Diagnosis and Treatment Guidelines for Down Syndrome in Dental Practice

2022

[Supervisor]
The Japanese Society for Disability and Oral Health (JSDH), Inc.

[Created by]
The Clinical Practice Guideline Committee of JSDH

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Message on Publication

While many subcommittees of the Japanese Medical Association and the Japanese Dental Association have published guidelines and manuals, our society has long sought an opportunity to contribute in this area. We recognized that standardization and equalization would be a difficult path, since many "disabilities" extend beyond ±1 SD of the normal distribution. However, Down syndrome one of the most frequently encountered conditions in both undergraduate and postgraduate education in special care dentistry, and one that presents characteristic features associated with chromosomal abnormalities represents essential knowledge for dental professionals. Dentists must be prepared to answer questions from parents and patients appropriately.

During my tenure as President, I asked Director Dr. Fumiyo Tamura to lead the development of these clinical practice guidelines. I am deeply grateful to the many dentists who devoted their time to reviewing this work. At the same time, I believe this effort has given our society a clear direction for promoting research.

I sincerely hope that these guidelines will be widely used by those involved in the care of individuals with Down syndrome. They may represent as small step within the dental field, but I am convinced they are a significant step toward improving the future of children and people with disabilities.

June 2022
Shouji Hironaka
President, FY2018~2021
The Japanese Society for Disability and Oral Health (JSDH)

Message on Publication

The Japanese Society for Disability and Dentistry, a public interest incorporated association, is pleased to announce the publication of the "2022 Guidelines for Diagnosis and Treatment in Dental Care for Down Syndrome." This document was completed through the efforts of the Society's Clinical Practice Guidelines Development Committee. Minds defines clinical practice guidelines as "documents that present optimal treatments based on evidence (scientific basis) and serve as important information for patients and healthcare professionals when making treatment decisions." Down syndrome presents numerous physical and mental characteristics, necessitating various considerations and judgments in dental care.

These guidelines, based on a detailed evaluation of numerous domestic and international papers, present current scientifically-based recommendations for dental treatment issues such as "dysphagia," "dental caries," "periodontal disease," "orthodontics," and "prosthetics" in Down syndrome. Dental care providers and healthcare recipients can use these guidelines as a reference for decision-making in clinical settings. We hope that these guidelines will be utilized to protect the oral health of individuals with Down syndrome and provide better medical care for them.

June 2022
Tadashi Ogasawara
President, FY2022~2023
The Japanese Society for Disability and Oral Health (JSDH)

Preface

Clinical practice guidelines are designed to support patients and healthcare providers as decision-making tools in clinical practice. They also aim to improve the quality of daily care by presenting the best available treatment methods based on up-to-date evidence. Because guidelines represent general practice, they are not always applicable to every individual case. In other words, guidelines serve as a compass for determining the most appropriate treatment for each patient.

The Committee for the Development of Clinical Practice Guidelines for Dentistry for Persons with Disabilities was established in 2018 in consultation with the then-President of the Japanese Society for Disability and Oral Health, Dr. Shoji Hironaka. In November of that year, the Committee convened its first meeting and began developing the present guideline. We first formulated clinical questions, and then invited highly specialized external members and a systematic review team to ensure the highest possible level of completeness. We had originally planned to hold a panel meeting with the cooperation of patients with Down syndrome and their families. However, this was not possible due to the ongoing COVID-19 pandemic. Instead, we requested the cooperation of the Japanese Down Syndrome Society and were able to receive valuable opinions through public comments, although regrettably not in person.

I would like to express my deepest gratitude to the Committee members, external experts, and collaborating academic colleagues for their cooperation over the past three and a half years. As chair, I went through a continuous process of trial and error, and although the work took time, we believe we have succeeded in producing a guideline that can be used by many. At the same time, research and treatment of Down syndrome continue to advance. We sincerely hope this guideline will be revised in the future to remain current and make it even more relevant.

July 2022
Fumiyo Tamura
Chairman of the Clinical Practice Guideline Committee of JSDH

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Method for Developing the Clinical Practice Guideline

This guideline was developed by the Clinical Practice Guideline Committee of the Japanese Society for Disability and Oral Health (JSDH), using the *Minds Clinical Practice Guideline Development Guide 2014* for processes up to the systematic review (SR) and the *Minds Clinical Practice Guideline Development Guide 2020* for processes after the SR, including recommendation/commentary development. Meetings were conducted both in person and online. First, five key clinical questions (CQs) were developed for Down syndrome, and work was divided into groups (feeding function group, caries management group, dentition development group, periodontal disease management group, and early aging prevention group). CQs were developed using the PICO framework to address critical clinical issues, and a panel meeting was used to decide on and adopt the CQs. Literature searches were conducted in MEDLINE (PubMed), the Cochrane Library, and Ichushi-Web (Japan Medical Abstracts Society database). Based on the SR results, the panel decided on the recommendations and prepared the commentary.

Public comments were solicited from and considered by the Japanese Society for Disability and Oral Health (JSDH), the Japan Down Syndrome Association, and the Japan Down Syndrome Society. This guideline was completed after deliberation and approval by the Board of Directors of the Japanese Society for Disability and Oral Health (JSDH).

Target Conditions

Down syndrome

Developing Organization

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Conflict of Interest

In an effort to ensure transparency and fairness, this guideline was developed without compensation. None of the Committee members report any conflicts of interest.

Funding

Funding was provided by the Japanese Society for Disability and Oral Health.

I Overview of the Down Syndrome

A History of Down syndrome

The oldest known record of Down syndrome is a clay figurine exhibiting features characteristic of the condition, discovered among the relics of the Tumaco-La Tolita culture in Mexico, and dating back to around 500 AD.[1)] The first documented description of Down syndrome was provided in 1838 by the French physician P. E. Esquirol, in a handbook in which he categorized individuals with distinctive facial features, body structure, and intellectual developmental disorder what would later be recognized as Down syndrome.[2-4)] In 1846, S. Edouard expanded upon P. E. Esquirol's description by adding features such as an open mouth, a protruding tongue, a small nose, and susceptibility to infections.[4)].

In 1866, in a three-page article in a medical journal, English physician J. Langdon Down first described the characteristics of Down syndrome, which he termed "Mongolian idiocy."[5)] Down's son, R. Langdon Down, also became a physician and described the "simian crease" (single transverse palmar crease), a finding often observed in individuals with Down syndrome.[3)]

In 1932, P. J. Waardenburg proposed that Down syndrome results from a chromosomal abnormality due to nondisjunction.[6)] In 1960, a translocation form of trisomy 21 was reported,[8)] and in 1961, mosaic trisomy 21 was described.[9)]

In 1961, *The Lancet* published a letter written by 19 prominent geneticists who suggested that the terms "Langdon-Down anomaly," "Down syndrome anomaly," "congenital acromicria," or "trisomy 21 anomaly" should be used instead of "Mongolian idiocy" [10)]. In 1965, a representative of the People's Republic of Mongolia unofficially requested that the World Health Organization (WHO) discontinue use of the term "mongolism" because it was offensive[11)]. In 1968, prenatal diagnosis of Down syndrome by amniocentesis was first reported.[12)]

In 2012, the United Nations General Assembly designated March 21 as World Down Syndrome Day; it has been observed since 2012 to promote awareness of Down syndrome and support for individuals with Down syndrome and their families.

(Tadashi Ogasawara)

B Patient Trends

Trisomy 21 most commonly results from meiotic nondisjunction; approximately 79.2% of cases are of maternal origin[1].The birth prevalence at maternal age 20 years is about 1 in 700–1,000. The frequency increases with maternal age: approximately 1/1,538 at 30 years, 1/840 at 35, 1/356 at 40, 1/94 at 45.[2] In Japan, social trends and the use of assisted reproductive technologies have increased pregnancy rates among older mothers, and the number of births of infants with Down syndrome recent is therefore expected to increase.[3] The annual number of births with Down syndrome in Japan from 2010 to 2016 is estimated to be around 2,200.[4] However, owing to prenatal diagnosis, the number of such births has not increased substantially.[4,5]

The average life expectancy of individuals with Down syndrome was less than 10 years until the 1950s.[6] However, with improvements in the treatment of complications such as congenital heart disease and congenital gastrointestinal disease, the average life expectancy has increased dramatically since the 1970s, and the current average life expectancy of individuals with Down syndrome is approximately 60 years.[6,7] The number of people with Down syndrome in Japan is estimated to be approximately 50,000. Although there are no reports on the number of patients by age group in Japan, it can be concluded that the population of people with Down syndrome is increasing, considering that the number of births has remained unchanged and the average life expectancy is long. In the United States, the number of births has been increasing since 1980, and the average life expectancy has been increasing, but the number of deaths of elderly people with Down syndrome has been increasing and estimated to be about 300,000 in 2008.[6]

(Tadashi Ogasawara)

C Etiology of Down syndrome

The Down syndrome phenotype results from an extra copy of all or part of the long arm 21q of human chromosome 21 (Hsa21).[1] Complete trisomy of Hsa21 occurs in ~95% of cases, followed by translocation (~4%) and mosaicism (~1%).[2] Hsa21 harbors more than 500 genes, including over 200 protein-coding genes, as well as microRNAs and other noncoding RNAs.[3] The majority of the genes on Hsa21 are still functionally unknown. Nevertheless, overexpression of these genes in trisomy disrupts the transcriptome (the set of all RNA transcripts present in a cell under given conditions) and the proteome (the complete set of proteins expressed in a cell, tissue, or state), affecting the development and function of multiple organs.[1] In the aging Down syndrome brain, marked cellular and transcriptomic changes have been described.[4] Because particular clinical features occur only in some individuals and vary in severity from case to case, the phenotype is thought to reflect the additive and interactive effects of many genes rather than a single Hsa21 gene.[1,11] Furthermore, the degree of intellectual developmental disorder varies from mild to severe, suggesting contributions from environmental and epigenetic factors in addition to genetic factors.[5,6]

(Junpei Murakami)

D Major Associated Conditions

1. Dementia

Dementia is currently a major cause of death in people with Down syndrome,[1] and the risk of developing dementia is estimated to be more than 90%.[2,3] Dementia develops in ~40% of individuals with Down syndrome by ages 55–60 years.[4]

2. Congenital heart disease (CHD)

CHD present in 54–66% of infants with Down syndrome[5-7] and increases the risk of pulmonary hypertension and death.[8] CHD has been reported to be more common in females than males with Down syndrome.[9-11] The recent increase in life expectancy in individuals with Down syndrome is largely attributable to the successful early treatment of CHD.[12] In a cohort of infants with Down syndrome with CHD, atrioventricular septal defects(AVSD), ventricular septal defects, and atrial septal defects were identified (rates reported in Allen[6]). Neonatal persistent pulmonary hypertension was also found in 5–34% of infants with Down syndrome,[7,12,13] and 44.8% of pulmonary hypertension cases in children with Down syndrome were attributed to CHD.[8]

3. Neurological Complications

(a) Epilepsy: Epilepsy is one of the most frequent neurological complications in Down syndrome, with an estimated prevalence of up to 13%.[14] The prevalence distribution is age-dependent and bimodal (infancy and adulthood).[15] The prevalence of infantile temporal epilepsy is 2–13%,[16] and the risk of later-life epilepsy has been reported to be low.[17] Infantile spasms/ West syndrome occur in 2–5%.[18] Up to 75% of adults with Down syndrome who have dementia develop late-onset myoclonic epilepsy[19], often beginning after ages 40–45 years, and this has been associated with Alzheimer's disease[19].

