ANODONTIA IN MONGOLISM.

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ANODONTIA is a physical anomaly which may present itself in varying degrees, and although cases of partial anodontia occur with comparative frequency, the condition of complete anodontia is rare.

In the recent literature one case of complete anodontia is described by Zilkens (1927) in a man of 25 years. Herbst and Apffelstaedt (1930) describe the same condition in a woman, whose two daughters had very undeveloped dentitions. Cases are described by Kjaer and Cavallaro in which one jaw was completely edentulous from birth.

The present case is that of a female mongolian imbecile who has complete anodontia of both first and second dentitions. She is rather remarkable also in the fact of her longevity, being 65 years of age; a 63-year-old mongol is mentioned by Tredgold.

The patient presents a number of the usual anomalies associated with this form of primary oligophrenia, namely, a small, brachycephalic skull with some flattening of the occipital and frontal regions, and, incidentally, absence of the frontal sinuses; there is obliquity of the palpebral fissures, accompanied by convergent strabismus, the tongue shows marked fissuring and hypertrophy of circumvallate papillæ, the mouth drooping with peripheral rhagades. The hands have the typical short incurved little fingers, with creasing of simian form. Mentally she is a low-grade imbecile, able to talk in monosyllables, in the deep guttural voice characteristic of this condition; she plays with simple toys and has to be supervised over her physical needs. The anodontia is confirmed by the detailed history furnished by three older sisters (all reliable witnesses) of complete absence of milk and permanent teeth, with no extractions at any time in her life. The mandibular gum margin has a well-formed hard superior ridging.

Radiography shows a well-formed mandible, with no evidence of retained teeth or other anomaly, but with typical senile changes, such as obtuseness of the mandibular angle (125°), and a wide angle between the coronoid process and condyle of the ramus. The maxilla is also clear of any evidence of retained teeth.

The association of the condition of complete anodontia with mongolism is of interest in considering their respective embryonic origins. In the development of the embryo (Arey) the dental *anlages* first make their appearance at six weeks, the dental lamina being laid down as an ectodermal invagination of the epithelium of the mouth. From this the enamel organs, the primordia of the enamel of the teeth, develop, the dentine, pulp and cement being developed from the mesenchymal tissue of the dental papilla later. The dental lamina is the ectodermal *anlage* of the buds of the milk dentition. Anodontia must be assumed to be due to an aplasia of the dental lamina.

Mongolism is a syndrome characterized by the occurrence of a number of physical anomalies of ectodermal derivation, each of which is the result of a variation from the normal of comparatively uniform extent when examined from case to case; so much so that to the casual observer acquainted with the condition it is readily recognized. A condition resulting in such uniformity of variation from the normal could arise at two stages in embryonic development; firstly the germ defect might be present from the time of fusion of the zygotes, or secondly, the condition must be due to a factor operating at a time when ectodermal differentiation is at its height.

The problem of causation of malformation, in particular, of critically timed developmental arrest has been studied by Stockard in experiments upon the marine minnow, *Fundulus*. The exact type of deformity which results, depends upon the time at which interruption in growth occurs, not upon the specific mode of interruption. The method used is that of diminution of oxygen supply, or lowering of temperature, resulting in either case in a reduced metabolism, associated with the retarding of oxidations, and this is identified as the cause of all developmental anomalies.

The developmental rates of various parts of the embryo vary in relation to its growth as a whole, and the depression of metabolism of a particular group of cells during a phase of dominant proliferative activity will produce a permanent defect in the fully developed individual, owing to the failure to utilize its period of differential ascendancy.

In this connection the theory of causation of mongolism advanced by Van der Scheer is of interest. He suggests that mongolism is due to the abnormal pressure of an excessively rigid amniotic sac operating at the sixth and seventh week of embryonic growth.

A number of published cases have, in addition to anodontia, an analogous series of defects of ectodermal origin referable to the period around the sixth and seventh week of embryonic development. In Zilkens' case there was syndactyly of the second and third toes of the left foot, together with anomalies of hair and of nipples—the digits are first indicated in the seventh week, the milk-line making its appearance at the sixth week, with mammary thickening in the seventh week. In Kjaer's case of partial anodontia in a man of 25, no nails were present in hands or feet. The classical case of the so-called

"Russian poodle man"—Adrian Zeftichejew—appears to be another such case. He had only one canine in his maxilla, associated with excessive hirsutes of the body.

The interest of the present case is chiefly in the correlation of the rare anomaly of complete anodontia with the commoner more generalized defect of ectodermal development of the mongol referable to the same period in ontogenic development.

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