ally recognized that the presence of *H. influenzae* in meningitis (Rivers, 1922; Zinnemann, 1946; Allibone *et al.*, 1951), and probably also in bronchiectasis (Allison *et al.*, 1943), is responsible for the formation of a very tenacious greenish-yellow mucopus. Such pus in the nasopharynx may well produce a cough resembling a whoop.

Summary

From per-nasal swabs of 650 children aged 0-4 years 32 capsulated *H. influenzae* strains have been isolated.

Type f was found 11 times, type e 8 times, and type b 6 times, the other three types occurring less often.

The fact that the most frequently isolated types were not *H. influenzae* type b, while type b was the causative agent in all of 41 cases of *H. influenzae* meningitis occurring during the same period in Leeds and various other parts of England and Wales, suggests that this type is more virulent for man than the other five known *H. influenzae* types.

We are greatly indebted to Professor J. W. McLeod for access to the bacteriological material. We are glad to acknowledge Dr. G. C. Turner's help in providing us with *H. influenzae* strains from the whooping-cough inquiry after one of us (B.D.) had left the department.

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INFANTILE CORTICAL HYPEROSTOSIS

BY

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Although a few isolated cases of infantile cortical hyperostosis had probably been recorded previously, the first full description of the disease was given by Caffey and Silverman (1945); their report was closely followed by those of Smyth *et al.* (1946) and Caffey (1946). The disease, which affects the sexes equally, always becomes manifest before the age of 6 months (Caffey, 1950). Its three cardinal signs are excessive irritability, localized swellings in the soft tissues, and periosteal thickening of various bones. This last feature is always seen in bones underlying the swellings of soft tissues, but is often demonstrable radiologically in other bones. Fever, leucocytosis, and anaemia may also be present. The course of the disease extends over months, with unpredictable remissions and relapses, but in all cases observed by Caffey (1950) complete clinical cure has resulted.

The majority of cases have been described by North American authors. MacGregor and Davies (1949), reporting the second recorded case in this country, found 28 in the literature; they included, however, a case described by Ellis (1939) which must be regarded as not proved, while two of the cases of Caffey (1946) and two of Smyth *et al.* (1946), occurring in children over 1 year old, are more likely to be examples of hypervitaminosis A, a view now shared by Caffey himself (1950). Up to 1949, therefore, 24 proved cases had been described: since then 15 have been added, three of which are in the British literature (Astley, 1950; Sakula, 1950; and Smitham and Palmer, 1950). Although the disease is probably not as uncommon in this country as would appear, its true incidence can be judged only if recognized cases are reported. It is of importance also to determine the mortality rate accurately, since the disease is not invariably as benign as in Caffey's series. Two cases have been described (van Zeven, 1948; Mossberger, 1950), in which the patient died of secondary infection, as did one of the two reported here.

Case 1

A female infant aged 15 weeks was first seen by one of us (W. J. M.) on July 28, 1950, because of fever and swelling of the face. The baby, an only child, was born at term after an uneventful pregnancy and normal delivery, and weighed 7½ lb. (3.3 kg.). She was bottle-fed and had been given cod-liver oil but not orange juice. The mother and father were both healthy. She had been well until a week previously, since when she had been feverish and fretful, although she had taken feeds well. On the previous day the right side of the face had been noticed to be swollen.

On examination her rectal temperature was 99.4° F. (37.5° C.) and pulse 166. Her weight was 14 lb. (6.4 kg.). She did not seem to be acutely ill and no abnormality other than the facial swelling was found. This was present over the whole length of the right mandible and appeared to be part of the bone itself. It was not tender and the skin was unaffected.

A provisional diagnosis of infantile cortical hyperostosis was made, skiagrams were taken, and the mother was asked to bring the child back in three days. Next day, however, she became alarmed at the condition of the baby, who, at her doctor's request, was admitted to hospital.

On admission the physical findings were essentially unchanged. There was a persistent pyrexia at 100-101° F. (37.8-38.4° C.). Eight days after admission she developed

FIG. 1.—Case 1. Periosteal thickening of the mandible.

diarrhoea and vomiting, which failed to respond to supportive therapy, and she died five days later.

Investigations.—Skiagrams showed periosteal thickening of both mandibles, especially on the right side (Fig. 1). No other bone was involved. A blood count on August 2 showed: Hb, 60%; white cells, 22,700 (polymorphs 16,570); and on August 8: Hb, 64%; white cells, 14,300 (polymorphs 5,434). Wassermann and Kahn reactions were both negative.

