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ORAL MANIFESTATIONS IN CHILDREN WITH DOWN SYNDROME

(MANIFESTASI ORAL PADA PASIEN ANAK DENGAN DOWN SYNDROME)

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ABSTRACT

Down syndrome is an autosomal genetic disorder that can occur in both men and women, with an extra chromosome 21. This literature review aimed to determine the oral manifestations and management of patients with Down syndrome. The method used in this literature review involved reviewing relevant national and international journal articles related to Down syndrome and its oral manifestations, retrieved from databases such as PubMed, ScienceDirect, and Google Scholar. The results of this literature review showed that children with Down syndrome have several clinical features, such as muscle hypotonia, brachycephaly, flat nose, hypersalivation, and various comorbid systemic diseases, such as congenital heart defects. Oral manifestations included mouth breathing, open bite, macroglossia, delayed tooth eruption, dental agenesis, caries, periodontal disease, and dental anomalies. Behavior management in patients with Down syndrome is something that must be considered so that tooth treatment can be carried out optimally.

Keywords: behavior management; down syndrome; oral

manifestations

ABSTRAK

Sindrom Down adalah kelainan genetik autosomal yang dapat terjadi pada pria dan wanita, dengan kromosom ekstra 21. Tujuan dari tinjauan literatur ini adalah untuk menentukan manifestasi oral dan penanganan pasien dengan sindrom Down. Metode yang digunakan dalam tinjauan pustaka ini melibatkan penelaahan artikel jurnal nasional dan internasional yang relevan terkait sindrom Down dan manifestasi oralnya, yang diambil dari basis data seperti PubMed, ScienceDirect, dan Google Scholar. Hasil tinjauan literatur ini menunjukkan bahwa anak dengan sindrom Down memiliki beberapa ciri klinis, seperti hipotonia otot, brachycephaly, hidung rata, hipersalivasi, dan berbagai penyakit sistemik komorbid, seperti kelainan jantung bawaan. Manifestasi mulut termasuk pernapasan mulut, gigitan terbuka, makroklosia, letusan gigi tertunda, agenesis gigi, karies, penyakit periodontal, dan anomali gigi. Manajemen perilaku pada pasien dengan sindrom Down merupakan hal yang harus diperhatikan agar perawatan gigi dapat dilakukan secara optimal.

Kata kunci*: sindrom down; manajemen perilaku; manifestasi oral*

INTRODUCTION

Down syndrome is an autosomal genetic disorder that can occur in both men and women, with an excess of one chromosome 21. This excess chromosome causes abnormalities in chromosome development and changes in the genetic balance of the body, which causes changes in physical and mental characteristics, intellectual abilities, and impaired physiological function. 1 Down syndrome was first described by a British doctor, Langdon Down, in 1866. According to the WHO, the incidence of the birth of a child with Down syndrome is 1 in 1000. There are 3,000–5,000 children born with Down syndrome each year. The etiology of Down syndrome is not yet known. Still, it is suspected that this chromosomal disorder is caused by genetics, the age of the mother

and father when having children over 35 years of age, radiation, infection, and autoimmune disease. Children with trisomy 21 develop differently from normal children. Children with Down syndrome have delays and limitations in all areas of development, so they have difficulty maintaining their hygiene, especially oral hygiene, and tend to depend on both parents and siblings throughout their lives.[1,2]

Children with Down syndrome have relatively similar physical characteristics even though they are of different races. In general, these signs are mental retardation, smaller than normal brain volume, small head, flat nose resembling Mongolian people, concave facial profile, hypotonia of orofacial muscles, and some have congenital heart defects, eyes Brush field spots accompanied by peripheral iris hypoplasia, myopia, broad toes, and hands show simian crease. Oral manifestations of the oral cavity in people with Down syndrome are class III malocclusion, crossbite, and open bite: large fissured tongue, fissure-forming lips, and narrow palate. The maxilla in people with Down syndrome is underdeveloped, the mandible is normal or slightly hypoplastic, and dental anomalies, delayed tooth eruption, and caries occur.[3,4] Oral disease is a significant health problem for individuals with Down syndrome, which has a higher prevalence and severity compared to the general population. These dental and oral problems will be discussed individually in this literature review to understand better the oral manifestations in the oral cavity of pediatric patients with Down syndrome and their management.