(b) Developmental disabilities: Autism spectrum disorder occurs in 10–18% of people with Down syndrome, and has been associated with attention-deficit/hyperactivity disorder (ADHD).[20]

(c) Moyamoya disease: Prevalence is up to 26-fold higher in Down syndrome than in the general population.[21]

4. Gastrointestinal (GI) Complications

(a) Structural problems: The prevalence of congenital GI abnormalities in Down syndrome is 3–13%.[22] ① Duodenal atresia (DA) is most common, with an estimated prevalence of 1–5%.[23] About 25% of infants with DA have Down syndrome.[24] ② Anal stenosis/occlusion occurs in 1–4%. ③ Esophageal atresia and tracheo-esophageal fistula occur in 0.3–0.8%, which is higher than in the general population.[23] ④ Hirschsprung disease presents with severe constipation, abdominal distention, acute obstruction, and neonatal fecal impaction; delayed transit occurs in 1–3% of infants with Down syndrome. Down syndrome accounts for ~5% of all Hirschsprung's disease.[25]

(b) Functional problems: ① Constipation is common in Down syndrome across childhood and adulthood. Possible contributors include hypotonia, poor diet,

inactivity, and hypothyroidism. In adults, 19% have severe constipation of unknown cause, and latent constipation is thought to be common.[26,27] ② Gastroesophageal reflux disease is associated with feeding difficulties and may be influenced by trunk strain in infants with Down syndrome. ③ Celiac disease prevalence is estimated at 5–7.5%.[28]

5. Endocrine and growth complications

(a) Hypothyroidism: Occurs in ~30% of adults with Down syndrome.[29,30] Symptoms include bradycardia, weight gain, fatigue, constipation, edema, impaired growth/cognition; hypothyroidism is a risk factor for dyslipidemia (hyperlipidemia), osteoporosis,[31] and mood disorders, and hormone replacement therapy is used. The incidence of congenital thyroid diseases is 26-fold higher than in the general population.[32] ① Congenital hypothyroidism (cretinism) occurs in ~1% of children with Down syndrome. ② Acquired hypothyroidism (e.g., Hashimoto disease) increases with age. ③ Subclinical/asymptomatic hypothyroidism occurs in 25–60% of children with Down syndrome and is often temporary.[33,34]

(b) Hyperthyroidism (e.g., Graves' disease): The prevalence of hyperthyroidism in Down syndrome is 0.65–3%.[33] Symptoms include tachycardia, weight loss, restlessness, diarrhea, palpitations, and tremor; risks include thyroid nodules, atrial fibrillation, osteoporosis, and mood/anxiety disorders.

(c) Type 1 diabetes mellitus: Type 1 diabetes mellitus is more common in Down syndrome,[35] especially in young children (<3 years).[36] A Danish study reported that a 4-fold increase versus the general population, with a peak incidence at the age of 8 years, (versus 14 years in the general population).[37]

(d) Growth: Height is more than two standard deviations below population norms, indicating slower growth.[38]

6. Respiratory complications

Complications of Down syndrome include immune dysfunction, structural abnormalities and congenital abnormalities. ① Airway infections account for 42% of hospitalizations in children with Down and are the second leading cause of death after CHD [39]. ② Acute tracheobronchitis occurs in up to 20% of cases [40] ③ Tracheomalacia and bronchiomalacia cause breathing and wheezing similar to asthma and croup.[41] ④ Subglottic stenosis is also common.[42] ⑤ Aspiration was observed in 56.3% of patients, and 90.2% of them were reported to be silent.[43]

7. Otolaryngological complications

(a) Hearing loss: Approximately 40% of children with Down syndrome have hearing impairment due to middle ear disease or other causes,[44] but the number increases with age, and it is reported that all children with Down syndrome have hearing impairment by age 60.[45] The proportion of people with Down syndrome with hearing impairment is higher than that of other populations.[46,47] Causes include inner ear malformations, temporal bone dysplasia, otitis media and pearls.[48-50] Hearing aids and cochlear implants are used from childhood.[51,52]

(b) Chronic rhinitis: It has been reported that chronic rhinitis is common in Down syndrome.[53]

(c) Sleep-disordered breathing: In children with Down syndrome, symptoms of sleep-disordered breathing, such as snoring, abnormal sleep posture, frequent awakenings, and daytime sleepiness,[54] are present in 20–80% of cases.[55,56]

8. Hematologic and oncologic complications

In neonates with Down syndrome, ① thrombocytopenia,② polycythemia,[57] and ③ transient abnormal myelopoiesis (TAM) may be observed. The prevalence of TAM is reported to be 10–15%,[58] and its clinical features include hepatosplenomegaly, bleeding, petechiae, pericardial effusion, pleural effusion, and skin rashes. Although 80% of cases resolve spontaneously, 20% progress to ④ Acute myeloid leukemia (AML).[59,60] Children with Down syndrome have a 150-fold increased risk of developing AML before the age of 5.[61] Prior to the onset of AML, myelodysplastic changes, GATA1 mutations, and acute megakaryoblastic leukemia may be observed; recent studies suggest a preleukemic phase initiated by trisomy 21, with subsequent progression to leukemia occurring occur independently of the trisomy.[62] The risk of ⑤ acute lymphoblastic leukemia (ALL) is also ~20-fold higher. While the onset patterns of ALL in Down syndrome do not differ significantly from those in the general population, children with Down syndrome tend to have poorer outcomes.[60]

9. Immune-function complications

The risk of infection is high for individuals with Down syndrome, for hospitalization and severe respiratory infection. The risk of death from infection is ~12-fold higher,[63] and the mortality rate from sepsis is more than 30%.[64]

(a) Acquired (adaptive) immune dysfunction: T- and B-cell numbers are decreased from infancy.[65] The thymus is small, and naïve/regulatory T cells are reduced.[66] Selective B-cell depletion is also observed.[67]

(b) Innate immune dysfunction: Abnormal neutrophil chemotaxis and phagocytosis have been reported.[68,69] Dysregulation of inflammatory cytokines is also observed,[70,71] and overexpression of TNF has been reported in most cases of Down syndrome,[72] suggesting effects on the immune system.[67]

10. Musculoskeletal complications

(a) The prevalence of arthropathy is 0.87% to 2%,[73] with joint swelling and symmetrical functional limitation of small and large joints; proximal interphalangeal joints, wrists, and metacarpophalangeal joints are commonly affected[74]. In addition, it has been reported that 20% of patients with Down syndrome have joint laxity and hypermobility [75].

(b) Foot deformities: ① Flat feet (70-90%),[76] ② Hallux valgus, ③ Syndactyly, etc. Other conditions include ④ scoliosis (0– 5%),[76] ⑤ dislocation due to hip instability (1– 7%),[77] and ⑥ foot deformity.

(c) Atlantoaxial instability: The prevalence of Down syndrome in adults is 2–20%, which is slightly lower than that of Down syndrome in children (15–20%), but higher than that of controls of the same age group.[45] There is excessive movement and ligamentous laxity between the C1 and C2 , which puts the patient at risk for spinal cord compression and death.[78] Symptoms are usually caused by compressive myelopathy, resulting in decreased muscle strength

and tone, hyperreflexia, and gait instability.[79,80] When secondary to severe cervical myelopathy, bowel and bladder symptoms are also present.[81]

11. Ophthalmologic Complications

The prevalence of ophthalmologic diseases in Down syndrome is 38% at less than 1 year of age, but increases to 80% at 5–12 years of age,[82] and the risk increases with age. The most common ophthalmic diseases include refractive error, astigmatism, strabismus, visual acuity loss, cataracts, blepharitis and nystagmus.[83]

12. Skin disease

Seborrheic dermatitis[84] is present in 36% of individuals with Down syndrome. Malassezia folliculitis (a type of dermatomycosis) is also more common. The prevalence of alopecia areata ranges from 1 to 11%.[85]

13. Obesity

It has been reported that 51.6% of men with Down syndrome and 40% of women with Down syndrome were classified as overweight/obese according to BMI.[86]

14. Osteoporosis

Adults with Down syndrome have low bone mineral density levels, with a particularly large decrease around the age of 40.[87] At the age of 40–49 years, the prevalence of osteopenia is 17% in women and 38% in men, and the prevalence of osteoporosis is reported to be 17% in women and 25% in men.[87]

15. Renal disease

Four percent of children with Down syndrome have congenital abnormalities of the kidneys and urinary tract.[88] Obstructive urinary tract disease is common, and other conditions such as posterior urethral valve, giant ureter, renal dysplasia, renal malformation (e.g., horseshoe kidney) suburethral cleft, and undescended testis are also seen.[89]

(Junpei Murakami)

E Rehabilitation (total care)

The average life expectancy of an individual with Down syndrome has increased significantly, and is now approximately 60 years.[1,2] This dramatic increase is largely due to improvements in medical care, education, community, family support, and other factors, as well as improvements in treatment techniques for congenital heart disease and other diseases. To support healthy lifestyles across the life span, preventive health care should, from early childhood, take into account expressive-language difficulties, motor delays, possible intellectual developmental disorder, and reduced executive functioning, so that children can spend meaningful time in the community and remain physically active.

In addition, appropriate medical management of associated conditions in Down syndrome is important. From a medical management, the life span is commonly divided into three stages: childhood (to 18 years), adulthood (19–40 years), and adulthood (41 years of older).[3] Because the major complications differ at each stage, seamless follow-up from birth through adulthood and into older age is necessary. The management of these conditions often spans multiple departments and specialties, which can lead to fragmented care and delays or missed evaluations and follow-up. To avoid this, effective care coordination is needed to provide comprehensive, seamless care.[4]

In infancy, respiratory and circulatory functions are immature, and early evaluation and treatment of various physical complications are prioritized. Physical therapy to promote gross motor development begins in infancy, with consideration given to the general condition, and in early childhood, the transition from physical therapy to occupational therapy and speech and language therapy is made. From this period, children begin to participate in group activities at rehabilitation facilities, where they are exposed to a variety of stimuli through activities with their peers, which promotes development in a wide range of areas, including physical, mental, and social aspects. In particular, it is important to nurture the child's desire to communicate with others while promoting the development of pronunciation and articulation, so that spoken language becomes a good means of communication.

From school age to adolescence, health and physical fitness are stable, and medical care may be less involved, but continuous follow-up of congenital diseases and attention to new diseases and conditions are also necessary. Schools include regular schools, special-needs classes, and special-needs schools, as well as home-visit education and in-hospital education, depending on the degree of medical care required.

In adulthood, behavioral and emotional problems may arise when individuals have difficulty adjusting to rapid changes after graduating from high school; attention is warranted. Down syndrome regression disorder—with psychomotor slowing, reduced conversation, apathy/indifference, weight loss due to decreased appetite, social withdrawal, and sleep disturbance—may be observed. New comorbidities often emerge and require medical management. Transitional care to adulthood involves more than transfer to an adult department; it also includes support for patient and family understanding of the condition, support for patient independence and autonomy, and adaptation for adult-care systems. Active collaboration and a seamless care system should be established to ensure appropriate medical care in adulthood.[5] Furthermore, pediatricians should remain available for consultation and back-up after transfer, and multidisciplinary

cooperation is important. For Down syndrome, in which the natural history has been well described, it is important to evaluate and treat complications longitudinally and to provide information on interpersonal and social characteristics, as well as on the natural history of the syndrome.

It is important to provide holistic support, including rehabilitation support that fosters social skills and welfare support that addresses the social environment.

(Tomoko Komatsu)

f Social welfare resources (systems)

1. Health Care System

Medical Expense Subsidy for Children with Specified Chronic Diseases: In order to reduce the burden of medical expenses of families with children suffering from chronic diseases of children from the viewpoint of sound upbringing, a portion of the out-of-pocket expenses for medical expenses is subsidized.

2. Disability Certificate System

The Ryoiku Techo (Certificate for Persons with Intellectual Disabilities): A certificate issued by prefectural governors or mayors of designated cities to individuals with intellectual disabilities, aimed at providing consistent guidance and consultation and facilitating access to various support services.

The Physical Disability Certificate:

Issued to individuals with physical impairments to provide access to support, consultation, and welfare services.

Mental Health and Welfare Certificate:

Issued to individuals with mental disorders or epilepsy to facilitate access to services and support.

3. Allowances and Pension Schemes

Pensions, benefits, allowances, and other programs are available to reduce the burden and stabilize the lives of people with disabilities and their families.

Special Child Support Allowance: For guardians, etc. who are taking care of children under 20 years old with moderate or severe physical or mental disabilities.

Welfare Allowance for Children with Disabilities: For home residents under 20 years old who are severely disabled and require special care in their daily lives.

Special Disability Allowance: For home residents aged 20 and over who are severely disabled and require special nursing care in their daily lives.

Disability Basic Pension: Individuals meeting the criteria for disability on the designated date of certification may be eligible. In the case of congenital conditions, the pension can be granted from their 20th birthday upon application.

Support systems may differ by prefecture or municipality, and other welfare services may be provided.

It is advisable to conduct research on websites or utilize consultation services such as the following.

4. Consultation Services and Developmental Support

Hospitals: In addition to physicians, patients and families can consult with rehabilitation specialists such as physical therapists, occupational therapists, and speech-language pathologists, as well as nurses and medical social workers who specialize in discharge planning and home support.

Health and Welfare Center: At the social welfare counter of the municipal office of the city, ward, town or village of residence, the Health and Welfare Center provides comprehensive consultation services related to health, welfare and medical care, and coordinates support with other organizations as needed.

