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RESEARCH ARTICLE

SUPERNUMERARY TEETH AND SYNDROMES ASSOCIATED

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ABSTRACT

Hyperdontia, commonly referred to as supernumerary teeth, is a well-known dental anomaly in humans. It is defined as the presence of extra teeth exceeding the normal count in either primary or permanent dentition. The occurrence has been documented to range from 0.2 to 3%, with a higher frequency observed in males compared to females. The etiology of this condition is heterogeneous and highly variable, and a majority of cases are idiopathic in nature. However, the presence of multiple impacted or erupted supernumerary teeth is rare and often associated with specific genetic syndromes. Early detection of abnormalities allows us to implement appropriate patient management strategies and plays a vital role in making well-informed decisions concerning long-term medical care and treatments.

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INTRODUCTION

Supernumerary tooth results from the continued proliferation of the permanent or primary dental lamina to form a third tooth germ. It can be familial, syndrome-associated, or non-syndromic. A higher proportion of males are affected in comparison to females, with a ratio of about 2 to 1.4 (Takahashi, 2017) Maxillary anterior region is the most common area of occurrence of supernumerary. The prevalence of supernumerary is found to be 0.2–3% in both primary and permanent dentitions (Subasioglu, 2015). This anomaly exhibits etiological heterogeneity and displays a high degree of variability, manifesting differences in numbers, location, morphology, and relationship with adjacent teeth (Lubinsky, 2016; de Souza Batista, 2017). The presence of supernumerary teeth can give rise to a range of complications, including impaction, failure to erupt, rotation, or displacement of adjacent teeth, root resorption, crowding, malocclusion, fistulas, cystic formation, as well as delayed or abnormal root development of permanent teeth (Subasioglu, 2015). The presence of mesiodens, or extra teeth between two maxillary incisors, and paramolars or distomolars, additional teeth in the molar regions adjacent to or distal from the usual sequence of teeth, are common dental anomalies. Accessory supplemental teeth, on the other hand, are less typical in form, often smaller, and described as tuberculate or conical. (2,1). The majority of supernumerary teeth are of unknown cause, with some cases showing autosomal dominant non-syndromic transmission (OMIM 108700), autosomal recessive inheritance, or linkage to the X chromosome (2,1). Nevertheless, the occurrence of multiple impacted or erupted supernumerary teeth is uncommon and is often linked to certain genetic syndromes. According to what is found in the literature, eight very different entities, including cleidocranial dysplasia, Gardners syndrome, Rubinstein-Taybi syndrome, Trichorhinophalangeal syndrome,Nance-Hooran syndrome, Opitz G/BBB syndrome, oculofaciocardiodental syndrome, and Robinow syndrome, present ST as distinct features and are reviewed in this article. The primary objective of this review is to provide a comprehensive overview of eight clinical conditions that exhibit ST as a significant finding, each with its own distinct inheritance pattern. By doing so, it aims to promote interdisciplinary medical care that encompasses dental evaluation and facilitates the timely provision of genetic counseling.(5)

CLEIDOCRANIAL DYSPLASIA: Cleidocranial dysplasia is an uncommon congenital skeletal defect with a dominant pattern of inheritance (5); it has been linked to mutations in many genes. It has been proposed that the illness process may be connected to cytogenetic anomalies in chromosomes 8q22 and 6p21. Recent research has revealed that the disease's primary cause is a mutation in the core-binding alpha-1 (Cbfa1) gene. (6). The main areas affected are the skull, the clavicle, and the dentition. The clavicle may not be present at all, and patients have abnormal mobility. Fontanelles may remain open or show a delayed closing. The skull suture is open, and there are several wormian bones present.

Micrognathia is present, and the maxilla is underdeveloped. The primary dentition is long-retarded, and the permanent dentition is delayed. There are numerous non-erupted teeth. (8) Supernumerary teeth and other dental abnormalities are seen in up to 94% of CCD patients. The development of extra teeth in CCD may also be caused by non-genetic reasons, including environmental influences, modifier genes, copy number variations, and epigenetic factors. (9) The incidence of ST in patients with cleidocranial dysplasia is observed at almost 22% in the maxillary incisor region and 5% in the molar region. (10)

Syndromes	Supernumerary tooth associated with the syndrome
Cleidocranial Dysplasia	22% in the maxillary incisor region and 5% in the molar region (10)
GardnersSyndrome	Anterior and canines (11)
TrichoRhinophalangeal Syndrome	All teeth except incisors (3)
Nance-HooranSyndrome	Maxillary incisors (14)
Opitz G/BBB Syndrome	Anterior mandible(18)
Oculofaciocardiodental Syndrome	Permanent canines (22)

GARDNER'S SYNDROME: Gardner's syndrome, a variant of familial adenomatous polyposis, is an autosomal dominant genetic disease caused by mutations of the APC gene (10). The most common symptoms are multiple polyposis of the large intestine,osteomas of the bones, multiple epidermoid or sebaceous cysts of the skin, the occasional occurrence of desmoid tumors, and impacted supernumerary and permanent teeth (6). 11-27% of patients with Gardner syndrome reported to have supernumerary teeth seen mostly between teeth in the alveolar bone or attached to the follicle of an impacted tooth. The most common sites of occurrence are around the anterior and canines. (11)