METHOD

This literature review was conducted by adjusting the topics related to oral manifestations in children with Down syndrome and their management. A literature review was conducted in several journals that discussed Down syndrome and its oral manifestations using the keywords Down syndrome, trisomy 21, genetic disorders, behavioral management, oral manifestations in Down syndrome, and its management to be used as a source of relevant material, retrieved from databases such as PubMed, ScienceDirect, and Google Scholar.

RESULT

Down syndrome is a genetic disorder that often occurs because of chromosomal abnormalities in both number and structure. An aneuploid condition or the addition of several chromosomes in the genome, Down syndrome, is called a trisomy 21 disorder because, in the nucleus, there is one chromosome number with three homologs.

In contrast, the other number still contains two chromosomes. Normal individuals have 46 chromosomes, but in people with Down syndrome, the total number of chromosomes is 47 because there is an additional chromosome 21. The extra chromosome 21 causes the number of specific proteins to be excessive, disrupting the normal growth of the body and causing changes in brain development that have been previously arranged, resulting in changes in physical characteristics, such as specific physical signs and a lack of intellectual abilities. Most children with Down syndrome have delayed physical development, learning disabilities, heart disease, even leukemia, and also experience mental retardation.[5]

The mother's age during pregnancy is one of the factors that affects the birth of a child with Down syndrome because the higher the mother's age during pregnancy, the greater the risk of giving birth to a child with Down syndrome. When the mother is 20-24 years old, the risk of Down syndrome is 1:1,490. When the mother is 40 years old, the risk of Down syndrome is around 1:106, while when the mother is 49 years old, the risk of Down syndrome is around 1:11 births.[6]

Down syndrome is categorized into several types based on the structure and number of chromosomes: classic trisomy 21, translocation, and mosaic. The first category is the classic trisomy 21. Down syndrome occurs in 94% of patients with Down syndrome. In trisomy 21, there is an additional chromosome 21. The second type is translocation, which has a frequency of occurrence of approximately 4%. All or part of the extra chromosome 21 breaks away during cell division and attaches to other chromosomes. Chromosome 21 joins chromosomes 13, 14, 15, and 22. In the translocation type, Down syndrome can be passed down from the parents to their children. The third type is mosaic, which has a frequency of occurrence of 1% or less. In this type, only a few cells have an excess chromosome 21. People with Down syndrome with the mosaic type will have a clinical picture and milder health problems compared to babies born with classic trisomy 21 and translocation Down syndrome.[6,7]

The cause of Down syndrome is not yet known, but failure in cell nucleus division that occurs during fertilization is a common cause. Down syndrome occurs due to an abnormality in the arrangement of chromosome 21, an excess of one copy of chromosome 21 in the genome, which is part of the Robertsonian translocation fusion, namely, the fusion of chromosome 21 with another acrocentric chromosome. Chromosomes are paired up to a total of 46

in normal numbers. Still, in people with Down syndrome, chromosome 21 is 3, so it is called trisomy, and the total number of chromosomes in people with Down syndrome becomes 47 chromosomes.[7]

Another cause of Down syndrome is anaphase lag, the failure of chromosomes or chromatids to join one of the daughter nuclei formed during cell division, resulting in delayed transfer or movement during anaphase. Chromosomes that did not enter the daughter cell nuclei disappeared. This phase occurs during meiosis or mitosis.8 Another cause of Down syndrome is anaphase lag, the failure of chromosomes or chromatids to join one of the daughter nuclei formed during cell division, resulting in delayed transfer or movement during anaphase. Chromosomes that did not enter the daughter cell nuclei disappeared. This phase occurs during meiosis or mitosis.[8]