Patient/family associations: Support in daily life includes nursery schools, kindergartens, schools, family-life support, and patient/family associations. Family associations are not limited to local communities; but also include national NPOs that expand the circle of connections across regions.

Consultation support for children with disabilities: Provides detailed support through care management to address issues faced by children with disabilities and to help them access appropriate services.

Places where specialized support is provided: Child development support centers; after-school day services; visiting day-care centers; medical-type child development support centers; and residential facilities for children with disabilities (welfare-type and medical-type). In addition to supporting independence in daily life and providing functional training, these services also serve as important sources of information.

5. Act on Providing Comprehensive Support for the Daily Life and Life in Society of Persons with Disabilities Welfare Service for Persons with Disabilities

Payment for Services and Supports for Persons with Disabilities:

Nursing care benefits: home care, daily nursing care, short-stay, and support for residential care in facilities

Special payment for training: employment transition support/continuous support for employment services/employment retention support, and communal living assistance (group home)

Consultation Support：Planning Consultation Support Benefits

Community life support services: consultation support, transportation support, provision of equipment for functional training, and support for using the adult guardianship system.

(Tomoko Komatsu)

g Genetic Counseling

Although definitions vary, the Japan Medical Association's *Guidelines on Genetic Testing and Diagnosis in Medicine* define genetic counseling as "a process that helps individuals understand and adapt to the medical, psychological, and familial implications of the genetic contributions to disease."[1)] This process includes ① interpreting family and medical histories to assess risk of occurrence/recurrence, ② education about genetic principles, testing, management, prevention, resources, and

research, and ③ counseling to promote informed choice and adaptation to risks and circumstances. The report emphasizes that genetic counseling is not a one-way delivery of information or persuasion; rather, through dialogue it helps clients (patients, research participants, etc.) make autonomous, informed choices and therefore requires great care. It is desirable that physicians experienced in the relevant condition and professionals skilled in genetic counseling work together as a team. In Japan, two types of specialists provide genetic counseling: clinical geneticists (jointly certified by the Japan Society of Human Genetics and the Japanese Society of Genetic Counseling) and certified genetic counselors, who work in collaboration with clinical geneticists.

Genetic counseling should be provided at the appropriate time. Prenatal counseling may be needed before and during pregnancy. If the child has a sibling with Down syndrome, genetic counseling should be provided at the appropriate time, such as when planning for a subsequent child. Genetic counseling may also be provided at an older age, if desired. Genetic counseling is necessary for prenatal testing during pregnancy. Support for pregnant women undergoing prenatal testing should not be provided by obstetricians and gynecologists alone, but should be provided in collaboration with pediatricians, clinical genetic specialists, midwives, public health nurses, nurses, psychologists, certified genetic counselors, social workers, peer supporters, and other professionals in various fields. Prenatal testing is broadly classified into *diagnostic tests*, which can confirm a diagnosis based on the results, and *non-diagnostic tests*, which cannot confirm a diagnosis. The former are genetic tests that detect the presence or absence of chromosomal or genetic abnormalities, and include procedures such as chorionic villus sampling (CVS) (performed between 10 and 14 weeks of gestation) and amniocentesis (performed from 15 weeks of gestation onward). The latter include maternal serum marker testing (performed at 15–21 weeks), combined testing, Non-Invasive Prenatal Genetic Testing (NIPT), and fetal ultrasound. Non-diagnostic tests that estimate fetal genetic information are used for risk assessment and screening. A definitive test to determine the genetic information of the fetus is necessary to diagnose the presence or absence of chromosomal abnormalities. Since April 2013, NIPT has been performed at a limited number of hospitals.[2)] The Japanese Society of Obstetrics and Gynecology (JSGY) has issued *Guidelines for New Prenatal Genetic Testing Using Maternal Blood,*[3)] which states that "it is important to understand diseases, pathological conditions, and genotypes based on genetic changes as human diversity and to respect such diversity and uniqueness, and that new prenatal genetic testing using maternal blood should be conducted in a manner that is sufficiently comprehensive and appropriate for the patient." In addition to various health care system issues, there are ethical, legal, and social issues (Ethical Legal Social Issues; ELSI) related to the implementation of NIPT. In 2019, an expert committee on prenatal testing such as NIPT of the Science and Technology Subcommittee of the Health and Welfare Science Council began discussions on the appropriate nature of prenatal testing, the development of consultation and support systems for providing information to pregnant women, and the nature of collaboration with seamless pediatric care and welfare measures from the fetal period. In 2021, a report[4] was prepared on the basic concept of prenatal testing, including views on prenatal testing and support systems.

Prenatal testing and diagnostic techniques are advancing rapidly with the development of microarrays and next-generation sequencing, making it possible to plan appropriate perinatal management and care for fetuses and newborns. However, there are concerns that prenatal testing/diagnosis may be used to exclude the birth of people with certain conditions, and ethical issues regarding the sanctity of life have also been pointed out. Therefore, prenatal testing/diagnosis must be performed with great caution and in accordance with the *Guidelines for Genetic Testing and Diagnosis in Medical Care*.[1)] There are also issues related to disclosure at birth. If severe physical complications are present, pediatricians often lead medical care. At this stage, it is important to make an accurate diagnose of the child's condition and take appropriate measures, such as explaining the situation to the parents and performing chromosomal testing. Genetic counseling by a clinical geneticist or other qualified specialist is required before chromosomal testing.

It is recommended that genetic counseling be provided again after the results are obtained.

It is also desirable to establish an ongoing bereavement-care system for people who experience perinatal loss (fetal or neonatal death). In addition, psychological support—as part of comprehensive support across pregnancy, childbirth, and childcare—and consultation support by social workers and peer supporters are also important.[4)]

(Tomoko Komatsu)

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II Clinical Questions

A The strength of the total evidence and recommendations

The quality of evidence was rated as A (strong), B (medium), C (weak), or D (very weak). Meta-analyses of randomized controlled trials (RCTs) and RCTs were rated up to A (strong), and observational studies were rated up to C (weak).

Quality of Evidence

A (Strong): Strongly recommended to do
B (Medium): Recommended to do
C (weak): Not enough scientific evidence, but recommended to do / not to do
D (very weak): No convincing scientific evidence

The strength of the recommendation was determined by considering the balance between the benefits and harms of the quality of evidence, the values of the guideline development committee, the current state of clinical practice, and whether the recommendation can be covered by dental insurance. In this guideline, when the strength of the recommendation cannot be determined, it is assumed to be “none”.

Strength of Evidence Recommendations

1: Strong: Recommend to do / not to do
2: Weak: Suggest to do / not to do
3: None: Can't say either way
Recommendations were listed with the quality of evidence (A, B, C, D) and the strength of the recommendation (1, 2, 3).

B Terminology

This guideline is unified as follows.
Eating function: including dysphagia
Videofluorography (VF)
Videoendoscopy (VE)

21

CQ1	Is assessment of general developmental status useful in predicting the acquisition of feeding function in Down syndrome?

[Perspective]
Evaluation of general developmental status may be useful in predicting the acquisition of feeding function. (2C)

Background and Objectives

In Down syndrome, the general features include short stature, obesity, short neck, short limbs, low muscle tone and short fingers,[1] and feeding dysfunction is often present.[2] We examined the usefulness of first assessing general developmental status when providing support for feeding function.

Explanation

Only two case-study articles[3,4] reported on general development and the acquisition of feeding function. Both papers examined the relationship between tongue protrusion swallowing (abnormal swallowing habit) and gross motor control, and found that tongue protrusion swallowing (abnormal swallowing habit) tended to decrease with the development of gross motor skills after the sitting position. However, there was no verification of confounding factors such as comorbidities other than Down syndrome or intellectual disability, and general developmental status was only a predictive factor.

The Japanese Guide to Support for Breastfeeding and Weaning (2019 revised edition)[5] lists the following as guidelines for the start of weaning: "the child`s neck is firmly planted and the child can turn over," "the child can sit up for at least 5 seconds," "the child rarely pushes a spoon or other food out of the mouth with the tongue (weakening of the feeding reflex)"and "the child shows interest in food".

Although no studies have directly examined the relationship between the acquisition of feeding function and head–neck posture in Down syndrome, it has been suggested that being able to sit unsupported may indicate maturation of tongue movement during feeding. However, the ability to sit up does not determine the timing of weaning; rather, it relates to immaturity of tongue movement—often described as tongue-protrusion ("tongue-thrust") swallowing. The average birth weight of infants with Down syndrome is lower than that of Japanese infants overall, and the rate of NICU admission is high due to preterm delivery.[6]

Although there is no established guideline for weaning in low birth weight or preterm infants, it is recommended that weaning should start at the corrected age in months.[5] In the case of Down syndrome, weaning should begin at 5-6 months of corrected age, based on an assessment of individual feeding function and developmental status.

Thus, evaluation of general developmental status and corrected age in months are useful in predicting the acquisition of feeding function in Down syndrome.

22

Literature

1) Kubodera Y, Murakami J, Morisaki I. Chromosomal abnormalities. In Japanese Society of Dentistry for Persons with Disabilities, ed. Special Needs Dentistry Dentistry for the Disabled. 2nd ed. Tokyo: Ishiyaku Shuppan; 2017. 170-2.
2) Ikeda S. Down syndrome. Ikeda S., Kuroki Y. Supervision. Syndromes and diseases diagnosed by mouth. Tokyo: Japanese Society of Dentistry for Persons with Disabilities; 2012.138-9.
3) Nakamura T, Ayuzawa K, Ozawa H. The Relationship between Tongue Thrusting during Swallowing and Gross Motor, Cognitive and Linguistic Development in Children with Down Syndrome: A Pilot Study. Journal of the Japanese Society for Dysphagia Rehabilitation 2017;21:200-8.
4) Mizukami M, Tamura F, Matsuyama M, et al. Association between gross motor skills and abnormal oral habits related to feeding in children with Down syndrome.-Journal of the Japanese society for disability and oral health. 2015;36:17-24.
5) Ministry of Health, Labour and Welfare: Support Guide for Breastfeeding and Weaning (2019 Revision). https://www.mhlw.go.jp/content/11908000/000496257.pdf (referenced 2022-4-25)
6) Shiraishi H, Morioka K, Takahashi N, et al. Clinical study of complications of Down syndrome treated in our NICU. The Journal of clinical pediatrics, 2019;67(1-6):21-6.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
35,998 (Pubmed), 54 (Cochrane Reviews)

#1. Down syndrome and Humans
#2. #1 and feeding
#3. #2 and feeding function
#4. #3 and development
#5. #4 and gross motor
#6. #4 and oral motor
Papers considered useful: 0
Medical Journal Search Results
3,628 (medical journal)
#1. (Down syndrome/TH or Down syndrome/AL) and (Down syndrome/TH or 21 trisomy/AL) and (PT=excluding conference proceedings)
#2. #1 and ((feeding /TH or feeding /AL) and (swallowing /TH or swallowing /AL))
#3. #2 and (cervical /AL)
#4. #2 and (respiration /TH or respiration /AL)
#5. #2 and (gross motor /AL)
Papers considered useful: 2

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

（FEEDING FUNCTIONS GROUP）

CQ2	Does Down syndrome complicate feeding dysfunction?

[Perspective]
Down syndrome may be accompanied by feeding disorders, but there are individual differences. (2C)

Background and Objectives

The characteristics of feeding dysfunction in Down syndrome include features related to intellectual development disorder and to decreased muscle tone (hypotonia).[1] Potential associated problems include feeding-behavior difficulties, swallowing problems, and delayed acquisition of feeding skills, which are discussed below.

Explanation

Although many case reports describe successful feeding therapy in Down syndrome, there are no studies that evaluate the degree of feeding dysfunction at the group/cohort level.

There have been retrospective studies using medical records and studies examining feeding dysfunction in children with Down syndrome in a population of children with disabilities, but the only comparison with typically developing children was reported in a questionnaire-based analysis by Anil et al.[2]

A study of Indian children aged 2-7 years with Down syndrome and their typically developing counterparts showed that feeding dysfunction was more frequent in the children with Down syndrome, with more problems with solid food intake. The children were also found to have physical, functional, and emotional problems related to feeding function, which were attributed to low sensory-motor skills.

