaux's description and illustrations. We can contribute no suggestion with regard to aetiology, but feel we should emphasize the benign course of the condition in spite of the florid clinicoradiological manifestations.

Summary

A case is described of chilblain-like lesions in the fingers with punched-out erosions in the phalanges on x-ray which appears to be an example of Maroteaux's phalangeal microgeodic syndrome of infancy.

Acknowledgement is made to Dr. Frank McCarthy who referred this case.

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Congenital varicella resulting from infection during second trimester of pregnancy

Congenital malformation of the newborn infant due to maternal varicella in early pregnancy has rarely been reported. Such a case is presented here with a review and discussion of published reports.

Case report

The infant was the third child of a 20-year-old woman. Her first pregnancy in 1970 resulted in the birth of an infant of 36 weeks' gestation who died at 8 days of age from disseminated herpes simplex infection, confirmed by liver histology and viral culture. The mother was noted to have exophthalmos antenatally, and vulvar herpes shortly after delivery. Her second pregnancy in 1972 resulted in a light-for-dates infant now alive and well. During this pregnancy she developed a subacute thyroiditis (de Quervain's) at the 15th week, which resolved spontaneously over the next 3 weeks, her thyroid function returning to normal.

Her third pregnancy in 1974 was complicated at 20 weeks by generalized varicella lasting approximately 10 days. 2 weeks after the start of this illness she was admitted because of undiagnosed abdominal pains. She had no vaginal bleeding and was discharged 12 days

later, but was readmitted at 30 weeks because of poor fetal growth and her bad obstetric history. Normal fetal growth had been recorded clinically until the time of her varicella infection and urinary total oestrogens and ultrasonic fetal biparietal diameters suggested poor fetal growth thereafter. The pregnancy ended with the spontaneous onset of labour at 34 weeks, a female infant being delivered by Wrigley's forceps on 13 October.

The infant's birthweight was 1580 g, length 39 cm, and head circumference 29.8 cm. Apgar scores were 7 at 1 minute and 10 at 5 minutes. Gestational assessment using Dubowitz's scoring system (Dubowitz, Dubowitz, and Goldberg, 1970) was 34 to 35 weeks, weight being below the 10th centile. Examination showed a left microphthalmia and enophthalmia, crusted vesicular skin lesions on the left hand and forearm, and a healed scar on the left side of the abdomen. She was initially nursed in an incubator, mild symptoms of respiratory distress settling over the first 48 hours. A maximum serum bilirubin of 144 $\mu\text{mol/l}$ was recorded on the third day. Assisted feeding by nasogastric tube was required for the first 4 weeks. A top-up transfusion of 30 ml of packed red cells was given at one month of age, when she had mild dyspnoea associated with a haemoglobin levels of 9.8 g/dl. Subsequent progress was uneventful.

The skin lesions healed with minimal scarring and good function of the left hand over the first 3 to 4 weeks. Examination of the eyes at 7 weeks of age showed corneal diameters of 10 mm on the right and 5 mm on the left. The right fundus appeared normal but the left was obscured by a central cataract. Neurological and developmental assessments have been otherwise normal up to 6 months after birth when outpatient follow-up was lost due to nonattendance.

Investigations. The viral titres obtained in the mother's and infant's serum are compared in the Table. An initially raised IgM of 0.48 g/l was found in the infant's serum (11 days after birth) and antiviral zoster IgM shown on a sucrose gradient. Skull and chest x-rays were normal. Peripheral blood film apart from the above-noted anaemia was normal. A thyroxine level and T_3 index, obtained because of the mother's

TABLE
Viral titres

	1st*	2nd	3rd	4th
Varicella-zoster				
Infant's sera	1/16	1/32	1/16	1/32
Mother's sera	<1/8†	<1/8	<1/8	<1/8
Cytomegalovirus				
Infant and mother's sera	<1/8	<1/8	<1/8	<1/8
Toxoplasmosis				
Infant and mother's sera	<1/8	<1/8	<1/8	<1/8
Herpes simplex				
Infant and mother's sera	1/64	1/64	1/64	1/64

*Serology sample separated by 10- to 14-day intervals.

†Low titre in the mother is compatible with infection 15 weeks earlier.