Children with Down syndrome can be recognized by several physical characteristics, such as a relatively smaller head shape compared to normal people (microcephaly) with a flat area at the nape, narrow eyes with the middle corners forming folds (epicanthal folds), a small mouth with a relatively large and fissured tongue (macroglossia) so that it looks protruding, smaller ear canals so that they are easily blocked and can cause hearing loss, straight/horizontal palm lines (simian crease), and decreased muscle tone (hypotonia). A flat nasal bridge (depressed nasal bridge) and small nostrils, so that the airway is smaller, cause children with Down syndrome to experience nasal congestion easily. Most children with Down syndrome do not reach the average adult height, so they have a short body, small chin (micrognathia), small teeth (microdontia), slower tooth eruption than normal people, and there are white spots in the iris of the eye (brushfield spots).[6,7,8]

Management of care for children with special needs, especially Down syndrome, needs to be considered by medical personnel. Care services for children with Down syndrome should not be differentiated from those of the general population. People with Down syndrome do have deficiencies in terms of development and mental retardation so that they are not treated the same as normal people, but in terms of health care, people with Down syndrome have the right to receive the same health care as other children.[9]

The limitations of children with special needs, such as the inability to clean themselves, including the oral cavity, will increase the risk factors for tooth decay and the surrounding soft tissue. One child with special needs had Down syndrome. The dental health of children with Down syndrome is generally poor. The most

common dental and oral health problems found in children with Down syndrome are tooth decay, periodontal disease, malocclusion, high palate, and dental anomalies.[10]

According to Ghaith et al., children with Down syndrome have fewer caries than people. In children with Down syndrome, salivary pH has a higher bicarbonate content and increases buffering, delayed eruption of deciduous and permanent teeth, loss of permanent teeth, and smaller than normal tooth size, causing a large enough space between the erupted teeth, making it easier to remove plaque.[11]

According to Ronald et al., some children with Down syndrome experience a higher increase in caries due to xerostomia and cariogenic foods. Hypotonia also increases the incidence of caries in DS children. Children with muscles that experience hypotonia have poor chewing habits, so that the food bolus is in the oral cavity for too long. This results in increased caries because the length of time the food is in the mouth without good chewing function makes the child wait for the food to be soft before swallowing the food.[12]

Periodontal disease is the most significant oral health problem among patients with Down syndrome. Periodontal diseases that often occur in children with Down syndrome are marginal gingivitis, gingival recession, periodontitis, and periodontal pocket formation. The occurrence of periodontal disease in patients with Down syndrome is caused by a damaged immune system rather than poor oral hygiene.[11,12]

In patients with Down syndrome, the immune system decreases, including nonspecific defense mechanisms and cellular and humoral immune systems. It is supported by reduced neutrophil chemotaxis, decreased phagocytic ability, and shorter neutrophil half-life in children with Down syndrome. The activity of polymorphonuclear leukocytes (PMN) against A. actinomycetemcomitans (AA) is reduced in individuals with Down syndrome. The abnormal number of PMNs in Down syndrome patients is included in the qualitative type, where there is a failure of bactericidal function and reduced bacterial neutrophil adhesion, damaged T cells, low levels of IgM immunoglobulin, and changes in the function of B cell lymphocytes.[11]

Malocclusion, which is often found in patients with Down syndrome, is an angle class III malocclusion. It is because the head in children with Down syndrome is brachycephalic. The round and wider head affected the cranial base. The cranial base is more upright with a smaller angle, so the

anteroposterior dimension of the middle cranial fossa is short, causing the maxilla to be located more posteriorly, and the horizontal length of the nasomaxillary complex is also relatively short. The result is a relatively retrusive nasomaxillary complex and a relatively more protrusive mandible.[7,11]

The craniofacial complex undergoes retrusion, and the profile of children with Down syndrome is relatively concave. The nose of children with Down syndrome tends to be flat, causing nostrils or small openings so that less air enters than normal people. It makes children with Down syndrome tend to breathe through the mouth. Buccinator contracts, causing the maxilla to become compressed and narrow, causing the palate to become high.[8]