Several case-control studies have addressed complications of feeding dysfunction. Jackson et al.[3] reported oral-phase disorder and pharyngeal-phase disorders (e.g., aspiration or laryngeal intrusion) in 63.8% and 56.3% of the patients, respectively, based on videofluoroscopic (VF) swallow studies (VFSS) performed at a medical center (mean age 2.1 years). In a report by Stanley et al.,[4] VFSS was performed in 174 infants with Down syndrome aged 0-6 months who were referred for thorough evaluation of dysphagia; oral-phase and pharyngeal-phase disorders were present in 55%, and dysphagia severe enough to warrant consideration of discontinuing oral feeding was present in 39%. Jackson et al.[5] reported that laryngeal intrusion or aspiration, or both, were found in 31.9% of children aged 0-5 months and 51.3% of children aged 6-12 months in a combined VFSS and videoendoscopic swallow study (VESS) of Down syndrome in children under 1 year. All of these reports recommend clinical screening for dysphagia in all Down syndrome groups, but as they were all conducted in the USA, validation in Japan is a future issue.

Literature

1) Ishizaki, A., Hironaka, S. Down syndrome. Eiichi Saito and Koichiro Ueda, eds. The Japanese Society of Dysphagia Rehabilitation No.1.3rd ed. Tokyo: Ishiyaku Shuppan; 2016.341-3.
2) Anil MA, Shabnam S, Narayanan S. Feeding and swallowing difficulties in children with Down syndrome, J Intellect Disabil Res 2019;63:992-1014.
3) Jackson A, Maybee J, Moran MK, et al. Clinical Characteristics of Dysphagia in Children with Down Syndrome. Dysphagia 2016;31:663-71.
4) Stanley MA, Shepherd N, Duvall N, et al. Clinical identification of feeding and swallowing disorders in 0-6 month old infants with Down syndrome. Am J Med Genet A 2019 Feb;179(2):177-82.
5) Jackson A, Maybee J, Wolter-Warmerdam K, et Al. Associations between age, respiratory comorbidities, and dysphagia in infants with down syndrome. Pediatr Pulmonol 2019 Nov;54(11):1853-9.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
35,998 (Pubmed), 54 (Cochrane Reviews)
#1. Down syndrome and Humans
#2. #1 and dysphagia
#3. #1 and swallow disorder
#4. #1 and eating dysfunction
#5. #2 and #3
#6. #2 and #4
#7. #5 and #6
#8. #1 and choking
#9. #1 and without chewing
#10. #1 and chewing dysfunction
#11. #1 and aspiration
#12. #1 and feeding issue
Papers considered useful: 3

Medical Journal Search Results
3,628 (medical journal)
#1. (Down syndrome/TH or Down syndrome/AL) and (Down syndrome/TH or 21 trisomy/AL) and (PT=excluding conference proceedings)
#2. #1 and (dysphagia /TH or dysphagia /AL))
#3. #1 and ((@ dysphagia /TH and @ dysphagia /TH) or dysphagia /AL) #4. #1 and (tongue protrusion /AL)
#5. #1 and (choking /TH or choking /AL)) #6. #1 and (swallowing whole /AL)
#7. #1 and (mammalian reflex /AL)
Papers considered useful: 0

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(FEEDING FUNCTIONS GROUP)

CQ3	Is a comprehensive assessment useful for evaluating feeding function in individuals with Down syndrome?

[Perspective]
The usefulness of a comprehensive assessment is recognized; however, results may vary depending on the assessment method and the person's level of cooperation. (2C)

Background and Objectives

We will next discuss the usefulness of evaluating feeding function in Down syndrome, which generally requires external observation and instrumental screening followed by a thorough examination.

Explanation

Two case-control studies[1,2] reported detailed examination of feeding function, including videofluoroscopic (VF) and videoendoscopic (VE) evaluation of swallowing, in people with Down syndrome.

As described in detail in CQ2, a study in infants younger than 1 year found a high rate of feeding dysfunction, but it evaluated only associations with comorbidities (e.g., respiratory disease), and did assess the usefulness of a detailed examination specifically for Down syndrome. Tolerability of detailed examinations is not unique to Down syndrome and may vary by age, as some patients refuse testing because of foreign-body sensation, discomfort, or pain. In addition, because feeding function changes with age in Down syndrome,[3] that report alone cannot describe age-related change or longitudinal assessment. Importantly, detailed assessment of feeding function is not intended solely to detect dysphagia and decide to discontinue oral intake; rather, it helps in planning effective support by evaluating functions that cannot be determined by external observation or screening. In Japan, VF and VE are covered by insurance, reducing the economic burden. Therefore, detailed evaluation of feeding function can be used to guide comprehensive support strategies.

Evaluation of feeding function in patients with Down syndrome as well as other disorders is basically performed by external observation and screening, followed by a detailed examination if necessary. Although there are no reports on the usefulness of the basic external observation evaluation, evaluation with a measuring device[4,5] may be performed as a screening test. However, there are cases in which measuring instruments are difficult to use due to the degree of cooperation and oral morphological abnormalities, and future investigation is warranted. Another method is the cervical auscultation method, which is minimally invasive, using only a stethoscope, and can evaluate swallowing function without placing undue stress on the subject. This method can be applied regardless of age or degree of cooperation, as long as the surgeon is skilled, but there are no reports limited to Down syndrome, so the usefulness of this method is unknown.

Literature

1) Jackson A, Maybee J, Wolter-Warmerdam K, et.al: Associations between age, respiratory comorbidities, and dysphagia in infants with down syndrome. Pediatr Pulmonol 2019;54:1853-9.
2) Stanley, Shepherd N, Duvall N, et.al. Clinical identification of feeding and swallowing disorders in 0-6 month old infants with Down syndrome. Am J Med Genet A 2019 Feb;179(2):177-82.
3) Lazenby T. The impact of aging on eating, drinking, and swallowing function in people with Down syndrome. Dysphagia 2008;23:88-97.
4) Isoda T, Tamura F, Kikutani T, et al.: Development of lip closing function during taking food into the mouth in children with Down syndrome. IJOM 2019; 45: 45. 31-45.
5) Hashimoto M, Inogari K, Ito A, et al: Tongue pressure during swallowing in adults with Down syndrome and its relationship with palatal morphology. Japanese Society of stomatognathic Function 2014;21:50-51 (abstract)

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
35,998 (Pubmed), 54 (Cochrane Reviews)
#1. Down syndrome and Humans
#2. #1 and dysphagia
#3. #1 and videofluorography
#4. #1 and VFSS
#5. #2 and videoendoscopy
#6. #2 and FEES
#7. #2 and ultrasound examination
Papers considered useful: 2

Medical Journal Search Results
3,628 (medical journal)
#1. (Down syndrome/TH or Down syndrome/AL) and (Down syndrome/TH or 21 trisomy/AL) and (PT=excluding conference proceedings)
#2. #1 and ((@ dysphagia /TH and @ dysphagia /TH) or dysphagia /AL)
#3. #2 and (swallowing contrast studies /AL)
#4. #2 and (tongue pressure /TH or tongue pressure /AL))
#5. #1 and (lip pressure /TH or lip pressure /AL))
#6. #1 and (swallow contrast examination /AL)
#7. #1 and (Cervical auscultation /AL)
Papers considered useful: 0

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(FEEDING FUNCTIONS GROUP)

Column 1 Feeding dysfunction in Down syndrome

In recent years, it has been recognized that Down syndrome is commonly associated with feeding and swallowing dysfunction.[1-6]Signs may include tongue protrusion, dysphagia, impaired bolus preparation/chewing, and masticatory dysfunction.[1-12]In addition, problems related to tooth number and dentition anomalies and to sensory integration disorders have also been reported.[2,7,9-12)]

Down syndrome is usually diagnosed soon after birth, and in recent years intervention for feeding and swallowing disorders has been provided from infancy.[2-15)] However, insurance coverage for feeding and swallowing therapy was not introduced in Japan until 1995, and there have been no reports of large-scale or longitudinal intervention studies over on the effectiveness of feeding and swallowing therapy in children with developmental disorders, including Down syndrome.

Many reports describe successful feeding and swallowing training in Down syndrome, including improvement in tongue protrusion during swallowing[6,13,14)] and improvement in mastication.[15,16)] On the other hand, effectiveness may depend on the timing of therapy initiation[6)]; tongue protrusion may persist or take time to improve[7,9,13)]; and motor/physical functional training alone may be insufficient when sensory integration disorders are present.[12)]

The acquisition of feeding skills is related to gross motor development.[17)] As head and neck stability are achieved, the muscular activities required for chewing and swallowing become more efficient, allowing stable feeding; this is particularly important from the early to middle stages of eating, when oral processing must stabilize. In addition, achieving a sitting posture and trunk stability is important for chewing and grinding with the alveolar ridge and primary teeth.

Feeding-function therapy requires a comprehensive response that includes the feeding environment, diet, and functional training, and depends not only on the patient but also on parent/caregiver cooperation. Even if clinical intervention studies are difficult to conduct, cohort studies that examine these factors should be undertaken to evaluate the effectiveness of the therapy.

Literature

1) Ikeda M. Down syndrome. Syndromes and Diseases Diagnosed by the Mouth (supervised by Masakazu Ikeda and Yoshikazu Kuroki). Tokyo JOURNAL OF THE JAPANESE SOCIETY FOR DISABILITY AND ORAL HEALTH; 2012. 138-9.
2) Shinozaki M. Down syndrome and dysphagia, Masaru Tazumi and Yoshiharu Mukai (eds.) Pediatric feeding and swallowing rehabilitation 2nd edition, Tokyo: Ishiyaku Shuppan; 2006. 238-241.
3) Ishizaki A, Hironaka S. Down syndrome. Ingestion and Swallowing Rehabilitation, 3rd ed. Tokyo: Medical and Dental Publishing ; 2016. 341-3.
4) Jackson, Maybee J, Moran MK, et al. Clinical Characteristics of Dysphagia in Children with Down Syndrome. Dysphagia 2016 Oct;31(5):663-71.
5) Stanley MA, Shepherd N, Duvall N, et al. Clinical identification of feeding and swallowing disorders in 0-6 month old infants with Down syndrome Am J Med Genet A 2019;179(2):177-82.
6) Jackson A, Maybee J, Wolter-Warmerdam K, et al. Associations between age, respiratory comorbidities, and dysphagia in infants with down syndrome.

Pediatr Pulmonol 2019;54:1853-9.
7) Mizukami M, Tamura F, Matsuyama M, et al. Relationship between gross motor function and oral parafunction habits relevant to eating in children with Down's syndrome. Journal of the Japanese society for Disability and oral health 2015;36:17-24.
8) Faulks D, Collado V, Mazille MN, et al. Masticatory dysfunction in persons with Down syndrome.
Part 1: a etiology and incidence. J Oral Rehabilitation 2008;35:854-86.
9) Nakajima R, Fujita H, Asahi R. Developmental Process of Oral Movement, Gross Motor Skills and Self-Feeding Motivation of Two Infants with Down Syndrome. the Japanese Journal of Dysphagia Rehabilitation 2012;16:290-8.
10) Nakamura T, Ayuzawa K, Ozawa H. The relationship between tongue thrusting during swallowing and gross motor,cognitive and linguistic development in children with down syndrome: a pilot study. the Japanese Journal of Dysphagia Rehabilitation. 2017;21:200-8.
11) yamazaki A, Fujiwara T, Nawa H, et al. Clinical statistical survey of patients with Down syndrome at the Orthodontic School of Aichi Gakuin University Dental Hospital. Orthodontic Waves-Japanese Edition. 2018; 77: 78-85.
12) Okubo M, Yamamoto M, Sugiyama T, et al. Sensory Processing Issues in Down Syndrome with Feeding Problems. the Japanese Journal of Dysphagia Rehabilitation. 2018;22:145-52.
13) Takahashi M, Tomita K, Hironaka S, et al. A Retrospective Study of the Feeding and Swallowing Function in Children with Down Syndrome ― A Review of Support Methods in Area Development and Disability Centers ―. the Japanese Journal of Dysphagia Rehabilitation. 2015;19:165-71.
14) Sasaki Y, Kamasaki Y, Hidaka K, et al. Promotion of growth and development in a Down syndrome infant with complications, Pediatrics International, 2010; Pediatrics International, 2010; 52: 653-656.
15) Faulks D, Collado V, Mazille MN, et al. Masticatory dysfunction in persons with Down syndrome: Part 2: management, J Oral Rehabilitation 2008;35:863-869.
16) Hayashi S, Endoh M, Saegusa Y, et al. A New Training Method for Masticatory Dysfunction in a Special Needs Patient, the Japanese Journal of Dysphagia Rehabilitation, 24: 56-63, 2020.
17) Mizukami M, Kikutani T ,Matsuyama M, et al. Investigating the fact estimating factors related t elated to the acquisition of masticatory function in Down syndrome children, International Journal of Orofacial Myology, 2019; 45(1):46-56.

