Dental Abnormalities in Congenital Ichthyoses: Case Report and Review of the Literature

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Background/Objective: Ichthyoses are disorders of cornification characterized by “fish-like” scaling of skin due to abnormal differentiation and desquamation resulting in a defective cutaneous barrier. Herein, we describe an infant with keratitis-ichthyosis-deafness (KID) syndrome and natal teeth and investigate spectrum of dental abnormalities, associated ichthyoses, and implications for long-term oral health.

Methods: A literature review of ichthyoses and dental abnormalities was conducted using Ovid MEDline (1946-2024) and EMBASE (1974-2024). Inclusion criteria were English-language articles that reported patient(s) with a variant of ichthyosis that manifested a dental abnormality. Exclusion criteria were non-English articles, articles with overlapping ectodermal dysplasia, pachyonychia congenita, or erythrokeratodermas, and articles that were unobtainable. Sex, age, country of origin, type of ichthyosis, dental findings, and genetic mutation were recorded when available. Ichthyoses were grouped into nonsyndromic and syndromic forms. Dental findings were categorized to reflect the type of anomaly: structure, eruption, number, and caries.

Results: Ninety-four articles included in our review described a total of 154 patients, with 74 (48.1%) males, 65 (42.2%) females and 15 (9.7%) patients with unspecified sex. Across all ichthyoses, the two most common dental anomalies reported were caries (51.3%) and structural abnormalities (40.9%), which include defects in shape, size, enamel, and color of teeth. Sjogren-Larsson, KID syndrome, trichothiodystrophy (TTD) and autosomal recessive congenital ichthyosis (ARCI) were most associated with dental anomalies.

Conclusion and Implications: We highlight the need for careful oral examination and dental evaluation of patients with ichthyosis. In rare cases of natal teeth, more emergent evaluation is warranted because natal teeth exhibit excess mobility and pose a risk for aspiration. Early referral to pediatric dentistry can help reinforce oral health habits, education, and infection prevention. Long-term follow-up can help limit extractions and improve oral health.