# ORAL DISORDERS ASSOCIATED WITH OCULAR DISEASE. II

## Disorders affecting Dentition

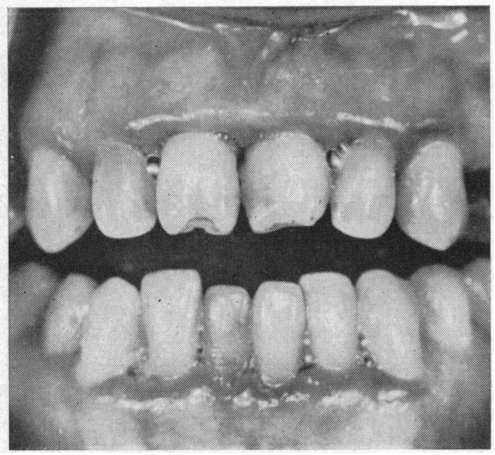

FIG. 1.—Hutchinson's teeth in congenital syphilis.

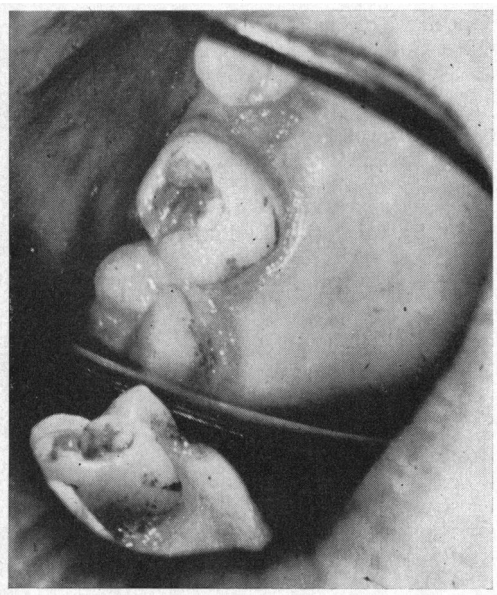

FIG. 2.—Moon's molars in congenital syphilis.

### Congenital Syphilis

*Oral Manifestations:* Angular stomatitis is common in the infectious infantile stage of the disease and may cause permanent scarring of the lips (rhagades). High arching and late gummatous perforation of the hard palate result from bone infection, while typical changes occur in the permanent dentition. The upper central incisors are stunted and peg-shaped, being narrowest at the incisal margin which is notched (Hutchinson's teeth) (Fig. 1). The first molars sometimes have maldeveloped cusps and a mulberry-like surface (Moon's molars) (Fig. 2).

*Ocular Lesions:* In infancy an infectious conjunctivitis is commonly associated with syphilitic rhinitis or "snuffles", while chronic dacryoadenitis and dacryocystitis occur rarely. Chorioretinitis is common at this stage, producing either the "pepper and salt" fundus appearance or disseminated scarring. Acute iridocyclitis is rare in infancy but commonly occurs in association with interstitial keratitis (IK). IK is the most typical ocular lesion and occurs in the post-infectious stage of the disease, most frequently between the sixth and twentieth years of life. The association of Hutchinson's teeth, IK, and labyrinthine deafness is termed Hutchinson's triad.

Basal meningitis in infancy, and syphilitic vascular disease or juvenile tabo-paresis in the later non-infectious stage, may cause optic atrophy, extra-ocular muscle palsies, and anomalous pupillary reactions. In these conditions the pupils are usually dilated and fixed, rather than being of the Argyll Robertson type.

**Rieger's Syndrome**

This dominantly transmitted hereditary disorder is characterized by mesodermal dysgenesis of the anterior segment of the eye and abnormal dentition, associated in some cases with skeletal and neurological abnormalities.

*Oral Manifestations:* There is partial suppression of dentition (anodontia) and those teeth which do erupt are peg-shaped, with hypoplastic enamel (Fig. 3). Premaxillary underdevelopment causes relative prognathia, and high arching of the palate sometimes occurs.

*Ocular Lesions:* The principal features are a posterior embryotoxon, abnormal mesodermal strands spanning the drainage angle, and hypoplasia or aplasia of the anterior stromal leaf of the iris, causing slit-pupil, corectopia, or pseudo-polycoria. These lesions are non-progressive, but juvenile glaucoma frequently occurs in the second or third decades of life.

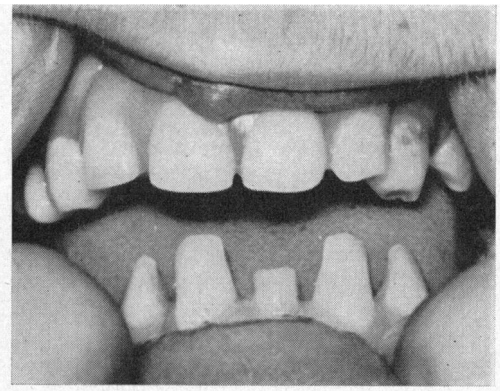

FIG. 3.—Rieger's syndrome.

**Van der Hoeve's Syndrome**

This dominantly transmitted hereditary disorder of mesodermal tissue is characterized in its complete form by fragile bones, blue sclerae and otosclerosis. Expressivity of the gene is variable so that only a quarter of cases exhibit all three features.

*Oral Manifestations:* Impaired dentine formation results in small teeth with a tendency to early wearing down, and with a characteristic pinkish opalescence (Fig. 4).

*Ocular Lesions:* The thin sclera composed of immature collagen is more translucent than normal so that the underlying uveal pigment becomes visible, resulting in the blue appearance of the sclera. Keratoconus and megalocornea are rare associated features.

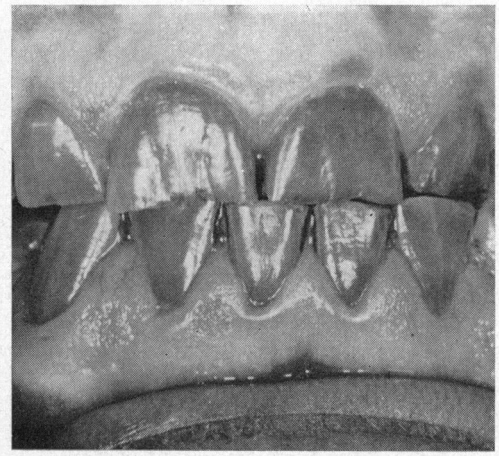

FIG. 4.—Van der Hoeve's syndrome.

*Illustrations:*
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