(FEEDING FUNCTIONS GROUP)

CQ4	Are primary teeth in children with Down syndrome less susceptible to dental caries than those in typically developing children?

[Perspective]
We found no evidence that primary teeth in children with Down syndrome are less susceptible to dental caries than those of children with disabilities unrelated to Down syndrome or typically developing children. (3D)

Background and Objectives

Children with Down syndrome are more difficult to treat than typically developing children. Therefore, assessment for caries morbidity is of high importance. The aim of this study was to investigate the susceptibility of Down syndrome to dental caries in primary teeth.

Explanation

Sixty-four articles on dental caries in individuals with Down syndrome were identified from PubMed and medical journals. Of these, five were eligible for the SR: two reported primary-dentition tooth counts and three reported permanent-dentition tooth counts (one article reported both). Many studies began from the premise that caries prevalence is low in Down syndrome and examined associations between the number of carious teeth and oral hygiene practices, levels of *Streptococcus mutans* (*S. mutans*) and *Lactobacillus spp*.,[1)] and salivary secretory immunoglobulin A (s-IgA) concentration.[2)] However, two reports found no significant difference in the number of carious primary teeth compared with typically developing children. Maclaurin et al.[3)] likewise reported no significant difference in the number of carious primary teeth between children with Down syndrome and typically developing children. Only the reports of Lee et al.,[4)] who investigated salivary S. mutans–specific antibodies, showed fewer carious primary teeth; however, that study included only children with higher intellectual levels and differed in background factors. Considering the results from tooth-surface and tooth-count indices together, the available evidence suggests that young children with Down syndrome are not more susceptible to caries that typically developing children; nevertheless, early prevention immediately after tooth eruption remains important.

The occurrence of dental caries is influenced by background factors, but findings for caries in the primary dentition in Down syndrome have been inconsistent after accounting for these factors, and few high-evidence studies are available. Further validation is needed.

Literature

1) Mathias M F, Simionato MR, Guaré RO. Some factors associated with dental caries in the primary dentition of children with Down syndrome. Eur J Paediatr Dent 2011;12:37-42.
2) Hashizume LN, Schwertner C, Moreira MJS, et al. Salivary secretory IgA concentration and dental caries in children with Down syndrome. Spec Care Dentist 2017;37:115-19.

3) Maclaurin ET, Shaw L, Foster TD. Dental caries and periodontal disease in children with Down syndrome and other mentally handicapping conditions. Paediatr Dent 1985;1:15-9.
4) Lee SR, Kwon HK, Song KB, et al. Dental caries and salivary immunoglobulin A in Down syndrome children. J Paediatr Child Health 2004;40:530-3.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search result
25,433 (Pubmed), 21 (Cochrane Reviews)
#1. “Down Syndrome”[MeSH]
#2. “Tooth, Deciduous“[MeSH]
#3. “Dental Caries“[MeSH]
#4. #2 OR #3
#5. #1 AND #4
#6. “Guideline”[Publication Type] #7. #5 AND #6
Papers considered useful: 6

Medical Journal Search Results
8,731 (Medical journal)
#1. deciduous tooth
#2. dental caries
#3. Down syndrome
#4. guidelines
#5. deciduous tooth caries
#6. #1 and #3
#7. #1 and #2
#8. #3 and #4
#9. #3 and #4 and #1
#10. #3 and #4 and #5
Papers considered useful: 0

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Caries Control Team)

CQ5	Are permanent teeth in children with Down syndrome less susceptible to dental caries than those in typically developing children?

[Perspective]
Permanent teeth in children with Down syndrome may be less susceptible to dental caries than those in typically developing children. (2D)

Background and Objectives

Whether the permanent teeth of children with Down syndrome are less susceptible to dental caries than those of typically developing children is an important question for oral health, quality of life, and overall health maintenance in children Down syndrome.

Explanation

This literature search identified 341 articles; one systematic review (including six studies) and five primary studies were included in the analysis.

Age and background factors influence caries occurrence. Cutress[1] reported significantly lower DMFT (Decayed, Missing and Filled teeth; permanent dentition) in individuals with Down syndrome aged 5–9, 10–14, 15–19, and 20–24 years compared with typically developing peers. Hosaka et al.[2] found significantly lower DMFT in teens with Down syndrome than in individuals with intellectual developmental disorder not due to Down syndrome, but no significant differences in those in their 20s and 30s—suggesting an effect of delayed eruption on post-eruptive caries experience.

In age-matched comparisons of Down syndrome and intellectual developmental disorder cohorts, some studies found significantly fewer DMFT in Down syndrome,[3] while others found no significant difference.[4] In comparisons of Down syndrome and siblings living in the same environment, DMFT was lower in Down syndrome than in typically developing siblings.[5] Reported contributions to lower DMFT in Down syndrome include fewer approximal (adjacent-surface) caries[3] and fluoride use; with aging, DMFT increases.[6] A meta-analysis of six studies by Deps et al.[7] reported a significantly lower DMFT in Down syndrome (mean difference -1.01 teeth, 95% CI -1.45 to -0.57), though study results were highly variable and the risk of bias was high.

Overall, the search suggested that children with Down syndrome may be slightly less susceptible to dental caries in the permanent dentition, but regular dental management is recommended because of environmental factors and aging influence caries experience.

Literature

1) Cutress T W. Dental caries in trisomy 21. Arch Oral Biol 1971;16(11):1329-44.
2) Hosaka K, Ogasawara T, Watanabe T, et al. Prevalence of Dental Caries in Down's Syndrome-Part 1 Clinico-statistical Survey on Incidence by Age-Journal of dentistry for the handicapped.1992;13:169-78.
3) Barnett ML, Press KP, Friedman D, et al. The prevalence of periodontitis and dental caries in a Down syndrome population J Periodontol 1986;57:288-93 .

4) Stabholz A, Mann J, Sela M, et al. Caries experience, periodontal treatment needs, salivary pH, and Streptococcus mutans counts in a preadolescent Down Spec Care Dentist 1991;11:203-8.
5) Fung K, Lawrence H, Allison P. A paired analysis of correlates of dental restorative care in siblings with and without Down syndrome. Spec Care Dentist 2008;28:85-91.
6) Fung K, Allison PJ. A comparison of caries rates in noninstitutionalized individuals with and without Down syndrome. Spec Care Dentist 2005;25:302-10 .
7) Deps TD., Angelo GL, Martins CC, et al. Association between Dental Caries and Down Syndrome: A Systematic Review and Meta-Analysis. PLoS One 2015;10:e 0127484.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search result
25,433 (Pubmed), 21 (Cochrane Reviews)
#1. "Down Syndrome"[MeSH]
#2. "Dentition, Permanent"[MeSH]
#3. "Dental Caries"[MeSH]
#4. #2 OR #3
#5. #1 AND #4
#6. "Guideline" [Publication Type] #7. #5 AND #6
Papers considered useful: 14

Medical Journal Search Results
6,865 (Medical Journal)
#1. permanent teeth
#2. dental caries
#3. down syndrome
#4. permanent dental caries
#5. guidelines
#6. #1 and #2
#7. #1 and #3
#8. #3 and #4
#9. #5 and #1
#10. #5 and #2
#11. #5 and #3
#12. #5 and #4
Papers considered useful: 4

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Caries Control Team)

CQ6	Is the application of fluoride useful for the prevention of dental caries in people with Down syndrome?

[Perspective]
We considered whether topical fluoride prevents dental caries in people with Down syndrome but found no Down syndrome-specific evidence of benefit compared with typically developing individuals. （None.）

Background and Objectives

The effectiveness of topical fluoride application in the prevention of dental caries in people with Down syndrome is not clear.

Explanation

Seven papers were selected: two systematic reviews on dental carries in Down syndrome (Moreira[1] and Deps[2]) without discussion of topical fluoride; two literature reviews (Shore[3] and Hennequin[4]) with no evaluation of fluoride effects; and three biochemical searches of exfoliated primary teeth in Down syndrome (Kusabe[5] and Nakano[6,7]), also without data on fluoride application. On this basis, we could not answer the question "Is topical fluoride useful for preventing dental caries in Down syndrome?"

However, although the influence of Down syndrome characteristics on fluoride application and mouthwash has not been clarified, it has been reported that "the daily use of fluoride toothpaste (1,100 to 1,400 ppmF) in combination with fluoride mouthwash (250 to 900 ppmF) for root surface caries in sedentary individuals can restore permanent teeth." The combination of a fluoride-containing toothpaste (1,100-1,400 ppmF) with a fluoride mouthwash (250-900 ppmF) daily has been shown to restore (harden and become inactive) active root surface caries in permanent teeth.[8] Gluzman[9] et al. reported the effectiveness of silver diamine fluoride not only in inhibiting caries progression in primary teeth but also in the primary prevention of root surface caries in people requiring nursing care. In Japan, the Ministry of Health, Labour and Welfare (MHLW) has approved the marketing of a high-concentration fluoride toothpaste as a quasi-drug with a fluoride ion concentration limit of 1,500 ppmF. The Japanese Society of Oral Hygiene and Preventive Medicine (JOSPH) has issued guidance on fluoride toothpaste use—"Application amount and fluoride ion concentration of fluoride toothpaste by age" and "Fluoride application by life stage"—though these are not Down syndrome-specific recommendations.[10] The toothpaste application of fluoride concentration of 500 ppmF for 6 months to 5 years of age, 1,000 ppmF for 6 to 14 years of age, and 1,000 to 1,500 ppmF for 15 years and older was indicated.[10] Currently, toothpaste applications with fluoride concentrations of 1,000 ppmF (about 1 to 2 mm for teething 2 years, 1,000 ppmF (about 5 mm) for 3 to 5 years, and 1,400 to 1,500 ppmF (about 1.5 to 2.0 cm) for 6 years to adults are indicated.[11]

Regarding dental caries in Down syndrome, observational studies and cross-sectional studies have examined the number of decayed teeth, but detailed studies on living environments, oral hygiene, and caries prevention methods have not been conducted. In particular, the method of fluoride application and the incidence of dental caries may be greatly influenced by the characteristics

of Down syndrome, the fluoride concentration in drinking water in the community, the attitudes of the patients and their guardians, and the state of oral function of the patients. Therefore, although topical fluoride may be used to prevent dental caries in people with Down syndrome, the influence of Down syndrome-specific characteristics on its use and effectiveness remains unclear. Because large-scale population studies are difficult, high-quality evidence should be accumulated through prospective data collection from infancy in controlled care settings (e.g., institutional environments).

Literature

1) Moreira MJ, Schwertner C, Jardim JJ, et al. “Dental caries in individuals with Down syndrome: a systematic review. “Int J Paediatr Dent 2016 Jan;26(1):3- 12.
2) Deps TD, Angelo GL, Martins CC, et al. Association between Dental Caries and Down Syndrome: A Systematic Review and Meta-Analysis. PLoS One 2015 Jun 18;10 (6):e0127484.
3) Shore S, Lightfoot T, Ansell P. Oral disease in children with Down syndrome: causes and prevention. Community Pract 2010 Feb;83(2):18-21.
4) Hennequin M, Veyrune JL, Buordiol P. Significance of oral health in persons with Down syndrome: a literature review. Developmental Medicine & Child Neurology 1999;41:275-83.
5) Kusabe Y, Nakano T, Okamoto T, et al. Distribution of fluoride and magnesium concentrations in deciduous tooth enamel of children with cerebral palsy and Ped Dent J 2012;22(2): 103-109.
6) Nakano T, Kawai T, Higashi K, et al. Bichemical Characteristic of Deciduous Enamel Before and After the Neonatal Line in the Down Syndrome. Pediatric Dentistry 2001;39(3): 561-7.
7) Nakano T. Biochemical Characteristic on Enamel Surface of Deciduous Tooth in Down Syndrome-F, Mg and Volume of Decalcification-. The Aichi-Gakuin Journal of Dental Science . 1999;37(1):175-86.
8) The Japanese Society of Conservative Dentistry (ed.): Guidelines for the treatment of dental caries. 3rd edition, Kyoto: Nagasue Shoten; 2020.
9) Gluzman R, Katz RV, Frey BJ, et al. Prevention of root caries: a literature review of primary and secondary preventive agents. Spec Care Dentist 2013May-. Jun;33(3):133-40.
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11) Recommended Use of Fluoride-containing Toothpaste by Four Academic Societies January 1, 2023. https://www.kokuhoken.or.jp/jsdh/news/2023/news_230106.pdf

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search result
25,433 (Pubmed), 21 (Cochrane Reviews)
#1. “Down Syndrome”[MeSH]
#2. “Dental Caries”[MeSH]

#3. “Fluorine”[MeSH] AND “Fluorides”[MeSH] #4. #1 AND #2 AND #3
#5. #1 AND #2
Papers considered useful: 5

Medical Journal Search Results
1,001 (Medical Journal)
#1. dental caries prevention
#2. fluorine
#3. fluoride application
#4. fluoride mouthwash
#5. fluoride
#6. fluoride application
#7. toothpaste containing fluoride
#8. fluoride concentration
#9. Down syndrome caries
#10. #1 and #2
#11. #1 and #3
#12. #1 and #4
#13. #1 and #5
#14. #1 and #6
#15. #1 and #7
#16. #1 and #8
#17. #1 and #9
#18. #1 and #2 and #9
#19. Guidelines
#20. #2 and #10 and #20
#21. #4 and #9
#22. Mouthwash
#23. #9 and #23
#24. toxic effects
#25. #2 and #25
#26. Acute poisoning and fluoride
#27. chronic poisoning and fluoride
#28. #2 and #25 and #20
#29. teeth and fluoride
#30. #20 and #30
#31. fluorine Chromosome
Papers considered useful: 2

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Caries Control Team)

CQ7

Is myofunctional therapy effective in improving lip closure and tongue function in individuals with Down syndrome?