FIG. 2.—Case 1. Mandible, showing numerous osteoblasts in periosteum. H. and E. ($\times 400$.)

At necropsy abnormal findings were confined to the mandible and the liver. The mandible showed along its whole length thickening of the periosteum, which could be stripped easily from the bone. Microscopically (Fig. 2) the periosteum was considerably thickened and very cellular, with numerous osteoblasts forming new bone of immature form with imperfectly developed Haversian systems. There was no evidence of inflammation or of trauma. The liver showed fatty infiltration.

Case 2

This patient, a male infant, was first seen by one of us (W. J. M.) on February 9, 1951, at the age of 4 months, because of swelling of the jaw. He was a full-term baby born to healthy parents after a normal pregnancy and delivery, weighing 6½ lb. (2.8 kg.). He was bottle-fed and had received vitamin supplements in adequate amounts. He was well until a month old, when swellings appeared over the lower jaw on each side; he became irritable, and refused to take full feeds. After about six weeks the facial swelling had subsided, but it recurred two weeks later and persisted. His appetite had gradually improved and was now normal. At no time had he been noticeably febrile.

On examination his temperature, pulse, and respirations were normal; weight 11½ lb. (5.1 kg.). The only abnormal findings on clinical examination were swellings overlying both mandibles and numerous small shotty glands in both posterior triangles of the neck. The swellings, which were hard and not tender, involved the whole length of the mandible and appeared to be part of the bone itself (Fig. 3). The overlying soft tissues did not seem to be involved.

FIG. 3.—Case 2. Showing swelling of lower jaw.

X-ray examination showed periosteal thickening involving both mandibles, both tibiae, and the left femur (Fig. 4). The Wassermann and Kahn reactions were both negative.

Progress.—The baby has been seen at intervals and has remained in good general health. He was last seen on August 24, when he was 10 months old and weighed 22 lb. (10 kg.). There was then still visible swelling of the right lower jaw, although this was less marked than it was originally. Skiagrams still showed periosteal thickening, which was fusing with the cortex.

FIG. 4.—Case 2. Periosteal thickening of left femur.

Discussion

These two cases show two of the three salient features of infantile cortical hyperostosis—namely, irritability and “cortical” thickening of bone. Neither showed swelling of the soft tissues. According to Caffey (1950) these appear long before there is any radiological evidence of bony involvement and disappear a considerable time before the bony changes. This would explain their absence in Case 2, which was not seen until three months after the onset of symptoms. In Case 1 it must be presumed that, the baby being fat, the facial swellings had passed unnoticed, since x-ray changes in the mandible were already evident when she was first seen, although the history was of only a week's illness.

Biopsies of the affected soft tissues have shown (Sherman, 1949; Sherman and Hellyer, 1950) necrosis and fibrosis of muscle with intimal proliferation in the blood vessels. Such intimal proliferation has also been found in the vessels of the periosteum, and this must lead to a diminution in the blood supply to the bone. It is known (Illingworth and Dick, 1949) that, while total deprivation of blood supply leads to necrosis of bone, diminution stimulates new bone formation, a process exemplified in the sabre tibia of congenital syphilis. Infantile cortical hyperostosis has been regarded as primarily a disease of bone, but the appearance of subcutaneous swellings as the initial manifestation and the microscopical evidence of changes in the blood vessels suggest that these are the primary disturbances and are responsible for the bony changes.

The term “infantile cortical hyperostosis” is a misnomer, since microscopical examination shows that it is in fact the periosteum which is thickened. Numerous fibroblasts appear in the periosteum, which is lifted off the bone; subperiosteal new bone is then formed, produces the bony swellings which may be palpable, and subsequently fuses with the cortex.

According to Sherman and Hellyer (1950) the only bones never reported to be involved are the phalanges, the small bones of the tarsus and carpus, the vertebrae, and the pelvis, but Sakula (1950) has described a case in which the pelvis was affected. Of all the bones, however, the mandible, clavicle, and ulna are most commonly involved (Caffey, 1950). When the mandible is involved, as in the present two cases, the child's appearance is characteristic enough for the disease to be suspected on sight.

The fever and leucocytosis which are commonly found point to an infection as the cause of the disease, and the peculiar age incidence suggests a prenatal origin. No micro-organism, however, has been recovered from affected tissues, although attempts have been made (van Zeben, 1948; Sherman and Hellyer, 1950), and the microscopical appear-

ance shows no evidence of infection (Fig. 2). Moreover, there was no evidence in the present two cases of maternal infection during pregnancy.

