

catheter was left *in situ* for six hours, later being withdrawn. The specimen of urine grew *Proteus vulgaris*. She was treated with Negram (nalidixic acid). Uneventful recovery followed.

## COMMENT

Spontaneous rupture of a kidney diseased as a result of chronic nephritis, stone, tuberculosis, tumour, pyelonephritis, aneurysm, hydronephrosis, or pyonephrosis is rare; the commonest disease associated with rupture is hydronephrosis due to stone, but even so rupture is almost always extra-peritoneal. Rupture into the peritoneal cavity is very rare because the peritoneum forms an additional barrier. Spontaneous intra-peritoneal rupture of a pyonephrosis is a most unusual occurrence and proves fatal if not dealt with promptly.

## Anodontia in Mongolism

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While partial anodontia occurs with comparative frequency, complete absence of teeth is very rare. Zilkens (1927) observed a case of complete anodontia in a labourer aged 25; Herbst and Apffelstaedt (1930) noted the condition in an otherwise healthy woman and cited six analogous cases collected by Busch; and Thomas (1939) recorded an example in a mongol.

Abnormalities of the teeth in mongolism (Down's disease) are well recognized (Benda, 1960). Thus the teeth are generally small, often abnormally shaped and hypoplastic, and they develop late. The dental system is thought to be immature and to evidence disharmonious growth, while unilateral anodontia is sometimes found (Spitzer and Robinson, 1955). A case of complete anodontia in a mongol is reported.

## CASE REPORT

The patient, a female mongol imbecile, showed complete anodontia of both dentitions. Her father, a farm labourer, was born in 1888, and her mother, formerly a domestic servant, was born in 1892. They married in 1919 and are known to belong to the dull group of a rural population, especially on the father's side. There was no consanguinity or history of mental disease in the ascendants. The family consisted of four children, three of whom were of normal intelligence and appearance; the fourth showed the features of Down's syndrome.

The patient was born in 1928 and admitted to an institution in 1930. She shows the typical stigmata of mongolism and her chromosome count is 47. On the Terman-Merrill scale she has an I.Q. of 41, draws at the 7½-year level, and reads at the 7-year level. Rapport is good but her responses are slow. Anodontia is confirmed by the detailed history available from the institution, where complete absence of milk and permanent teeth has been noted. Her mandibular gum margin shows well-formed superior ridging, and radiography has demonstrated neither retained teeth nor other abnormality in maxilla or mandible. Dentition in the sibs was normal.

## COMMENT

Though the presence of an additional chromosome in mongolism is now well recognized, aetiology continues to be a matter of debate, the only generally accepted causative factor being that of increased maternal age (Penrose, 1954).

Zaidi (1959) reported intraperitoneal rupture of a pyonephrotic kidney in a child of 27 days—the only case on record in an infant.

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The association of complete anodontia with mongolism is of significance in relation to their respective embryonic origins. In the embryo the first indication of oncoming tooth development is an epithelial plate, the dental lamina, which arises during the seventh week just gumward of the labial lamina. This soon becomes a horizontal shelf which projects perpendicularly from the labial lamina and extends well into the substance of the primitive gum. This is brought about as the gum, upgrowing, buries the dental lamina deeper and deeper. Each dental lamina thereby courses alongside the curving labial groove and lies just gumward of it. At intervals along the epithelial lamina there develops simultaneously a series of knob-like thickenings called the enamel organs which will produce the enamel and serve as the moulds for the future teeth. Early in the third month the deeper side of each enamel organ presses against a dense accumulation of mesenchyme, and the epithelial surface of contact invaginates and grows round the mesenchymal mound until the whole organ is hollowed like a thick cup. The concavity thus formed is occupied by the condensed mesenchymal tissue of the dental papilla, which is destined to differentiate into dentine and pulp (Arey, 1954).

Essentially, mongolism is a syndrome characterized by the occurrence of a number of physical anomalies of ectodermal derivation. The mongol child is supposed to go through a fairly normal organogenetic period in the first few weeks of foetal development, but thereafter developmental deceleration takes place and anomalies of brain, heart, bones, and dentition become apparent. A number of the published cases showed, in addition to anodontia, analogous defects of ectodermal origin referable to the period around the sixth and seventh weeks of development. Thus Zilkens's patient showed webbing of toes and developmental disturbances of hair and nipples.

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