[Perspective]
If orofacial myofunctional therapy is undertaken, lip closure and tongue function may improve. (2D)

Background and Objectives

People with Down syndrome often show abnormal tongue rest posture and atypical swallowing; lip incompetence (open-mount posture) at rest is also common. This CQ evaluates the usefulness of myofunctional therapy for correcting tongue rest posture/swallowing and lip incompetence.

Explanation

Down syndrome is characterized by difficulty in lip closure at rest. There are various possible causes, such as the tongue apex being positioned anterior to the lips, difficulty in lip closure, nasal breathing failure due to mouth breathing, and opening of the mouth due to the muscle relaxation also seen in the mouth-closing muscles, which is a characteristic of Down syndrome.[1)] To promote lip closure at rest, if the apex of the tongue is positioned anterior to the lips, it is necessary to position the tongue in the native oral cavity at rest, and if the tongue can be placed in the native oral cavity during swallowing, the tongue position may be improved and lip closure may be achieved. In the case of mouth breathing, if there is no problem with nasal ventilation, nasal breathing can be trained and lip closure can be achieved. In the case of muscle relaxation, lip closure may be possible if the strength of the mouth-closing muscles is increased.

The literature reviewed in this study included children with Down syndrome. In order to promote lip closure at rest, it is important to check for nasal breathing, including nasal obstruction and adenoids, as well as the presence of underlying cardiac disease, which may cause decreased arterial oxygen saturation. It is also necessary to check for morphological abnormalities such as tooth inclination and upper and lower jaw bones that would make lip closure difficult. In addition, it is important to improve the tongue position during lip closure; to strengthen the orbicularis oris muscle and mouth-closing muscles, which are important for lip closure; and to acquire the habit of nasal breathing during lip closure.

The results of the present search identified case reports of myofunctional therapy for lip closure at rest in individuals with Down syndrome.[2-4)] Each case report was a comparison of pre- and post-intervention for individuals with Down syndrome with good cooperation. Among them, it was reported that myofunctional therapy aimed at lip closure could be evaluated to a certain degree.[1-31)] Tongue position was also generally favorable, with a tendency to be able to hold the tongue in the native oral cavity.[1-3)] Therefore, it was reported that if the resting tongue position could be improved, the tongue movement and lip closure during feeding and swallowing could also be functionally improved.[2-4)]

Based on the results of this study, it is necessary to confirm which active training is applicable to the target individuals with Down syndrome if they are

able to imitate. Even if the patient has difficulty with imitation, if communication is possible using picture cards or talking to the patient, active training may be method.[5] The number of times and duration of various types of myofunctional therapy are also dependent on the patient's acceptance, so the evidence is unknown. In the future, it is necessary to further verify the training methods and the duration of training according to the acceptance of the patient.

Literature

1) Mizukami M , Tamura F, Matsuyama M, et al.Relationship between Gross Motor Function and Oral Parafunction Habits Relevant to Eating in children with Down syndrome. J. Jpn.Soc. Disability Oral Health 2015;36(1):17-24.
2) Ide T, Uechi R, Tamai H. Mouth care for children with Down syndrome. Japanese Journal of Down Syndrome Rehabilitation Research 2019;2433-376(12)
:52-4.
3) Lee AS, Gibbon FE. non-speech oral motor treatment for children with developmental speech sound disorders Cochrane Database Syst Rev 2015(3), 2015. CD CD009383. doi: 10.1002/14651858.
4) Saccomanno S, Martini C, D'Alatri L, et al. 3 A specific protocol of myofunctional therapy in children with Down syndrome. A pilot study. Eur J Paediatr Dent 2018 Sep;19(3):243-6. doi: 10.23804/ejpd.2018.19.03.14.
5) Edited by Kazuhiko Omoto, supervised by Yoshihiro Kaneko. Eating, Swallowing, and Respiratory Rehabilitation of Children with Disabilities: Basics and Practice. Tokyo: Medical and Dental Publishing,,2005. 293-5.

Search expression

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
15,612 (Pubmed), 21 (Cochrane Reviews)
#1. “down syndrome”[Title/Abstract].
#2. “myofunction”[All Fields] OR “myofunctional”[All Fields]
#3. “#1 and #2 and (therapy)
#4. (#3) and (tongue)
#5. (#4) and (lip)
#6. (#3) and (lip)
Papers considered useful: 2

Medical Journal Search Results
3,964 (Medical Journal)
#1. (Down syndrome /TH or Down syndrome /AL) and (PT= except conference proceedings)
#2. (Myofunctional Therapy /TH or Myofunctional Therapy /AL) and (PT= except meeting minutes)
#3. (Lip Closure /AL) and (PT= except meeting minutes)
#4. (tongue function /AL) and (PT= except meeting minutes)
#5. #1 and #2
#6. #3 and #5
#7. #4 and #5

Papers considered useful: 2

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Dental Growth Team)

CQ8

Is the use of the Castillo-Morales palatal plate effective in improving tongue function in individuals with Down syndrome?

[Perspective]
If the Castillo-Morales palatal plate is accepted, there is a possibility that tongue function will improve with long-term use. (2D)

Background and Objectives

Down syndrome tends to cause abnormal tongue position at rest and during swallowing. The purpose of this study was to investigate whether the Castillo-Morales palatal plate could improve this condition.

Explanation

(For more information on myofunctional therapy, see CQ7: Is myofunctional therapy effective in improving lip closure and tongue function in individuals with Down syndrome? for more information on myofunctional therapy).

The Castillo-Morales palatal plate (CM plate) was first reported in 1982.[1)] This intraoral appliance aims to correct tongue posture at rest and during swallowing by providing sensory–motor stimulation with a rotating bead mounted on the anterior portion of the plate, stimulating the tongue and the oral vestibule. In some cases, a protrusion is placed on the palate to guide the tongue position. In any case, since the device is worn only at rest, its purpose is to control tongue protrusion and induce lip closure at rest and during salivary swallowing, with the aim of improving articulatory function, feeding and swallowing function, and drooling except at rest.

In this search, case reports were identified. For the tongue position, the tongue position at rest tended to improve when the device was worn for one hour twice a day at an early age (after 3 years of age).[2-6)] Although the degree of underlying disease or intellectual developmental disorder was not stated, there was a trend toward improvement, including tongue movement during conversation.[2-6)] For lip closure, when the device was applied twice a day for 1 hour at an early age (after 3 years of age), there was a trend toward improvement in lip closure at rest.[2–6a)] For the jaw position, the resting lip closure was improved at 13.4±5.0 months. In a comparison using cephalometric analysis before and after the use of the device for one month, three times a day for one hour, the Class III tendency was not substantially changed.[7)] Therefore, the device was considered to be effective in improving lip closure, tongue movement at rest and during conversation, and drooling. However, patients needed to adapt to the appliance; many reports described use for approximately 1 hour per session, and a trend toward improvement was observed.

In the case report, the minimum age of use was more than 15 months. The youngest age at which the device was applied was 3 months, but since the sucking reflex remained, it is considered premature to improve the tongue position.[3,4)] In Japan, five case reports have been published, showing that tongue function tended to improve on the CM floor.[8-12)]. Therefore, it was considered possible that the device could be improved by confirming its acceptance and

long-term use as early as possible after the disappearance of the primitive reflexes related to sucking, including the sucking reflex.

Literature

1) Castillo-Morales R, Crotti E, et al. Orofaziale Regulation beim Down-Syndrome durch Gaumenplatte. Sozialpadiatrie 1982;4:10-17.
2) Javed F, Akram Z, Barillas AP, et al. Outcome of orthodontic palatal plate therapy for orofacial dysfunction in children with Down syndrome: A systematic review. orthod Craniofac Res 2018Feb;21(1):20-6.
3) Carlstedt K, Henningsson G, Dahllöf G. A four-year longitudinal study of palatal plate therapy in children with Down syndrome: effects on oral motor Acta Odontol Scand 2003 Feb;61(1):39-46. doi: 10.1080/ode.61.1.39.46.
4) Bäckman B, Grevér-Sjölander AC, Bengtsson K, et al. Children with Down syndrome: oral development and morphology after use of palatal plates between 6 Int J Paediatr Dent. 2007.
5) Carlstedt K, Henningsson G, McAllister A, et al. Long-term effects of palatal plate therapy on oral motor function in children with Down syndrome Acta Odontol Scand 2001 Apr;59(2):63-8. doi: 10.1080/000163501750157117.
6) Zavaglia V, Nori A, Mansour NM. Long-term effects of palatal plate therapy for orofacial regulation in children with Down syndrome. J Clin Pediatr Dent 2003 Fall;28(1):89-93.
7) Korbmacher H, Moeller HC, Klocke A, et al. Cephalometric evaluation of children with Down syndrome after early intervention with a stimulating plate. Spec Care Dentist Sep-Oct 2005;25(5):253-9.
8) Fujita H, Yabushita Y, Imamoto H, et al. A case of Castillo-Morales paratarsal plate in a patient with dysarthria during feeding rehabilitation. Journal of the Hokkaido Dental Association 2005;60: 59-62.
9) Ogata K. Oral functional training with paratarsal plates Castillo-Morales' oral-facial coordination training. Journal of the Japanese Dental Association 1991;44:257-64.
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11) Hironishi M, Daikoku H, Fukui N, et al. Application and clinical evaluation of Castillo-Morales palatal floor in dentistry for the handicapped. J. Jpn. Soc. Disability Oral Health 1998;19:227-35.
12) Daikoku H, Amano A, Fukui N, et al. Clinical evaluation of orofacial regulation therapy for Down syndrome children using Castillo- Morales palatal plate, Pediatric Dental Journal 2000;10: P133-137.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
40,351 (Pubmed), 21 (Cochrane Reviews)
#1. down syndrome

#2. #1 AND (myofunctional) AND (therapy) AND (palatal plate)
Papers considered useful: 6

Medical Journal Search Results
3,964 (Medical Journal)
#1. (Down syndrome /TH or Down syndrome /AL) and (PT= except conference proceedings)
#2. (Occlusal Splint /TH or Intraoral Device /AL) and (PT= except meeting minutes)
#3. (Occlusal Organ Implant /TH or Prosthetic /AL) and (PT= except meeting minutes)
#4. (tongue function /AL) and (PT= except meeting minutes) #5. #1 and #2
#6. #1 and #3
#7. #1 and #4
Papers considered useful: 0

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Dental Growth Team)

CQ9	Are surgical interventions recommended for delayed tooth eruption in individuals with Down syndrome?

[Perspective]
Surgical treatment for delayed tooth eruption is possible if the procedure is acceptable, but the evidence for recommending surgical treatment is unclear. (3D)

Background and Objectives

The purpose of this study was to investigate the usefulness of surgical procedures such as tooth extraction and window opening for late eruption of primary teeth and delayed eruption or eruption of permanent teeth in Down syndrome.