Various dental anomalies, including tooth number, shape, structure, and position, are often found in patients with Down syndrome. Dental anomalies can occur in deciduous and permanent teeth. The most commonly observed dental anomalies are dental morphological anomalies, hypodontia, hypoplasia, delayed eruption, and supernumerary teeth.[13]

Dental anomalies often occur in the maxillary lateral incisors; these teeth have smaller crown sizes, are tapered, and are conical. Tooth agenesis is also a dental anomaly frequently found in patients with Down syndrome. The incidence in the mandible is more frequent than that in the maxilla, especially in the left mandible. Patients with taurodontism are also often diagnosed with Down syndrome. Teeth with taurodontism have abnormally short roots, which reduce the attachment of periodontal tissue and cause tooth mobility. Delayed eruptions in patients with Down syndrome have been reported in several studies. Patients with Down syndrome experience delayed tooth growth, which should have grown at 12–14 months but can be delayed until 24 months, and have complete teeth at 4-5 years of age. Likewise, permanent teeth in people with Down syndrome experience delayed growth, where the first molar teeth should have erupted at 6-7 years of age. People with Down syndrome are reported to have erupted at 8-9 years of age.[13]

The management approach in carrying out dental care on children with Down syndrome is slightly different from that of normal children, adjusted to the child's behavior. Children with Down syndrome generally have less controlled behavior; therefore, dentists must use an appropriate approach. According to the Frankl Behavior Rating Scale, children with special needs are included in the number one rating category, which is negative. Children refuse treatment clearly because

they have uncontrolled behavior, excessive anxiety, active or passive oppositional behavior, and even crying loudly. [14]

Management of dental care in Down syndrome patients who have a lot of caries in the study of Khandelwal, D. et al. reported that the restoration of a 7-year-old boy on teeth 54, 55, 74, and 84 using GIC and extraction of teeth 75 and 85 was carried out using general anesthesia. General anesthesia was used to reduce unwanted risks. It is because patients with Down syndrome have uncontrolled behavior, such as sticking out the tongue, unchecked head and body movements, choking, biting instruments inserted into the mouth, and closing the mouth suddenly. [15]

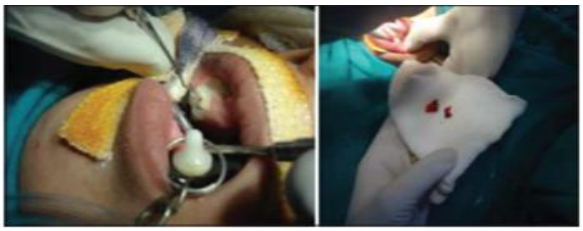
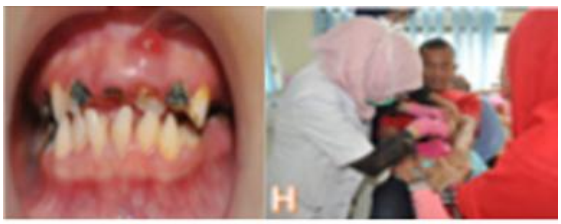


Figure 1. Restoration of teeth 54, 55, 74, 84 with GIC and Extraction of teeth 75 and 85 using general anesthesia.

In another journal, it was reported that the management of dental care uses behavioral management with a visual approach through photos of dental care, tooth models, and YouTube videos on dental care in children. Some images included pictures of dental care, tools used in dentistry, toothbrushes, and tooth models. A modeling approach was also reported in this study. Patients were invited to visit the clinic room to see friends of the same age when being treated and a storytelling show explaining the treatment that would be carried out. A non-pharmacological approach was also used in a 7-year-old patient with Down syndrome. The approach used was protective stabilization of physical restraint with parental consent to perform tooth extraction 61 due to pulp gangrene accompanied by decubitus ulcers. This approach involves having the child held by the parent, the parent holding the child's head, and the fellow operator restraining the child's movement. Several other people also held the patient to prevent unwanted movements and the risk of injury. Tooth extraction 61 was performed using topical anesthesia and chloroethyl. Extraction was performed in one movement, and the child did not cry. Teeth 51, 71, and 81 were also extracted in the same way as tooth 61 in stages.[15]



A B

C

Figure 2. A. Intraoral photo during occlusion. B. Non-pharmacological approach with Physical Restraint Technique. C. Intraoral photograph after extraction during occlusion.