A prominent feature of this condition is extreme irritability, which may be due to pain in the bones; from observation of Case 1 it was thought that the child suffered from pain in the affected jaw. It is possible that, when the part becomes warm, pain is produced by vascular dilatation compressing the nerves in Haversian canals narrowed by new bone formation. This explanation is supported by Mossberger's (1950) finding of newly formed cancellous bone filling the Haversian spaces. The long course and marked irritability suggest an analogy with pink disease. It is notorious that cases of the latter are prone to secondary infection, and, to judge from reported cases, there seems to be a similar tendency in infantile cortical hyperostosis. One of our patients died in hospital of secondary infection, as did one of van Zeven's (1948). Mossberger's patient (1950) died suddenly at home, and necropsy indicated that an infection in the gut was the cause of death. Since three of the 41 recorded cases have ended fatally, this disease cannot be considered as benign as has been thought, and it would be sound practice to exclude these cases from hospital unless their admission was strongly indicated.

Summary

Two cases of infantile cortical hyperostosis are described; in one of these the patient died of secondary infection, to which these cases appear to be prone. The disease is therefore not invariably benign, and it would be prudent to avoid admission to hospital.

It is suggested that the bony changes are dependent on diminution in the blood supply to the bone due to intimal thickening, and that the disease is not primarily one of bone.

We wish to thank Dr. J. V. C. Braithwaite for the hospital notes of Case 1, which was admitted under his care, Dr. M. Bodian for the histological report, Dr. D. Forbes Lawson for the skiagrams, Mr. R. W. T. Gaze for the photomicrographs, and Drs. C. K. McCoan and T. M. Gibson for referring the patients to us.

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At the first annual general meeting of the Edinburgh Group of the Infantile Paralysis Fellowship the secretary gave a short outline of the history and work of the group (*Scotsman*, March 14). It is already acting as an information centre and has contacts with the Ministry of Labour and with the Edinburgh Council of Social Services. It has also offered to finance members who wish to take a degree by postal course. Priority at cinemas for members means that they do not have to wait in queues. Another project is a plan to supply television sets to patients in iron lungs. Also the group has managed to get increased supplies of steel needed for orthopaedic appliances.

OUTBREAK OF BORNHOLM DISEASE IN WEST FIFE PRACTICE

BY

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Quite a considerable amount of literature on the subject of Bornholm disease has accumulated within the past 20 years. The disease has now become more generally recognized as a definite entity.

The attention of one of us (W. W. W. McN.) was drawn to the symptomatology of the illness by Hopkins (1950), who described cases which had occurred in his practice. Similar cases had occurred from time to time in the practice here, but had gone unrecognized as Bornholm disease. In May, 1951, the appearance of the cases of two boys proved in retrospect to be the beginning of an outbreak, which continued during June and July. During that time 24 cases from a scattered community came under our care.

Bornholm disease, as we now know it, seems to be an infection of the diaphragm. Ronald (1942) made out a good case for this assumption and thought that the infection occurred primarily in the diaphragmatic muscle, but suggested it might also be primarily in the diaphragmatic pleura. Hopkins believed that when pleurisy did occur it was an extension of the disease from the muscle. The Coxsackie group of viruses is considered by Findlay and Howard (1950) to be the infecting agent.

The cases were investigated from the general practitioner's point of view. The first cases were seen at the end of May and the last on July 30. The outbreak was, therefore, almost exclusively confined to the months of June and July. Four patients were under 10 years of age, six were 10–19, seven 20–29, five 30–39, and two were 40 and over. There were seventeen males and seven females.

Symptoms and Signs

In almost every case the onset was sudden, pain in the upper abdomen or lower thorax being the outstanding feature. The site of pain and the frequency of occurrence are shown in Table I. The pain was definitely related

TABLE I.—Site and Frequency of Pain

Abdominal	20
Right upper subcostal	7
Bilateral subcostal	3
Vague upper	4
Central epigastric radiating to either side	3
Epigastric, spreading to lower left chest	3
Thoracic	4
Left lower anterior	2
Bilateral lower posterior	1
Right lower posterior	1

to respiration in most of the cases. In 20 it was much worse on breathing. Some complained that it was like a stitch in the side; others gasped on account of its severity and were afraid to breathe. Various adjectives were used to describe it: knife-like, terrible, sharp, shooting, unbearable. Some who initially had pain in the upper abdomen had pain in the lower thorax, on one or other side, within a few days. Others had transient pains in the lower thorax before the onset of the acute crippling pain in the abdomen. One patient had pain radiating into the right iliac fossa, which made him think he had appendicitis; another com-