Explanation

Surgical procedures such as tooth extraction and window opening for late remaining primary teeth and delayed eruption or eruption of permanent teeth in children with Down syndrome are the same as those for typically developing children.

In patients with cardiac disease, prevention of infective endocarditis requires preoperative antimicrobial prophylaxis in accordance with the *Guidelines for the Prevention and Treatment of Infective Endocarditis, Revised 2017.*[1] This indication reflects the underlying cardiac comorbidity that may co-occur with Down syndrome, not Down syndrome itself.[2]

If the patient is unable to cooperate with dental treatment, it is necessary to confirm whether behavioral therapy is acceptable, and if surgical treatment is judged to be difficult, it is necessary to consider physical behavioral modification (physical restraint) or pharmacological behavioral modification (general anesthesia). In addition, it is also necessary to consider suture options, such as not suturing if stitches cannot be removed, or using absorbable threads if stitches are necessary.

In our search, we found a case report of a surgical procedure (window opening) for delayed tooth eruption in Down syndrome performed under general anesthesia.[3] Although not an awake procedure, it was considered to be feasible if the patient could cooperate in the preoperative and postoperative procedures under general anesthesia, but the evidence is unclear.

Therefore, the decision to perform an open surgical procedure depends on the patient's acceptance of the procedure, and it is important to evaluate the risks and benefits of the procedure according to the patient's symptoms. Further verification of surgical treatment for delayed tooth eruption is needed.

Literature

1) Japanese Society of Cardiology, Guidelines for the Prevention and Treatment of Infective Endocarditis 2017 Revision .2018. https://www.j-circ.or.jp/cms/wp-

content/uploads/2020/02/JCS2017_nakatani_h.pdf (see 2022- 4-23)
2) Japanese Society for Dental Health Care for the Sick, Japanese Society of Oral and Maxillofacial Surgery, Japanese Society of Geriatric Dentistry Editors, Guidelines for the extraction of teeth in patients undergoing antithrombotic therapy 2020 Edition, Academic Publisher, Tokyo ,2020. https://minds.jcqhc.or.jp/docs/gl_pdf/G0001242/4/Exdontia_in_patient_with_antithrombotic_treatment.pdf (ref 2022-4-23)
3) Li S. et al. Eruptive Guidance of Upper Impacted Canine with a follicular dental cyst in a patient with Down syndrome. J. Jpn. Soc. Disability Oral Health. 2009;30:120-4.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
40,351 (Pubmed), 21 (Cochrane Reviews)
#1. down syndrome
#2. Teeth
#3. delayed eruption
#4. tooth extraction
#5. Fenestration
#6. Surgical
#7. #1 and #2
#8. #3 and #7
#9. #4 and #7
#10. #5 and #7
#11. #6 and #7
Papers considered useful: 0

Medical Journal Search Results
3,964 (Medical Journal)
#1. (Down syndrome /TH or Down syndrome /AL) and (PT= except conference proceedings)
#2. (unmigrated teeth /TH or delayed eruption /AL) and (PT= except meeting minutes) #3. (surgical procedures /TH or surgical procedures /AL) and (PT= except meeting minutes) #4. (teeth /TH or teeth /AL) and (PT= except meeting minutes)
#5. #1 and #2
#6. #1 and #3
#7. #5 and #4
#8. #6 and #4
Papers considered useful: 1

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Dental Growth Team)

CQ10	To what extent is orthodontic treatment (excluding surgical orthodontic treatment) recommended for individuals Down syndrome?

[Perspective]
Treatment is possible if the patient can accept orthodontic treatment, but the evidence for the indicated cases is unknown. (3D)

Background and Objectives

There have been case reports of orthodontic treatment (excluding surgical orthodontic treatment) for Down Syndrome, but the criteria are unclear. Therefore, we aimed to investigate the extent to which orthodontic treatment is actually recommended.

Explanation

Orthodontic treatment for Down syndrome is covered by insurance at medical institutions that have notified the Director of the Regional Bureau of Health, Labour and Welfare of their compliance with the facility standards specified by the Minister of Health, Labour and Welfare. Therefore, treatment can be covered by dental insurance at a dental clinic or hospital dental institution that has been designated by the Minister of Health, Labour and Welfare under the Medical aid for children with potential disability (ikusei-iryo) for patients under 18 years of age, or the Medical rehabilitation service (kousei-iryo) for those aged 18 and over.

The diagnostic criteria for malocclusion are the same as those for typically developing children, but there are no criteria for how far the chief complaint should be treated. In the present study, all the case reports showed a tendency to confirm the patient's adaptation to orthodontic treatment at the time of treatment, based on the patient's general condition, the level of cooperation with treatment, and the parents' wishes. [1)]

Therefore, the evidence for the indicated cases is unknown, and diagnosis and treatment may be necessary depending on the chief complaint and the degree of cooperation with dental treatment.

Even if the patient is able to cooperate with orthodontic treatment, if oral hygiene deteriorates due to the orthodontic appliance, caries may occur more frequently and periodontal disease may progress. Maintenance and improvement of oral hygiene are also important for the continuation of treatment.

In cases of cardiac disease, it is important to consult with the patient's primary physician regarding the pros and cons of orthodontic treatment.[2)] When orthodontic treatment is performed, care should be taken to avoid possible damage to the oral mucosa caused by the device.

Literature

1) Horie Y, Miyazaki H, Kosaka T. et al, Clinical survey of orthodontic treatment in Down's syndrome patients. Dental Science Journal 2009;109: 381-7.

2) Japanese Society of Cardiology. Guidelines for the Prevention and Treatment of Infective Endocarditis 2017 Revision .2018. https://www.j-circ.or.jp/cms/wp-content/uploads/2020/02/JCS2017_nakatani_h.pdf (see 2022- 04-23)

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
40,351 (Pubmed), 21 (Cochrane Reviews)
#1. down syndrome
#2. orthodontic treatment
#3. Outcome
#4. Possibility
#5. Recommendation
#6. Effects
#7. #1 and #2
#8. #3 and #7
#9. #4 and #7
#10. #5 and #7
#11. #6 and #7
Papers considered useful: 0

Medical Journal Search Results
3,964 (Medical Journal)
#1. (Down syndrome /TH or Down syndrome /AL) and (PT= except conference proceedings)
#2. (Orthodontic Treatment /TH or Orthodontic Treatment /AL) and (PT= except meeting minutes) #3. #1 and #2
Papers considered useful: 1

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Dental Growth Team)

CQ11	Are younger children with Down syndrome more susceptible to periodontal disease than younger children with non-Down syndrome intellectual developmental disorder or typically developing children?

[Perspective]
Younger children with Down syndrome are more susceptible to periodontal disease than children with other intellectual developmental disorder or typical development. (2C)

Background and Objectives

Individuals with Down syndrome are more likely to develop periodontal disease at an early age and to develop severe periodontal disease. The purpose of this study was to examine the epidemiological data on the incidence of periodontal disease in young individuals with Down syndrome compared with other children with intellectual developmental disorder and typically developing children.

Explanation

A systematic review of the prevalence of periodontal disease in young (6–19 years) individuals with Down syndrome with other intellectual developmental disorder or stereotyped development was conducted using the degree of gingival inflammation, plaque adhesion, and calculus (tartar) deposition as outcomes in controlled studies of children with other intellectual developmental disorder or stereotyped development. All of the selected references were observational studies.

This been reported that the gingival index (GI)[1] and gingival bleeding index (GBI)[2,3] are significantly higher in individuals with Down syndrome younger than 4 years compared to normal children of the same age, and that the PMA index is also significantly higher in individuals with Down syndrome than in normal children of the same age[4]. It should be noted that gingival inflammation tends to occur at an early age. In a study evaluating the degree of plaque adhesion, the simplified oral hygiene index (OHI-S) was significantly higher in individuals with Down syndrome compared with controls[5], while another study reported no significant difference in the degree of plaque adhesion in individuals with Down syndrome[2].In studies evaluating the degree of calculus accumulation[6-8], no significant differences were found between individuals with Down syndrome and control groups. In the meta-analysis, there was no significant difference in the degree of plaque adhesion and calculus accumulation, but there was a significant difference in the degree of gingival inflammation (mean difference 1.07, 95% confidence interval: 0.05–2.10).These results suggest that individuals with Down syndrome are more susceptible to periodontal disease due to a greater degree of gingival inflammation than other intellectual developmental disorder or typically developing children. The factors that contribute to the development of periodontal disease in young individuals with Down syndrome include not only poor oral hygiene, but also abnormal tooth morphology as a plaque retention factor, malocclusion, bad habits, and host and genetic factors due to immune

dysfunction, although the mechanisms remain unclear. However, the mechanism is still unclear. Further investigation of periodontal disease in young patients with Down syndrome is needed, including elucidation of the mechanism of the disease.

Literature

1) Al Habashneh R, Al-Jundi S, Khader Y, et.al . Oral health status and reasons for not attending dental care among 12- to 16-year-old children with Down syndrome in special needs centres in Jordan Int J Dent Hyg 2012;10(4):259-64.
2) Faria Carrada C, Almeida Ribeiro Scalioni F, Evangelista Cesar D, et al. Salivary Periodontopathic Bacteria in Children and Adolescents with Down Syndrome. PLoS One 2016 Oct 11;11(10):e0162988. doi: 10.1371/journal.pone.0162988. eCollection 2016.
3) Sakellari D, Arapostathis KN, Konstantinidis A. Periodontal conditions and subgingival microflora in Down syndrome patients. J Clin Periodontol 2005 J Clin Periodontol 2005;32(6):684-90.
4) Morinushi T, Lopatin DE, Van Poperin N. The relationship between gingivitis and the serum antibodies to the microbiota associated with periodontal disease in children with Down's syndrome. J Periodontol 1997;68(7):626-31.
5) HabibeC-H, Yoshida R-A, Gorjão R, et al. Comparison of salivary cytokines levels among individuals with Down syndrome, cerebral palsy and normoactive. J Clin Exp Dent 2020;12(5):e446-e51.
6) Barr-Agholme M, Dahllöf G, Modéer T, et al. Periodontal conditions and salivary immunoglobulins in individuals with Down syndrome. J Periodontol 1998;. 69(10):1119-23.
7) Barr-Agholme M, Dahllöf G, Linder L, et al. Actinobacillus actinomycetemcomitans, Capnocytophaga and Porphyromonas gingivalis in subgingival plaque of adolescents with Down's syndrome. Oral Microbiol Immunol 1992;7(4):244-8.
8) Modéer T, Barr M, Dahllöf G. Periodontal disease in children with Down's syndrome. Scand J Dent Res 1990;98(3):228-34.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
40,351 (Pubmed), 21 (Cochrane Reviews)
#1. down syndrome
#2. periodontal disease OR periodontitis OR gingivitis
#3. healthy OR typical development OR intellectual disability #4. pediatric OR child OR young
#5. #1 AND #2 AND #3 AND #4
Papers considered useful: 3

Medical Journal Search Results
9,581 (medical journal)

#1. Down syndrome /TH or Down syndrome /AL
#2. (Periodontal disease /TH or Periodontal disease /AL) or (Periodontitis /TH or Periodontitis /AL) or (Gingivitis /TH or Gingivitis /AL)
#3. (Child /TH or Child /AL) or Young /AL or (Minor /TH or Minor /AL)
#4. healthy OR/AL and regular development /AL or (intellectual disability /TH or intellectual disability /AL)
#5. (#1 and #2 and #3 and #4) and (PT= except meeting minutes)
Papers considered useful: 19

Resolution Decision

Acceptable 13 Not acceptable 0 Correction required 0

(Periodontal Disease Management Team)

CQ12	Are adults with Down syndrome more susceptible to periodontal disease than adults with other intellectual developmental disorder from causes other than Down syndrome or adults without intellectual developmental disorder?

[Perspective]
Adults with Down syndrome are more susceptible to periodontal disease than other intellectual developmental disorder or typically developing individuals. (2C)

Background and Objectives

It is known that people with Down syndrome are susceptible to various infectious diseases due to their physical characteristics. In the oral cavity, it has been recognized that periodontal disease occurs early in life and is frequently severe, and many studies have been conducted in this area. The purpose of this study was to compare the susceptibility to periodontal disease among adults with Down syndrome, intellectual developmental disorder unrelated to Down syndrome, or typical development.