Treatment of patients with Down syndrome with a narrow maxilla can be performed by installing RME. RME widens the maxilla, corrects malocclusion, reduces the number of infections in the superior respiratory tract, increases nasal permeability, improves tongue mobility, and improves aesthetics. In addition, rapid upper jaw expansion can provide a good airway for patients with DS. [16]

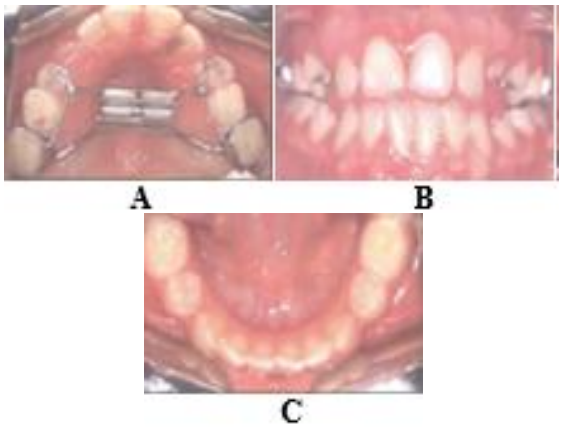


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Figure 3. A. Use of RME. B. patient at occlusion. C. Maxillary widening results after RME removal.

DISCUSSION

Down syndrome, also known as trisomy 21, is a collection of symptoms caused by chromosomal abnormalities that cannot be separated during meiosis, resulting in individuals with 47 chromosomes, whereas normal individuals have 46 chromosomes. Down syndrome is not a disease but rather a genetic disorder that causes various symptoms. Individuals with Down syndrome experience cognitive delays that usually have mild-to-moderate effects. People with Down syndrome have gross motor, fine motor, language, and social and personal delays. In addition, physically, people with Down syndrome experience muscle hypotonia, hypersalivation, and various accompanying systemic diseases such as heart defects. Oral and systemic manifestations in people with Down syndrome are something that needs to be known so that they can be handled properly as early as possible. Oral manifestations of patients with Down syndrome include mouth breathing, open bite, macroglossia tongue, delayed tooth eruption, chapped lips, tooth agenesis, caries, periodontal disease, and dental anomalies.[1,4]

Preventive care is prioritized by paying attention to oral hygiene, which is regularly checked. Behavior management in patients with Down syndrome is also something that must be considered so that treatment can be carried out optimally.

Cooperation among parents, dentists, and nurses is necessary to avoid unwanted risks. Patients with Down syndrome who have congenital heart disease must first be administered prophylactic antibiotics when tooth extraction and scaling are performed. Pulp treatment on deciduous teeth is contraindicated due to the risk of bacteremia, but it is still allowed if there is a good apical seal on permanent teeth.[6,7,8]

Patients with Down syndrome must be educated on good oral hygiene. Please consult with the family about what the child likes and dislikes and understand the level of communication for each action of the treatment process because it is important for the dentist to know detailed information about the patient to support the treatment process. Parental support is also important in dental care, including preventive measures, practices in maintaining oral hygiene, and motivation for a healthy diet. Patients with Down syndrome are advised to visit the dentist every 3 months or more often if needed.[9,11,12]

CONCLUSION

Down syndrome is a genetic disorder that often occurs due to abnormalities in chromosome 21. Oral manifestations that appear in people with Down syndrome include periodontal disease, macroglossia of the tongue, open bite, crossbite, mouth breathing, and dental anomalies such as microdontia, oligodontia, taurodontia, and enamel hypoplasia. The treatment of patients with Down syndrome requires good cooperation between dentists, parents, and nurses. Dentists must thoroughly understand the child's condition to provide good dental care.

CONFLICT OF INTEREST

The authors reported no potential conflict of interest.

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