TRICO-RHINO-PHALANGEAL SYNDROME: Trichorhinophalangeal syndrome, type I (TRPS1; OMIM 190350) is an autosomal dominant disease.(12) A distinctive facies featuring sparse scalp hair, a bulbous nose tip, a long, flat philtrum, a thin upper vermilion, and a projecting ear are among the skeletal abnormalities with cone-shaped phalangeal epiphyses.(12) In addition to the characteristic traits of the syndrome, there have been reports of dental anomalies, including microdontia, delayed tooth eruption, ST, and malocclusion.(13) The mandible and maxilla were affected in every patient with multiple ST, while the incisors were not always affected (3).

NANCE–HORAN SYNDROME: An X-linked syndrome known as Nance-Horan syndrome involves both oral malformations and ophthalmological findings.(12,13) On chromosome Xp22, mutations in the NHS gene are the cause of it.(16,17). Facial dysmorphism, eye and tooth abnormalities are some of the characteristics. The facial dysmorphia includes an extended face, a distinguished nose and nasal bridge,mandibular prognathism, enlarged ears with anteverted pinnaries, and increased folds. Eye abnormalities include bilateral congenital cataract (100%), microcornea (96%), and microphthalmia. In 93% of the cases, ocular abnormalities cause severe nystagmus (93%), and sometimes strabismus (16). Dental abnormalities encompass supernumerary maxillary incisors (mesiodens), diastema among the teeth, and screwdriver-blade-fashioned incisors.(13) It is most usually visible within the imperative maxillary area, such as in mesiodentes. Males have a tendency to be significantly affected. It is because of mutations in NHS (OMIM 302350), which regulate actin transforming and mobileular morphology (3).

OPITZ G/BBB SYNDROME Opitz: G/BBB syndrome is an unprecedented genetically heterogeneous condition, with each an autosomal dominant form (OMIM 145410) that's because of mutation in SPECC1L gene placed on chromosome 22q11.2,46,47 liable for the manufacturing of cytospin-A, which interacts with cytoskeletal factors and microtubule stabilization,48and X-linked form because of mutation in MID1 gene placed on chromosome Xp22.2,46,49 and produces the midline-1 protein liable for microtubule binding (5). A study on dental anomalies in Opitz G/BBB syndrome and cleft lip and palate associated with hypertelorism found that 33.33% of the sample group had supernumerary teeth in the anterior mandible (17).

OCULOFACIOCARDIODENTAL SYNDROME: The BCOR (BCL-6 interacting corepressor) gene mutations were detected during the genetic analysis of individuals diagnosed with OFCD (Oculofaciocardiodental syndrome). This X-linked condition is distinguished by ocular, facial, cardiac, and dental abnormalities, as outlined by Hayward in 1980. (18,19,20). The occurrence of this syndrome has solely been observed in females who are heterozygous. It is believed to have lethal effects on affected males, as there are no documented cases of males developing this condition (21). The BCOR gene (M# 300485) on chromosome Xp11.4 is associated with this syndrome, and it can be caused by deletions, substitutions, or splice site mutations that result in a frameshift leading to a premature stop codon (19,20,18). BCOR is expressed in a wide range of tissues and plays multiple important roles during early embryogenesis, including maintaining tissue homeostasis and gene silencing through epigenetic mechanisms. These diverse functions of BCOR contribute to the varied symptoms observed in individuals with OFCD (22).

The phenotypic spectrum of OFCD (MIM# 300166) encompasses various features, including microphthalmia, congenital cataracts, and cardiac manifestations such as atrial septal defect, ventricular septal defect, or mitral valve prolapse (23,21). Individuals with OFCD often exhibit distinct facial characteristics, such as facial elongation with a prominent nasal bridge and broad nasal tip, along with separation of the anterior cartilage(23,24). Dental abnormalities associated with OFCD include delayed tooth eruption and prolonged retention of primary teeth. However, the most distinctive diagnostic symptom for OFCD is radiculomegaly, also referred to as dental root gigantism (23,25,26). In some cases, the roots of canines (or occasionally incisors or lateral incisors) continue to develop until they reach the cortical plate of the orbit or mandible (26,27). According to Schulze *et al*. (1999), patient number three looked to have duplicated permanent canines.

CONCLUSION

The presence of supernumerary teeth can be linked to specific syndromes, as previously mentioned, or it can be non-syndromic. It is important to identify supernumerary teeth early, evaluate them radiologically, and treat them promptly if necessary. When supernumerary teeth are paired with other clinical characteristics, the dentist can use this information to identify syndromes early and refer patients for appropriate care.

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ABBREVATIONS

CCD-Cleidocranial dysplasia, NHS-Nance-Hooran syndrome, OFCD –oculofaciocardiodental, ST-supernumerary teeth

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