Explanation

We searched for studies that investigated periodontal disease in adults with Down syndrome compared to age-, gender-, and race-matched individuals with intellectual developmental disorder or typical development, focusing on the degree of gingival inflammation, periodontal pocket depth (probing depth; PD), attachment level (clinical attachment level; CAL), degree of tooth mobility, amount of plaque accumulation, and amount of calculus (tartar) accumulation. There were no studies that adequately evaluated tooth mobility and calculus accumulation. In a cross-sectional study that investigated the degree of gingival inflammation, the bleeding on probing (BOP)[1,2] and gingival index (GI)[1,2] were evaluated in adults with Down syndrome compared with those in adults with typical development. The plaque index (PlI)[1] showed an increase in the amount of plaque adhesion and periodontal pockets of 4 mm or more.[2] As for CAL, the average bone resorption of the alveolar bone of the whole jaw measured by dental X-ray showed that bone resorption was significantly more advanced in patients with Down syndrome (3.0±1.5 mm) than in those with intellectual developmental disorder (1.9±1.1 mm).[3] In CAL by probing attachment, loss was greater in patients with Down syndrome than in those with intellectual developmental disorder unrelated to Down syndrome or those with typical development,[1,2] and there was no relationship between CAL and institutionalization, home care, or frequency of dental visits.[1] In addition, the results of regression analysis showed a significant association between PlI and CAL.[4] However, in the present meta-analysis, the mean difference in periodontal pocket depth was 1.54, and the only significant finding was an increase in periodontal pocket depth in people with Down syndrome (95% confidence interval: 0.66–2.43). The above findings suggest that adults with Down syndrome are more susceptible to periodontal disease than typically developed individuals. The factors involved in the onset and progression of

periodontal disease in individuals with Down syndrome include not only poor oral hygiene, but also local factors such as abnormal tooth morphology that can cause occlusal trauma, malocclusion, and bad habits. In addition, host and genetic factors such as immune dysfunction are also considered, although many aspects of the underlying mechanisms remain unclear. In the future, it will be necessary to elucidate the disease mechanisms through basic research and to examine various confounding factors related to oral health care, such as daily oral care, dental visits, and lifestyle background, including comorbidities and cooperative relationships, through longitudinal studies.

Literature

1) Khocht A, Janal M, Turner B. Periodontal health in Down syndrome: contributions of mental disability, personal, and professional dental care. Spec Care Dentist 2010;30:118-23.
2) Sakellari D, Arapostathis KN, Konstantinidis A. Periodontal conditions and subgingival microflora in Down syndrome patients A case- J Clin Periodontol 2005;32:684-90.
3) Knoll S, Janal M, Khocht A. Radiographic assessment of periodontitis in African-Americans with Down syndrome. J Int Acad Periodontol 2008;10:16-21.
4) Khocht A, Russell B, Cannon JG, et al. Oxidative burst intensity of peripheral phagocytic cells and periodontitis in Down syndrome. J Periodontal Res 2014;49:29-35.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
40,351 (Pubmed), 21 (Cochrane Reviews)
#1. down syndrome
#2. periodontal disease OR periodontitis OR gingivitis
#3. healthy OR typical development OR intellectual disability
#4. #1 AND #2 AND #3 filters:19+years
Papers considered useful: 14

Medical Journal Search Results
9,581 (medical journal)
#1. Down syndrome /TH or Down syndrome /AL
#2. (periodontal disease /TH or periodontal disease /AL) OR (periodontitis /TH or periodontitis /AL) OR (gingivitis /TH or gingivitis /AL)
#3. healthy OR/AL and regular development /AL OR (intellectual disability /TH or intellectual disability /AL)
#4. (#1 and #2 and #3) and (PT= except conference proceedings) papers considered useful: 3

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Periodontal Disease Management Team)

CQ13	Are there differences in oral microflora among children with Down syndrome, children with intellectual developmental disorder from causes other than Down syndrome, and typically developing children?

[Perspective]
Down syndrome do not show differences in periodontal pocket flora compared to intellectual developmental disorder, cerebral palsy, or typically developing children. (3D)

Background and Objectives

To test whether the oral microflora is unique among the risk factors for the development and progression of periodontal disease in Down syndrome.

Explanation

We compared the types of periodontopathogenic bacteria detected in children with Down syndrome and in typically developing children or children with intellectual developmental disorder or cerebral palsy of the same age group. The types of bacteria included in the search were the typical periodontopathogenic bacteria *Porphyromonas gingivalis* (*P. gingivalis*), *Tannerella forsythia* (*T. forsythia*), *Toreponema denticola* (*T. denticola*), *Aggregatibacter actinomycetemcomitans* (*A. actinomycetemcomitans*), and *Prevotella intermedia* (*P. intermedia*). A meta-analysis was performed on a total of five references that showed the frequency of detection of bacteria in subgingival plaques: two references for controls with typical development, two references for participants with intellectual developmental disorder, and one reference for patients with cerebral palsy. In a study targeting younger children 2–13 years, a significantly higher detection rate of four of these bacterial species (with the exception being *P. intermedia*) was observed in children with Down syndrome compared to typically developing children.[1)] In another study targeting individuals aged 9–21years,[2)] only *P. gingivalis* and *A. actinomycetemcomitans* were evaluated. No significant difference in the detection rate of *P. gingivalis* was observed, while *A. actinomycetemcomitans* showed a significantly higher detection rate. On the other hand, in a study comparing adults aged 20–35 years with intellectual developmental disorder,[3)] no significant differences were found in the detection rates of all five bacterial species. Regarding *P. gingivalis*, a comparison of the distribution of *FimA* genotypes was also conducted, but no significant differences were observed. Similarly, in another study targeting adults with Down syndrome,[4)] no significant difference was found in the detection rates of periodontal pathogens when compared to individuals with intellectual developmental disorder. In contrast, a study of individuals with cerebral palsy[5)] reported that they had a higher rate of periodontal pathogen detection. A meta-analysis of these reports showed no significant differences in all bacterial species, and thus Down syndrome cannot be considered to be associated with a specific oral microflora. However, the level of evidence is very weak because these reports are not

consistent in terms of race, age, sample conditions, and bacterial detection methods. In recent years, periodontitis has come to be understood as being caused not by specific bacteria, but by a pathogenic bacterial community. In the future, analysis of the oral microbiome using next-generation sequencing is expected.

Literature

1) Amano A, Kishima T, Kimura S, et.al . Periodontopathic Bacteria in Children With Down Syndrome. J Periodontol 2000;71:249-55.
2) Barr-Agholme M, Dahllöf G, Linder L, et.al. Actinobacillus actinomycetemcomitance, Capnocytophaga and Porphyromonas gingivalis in subgingival plaque of adolescents with Down's syndrome. Oral Microbiol Immunol 1992;7:244-8.
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Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
40,351 (Pubmed), 21 (Cochrane Reviews)
#1. down syndrome
#2. periodontitis OR gingivitis
#3. Bacteria
#4. #1 AND #2 AND #3
Papers considered useful: 10

Medical Journal Search Results
9,581 (medical journal)
#1. down syndrome /TH or down syndrome /AL
#2. oral bacteria /TH or oral bacteria /AL #3. #1 AND #2
#4. (#3) and (PT= except conference proceedings) Papers considered useful: 0

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Periodontal Disease Management Team)

CQ14 Is toothbrushing instruction effective for individuals with Down syndrome?

[Perspective]
Toothbrushing instruction for individuals with Down syndrome is effective. (2D)

Background and Objectives

Individuals with Down syndrome are susceptible to periodontal disease from early ages. We investigated the effectiveness of toothbrushing instruction in Down syndrome.

Explanation

CQ outcomes included reduction of plaque accumulation, reduction of calculus (tartar) accumulation, maintenance of the current number of teeth, improvement in gingival inflammation, and reduction in caregiver burden. In the systematic review, one pre–post intervention study was identified for each of two indices: plaque accumulation and gingival inflammation. In the plaque-accumulation study, 135 institutionalized individuals with disabilities aged 12–46 years (mean 23.81 years), were enrolled, including 17 with Down syndrome. Weekly toothbrushing instruction for participants and oral-health education for caregivers were provided over a period of three months. Among the 10 individuals with Down syndrome who completed the program, the Simplified Debris Index (DI-S) decreased significantly.[1)]

In the gingival-inflammation study, 112 individuals with Down syndrome aged 11–22 years (mean 14.8 years) received toothbrushing instruction and oral-health education at school every two weeks for three months; a significant reduction in the Gingival Index was reported.[2)]

The former study involved instruction including caregivers, while the latter demonstrated effectiveness among individuals with Down syndrome who had mild intellectual developmental disorder. However, because each outcome was supported by only a single pre–post study and meta-analysis was not possible, the level of evidence is very weak. In actual clinical practice, toothbrushing instruction should be tailored to the person's level of intellectual development and degree of cooperation. Guidance for caregivers is also an important factor, taking into account caregivers' knowledge of oral diseases and care, feasible intervention techniques, caregiver manual dexterity, and life circumstances (including caregiver burden).

Further intervention studies are warranted to evaluate the effects of toothbrushing instruction and to examine in detail the confounding factors that influence outcomes for both patients and caregivers. In particular, intervention studies using standardized protocols based with detailed stratification by background factors are desirable.

Literature

1) Bizarra F, Ribeiro S. Improving toothbrushing behaviour in an institution for the disabled in Lisbon, Portugal Int J Dent Hyg 2009;7:182-7.
2) Shyama M, Al-Mutawa SA, Honkala S, et al. Supervised toothbrushing and oral health education program in Kuwait for children and young adults with Down syndrome. Spec Care Dentist 2003;23:94-9.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
40,351 (Pubmed), 21 (Cochrane Reviews)
#1. down syndrome
#2. toothbrushing instruction OR dental hygiene instruction OR toothbrushing
#3. #1 AND #2
Papers considered useful: 3

Medical Journal Search Results
9,581 (medical journal)
#1. Down syndrome /TH or Down syndrome /AL
#2. brushing instruction /AL or oral cleaning instruction /AL or (brushing /TH or brushing /AL)
#3. (#1 and #2) and (PT= except meeting minutes)
Papers considered useful: 0

Resolution Decision

Acceptable 13 Not acceptable 0 Correction required 0

(Periodontal Disease Management Team)

CQ15 Is basic periodontal therapy effective for adults with Down syndrome?

[perspective]
Scaling and root planing are effective in improving periodontal pockets in the periodontal treatment of adults with Down syndrome. (2C)

Background and Objectives

We will examine whether scaling and scaling root planing (SRP) are an effective part of basic periodontal therapy in people with Down syndrome, who are considered to be at high clinical risk of periodontal disease.

Explanation

In this CQ, to evaluate the effectiveness of basic periodontal treatment—e.g., scaling and SRP—the following outcomes were selected from the literature: "probing depth (PD)," "bleeding on probing (BOP) or gingival index (GI)," "clinical attachment level (CAL)," and "periodontal pocket depth (PD)." The "clinical attachment level (CAL)" was used as the outcome measure to evaluate the effectiveness of periodontal prophylaxis (PP).

In a study that evaluated the effect of scaling[1)] there was no improvement in PD, GI, or CAL to 12 weeks postoperatively. In studies that evaluated the effect of SRP up to 6 or 12 months postoperatively, PD, BOP or GI, and CAL were all significantly improved, respectively.[2-4)] These studies provided caregivers with instruction on the use of interdental cleaning appliances as well as toothbrushes, and plaque control was also improved. All of these studies also used mouthwashes with 0.12% or 0.2% chlorhexidine and chemical plaque control using 1.0% chlorhexidine toothpaste. In Japan, chlorhexidine is restricted to an undiluted concentration of 0.05%, so these SRP results must be treated only as a reference.

A meta-analysis was performed on one randomized controlled trial,[5)] two non-randomized controlled trials,[1,2] and one case series,[3)] in which the mean values of clinical parameters before and after treatment could be calculated. The results showed a statistically significant improvement in PD with a weighted mean of 0.73 mm and confidence interval of -1.30 to -0.15. This suggests that basic periodontal therapy for Down syndrome is effective in improving PD, but may be contingent on improved plaque control. All studies with improved clinical parameters continued to receive recalls at 4- to 6-week intervals during the study period, suggesting that maintenance therapy at 1- to 2-month intervals is effective in Down syndrome after periodontal treatment.

Literature

1) Cichon P, Crawford L, Grimm W-D. Early-Onset Periodontitis Associated With Down's Syndrome-A Clinical Interventional Study. Periodontol 1998;3:370-80.

2) Zaldivar-Chiapa RM, Arce-Mendoza AY, De La Rosa-Ramírez M, et al. Evaluation of Surgical and Non-Surgical Periodontal Therapies, and Immunological J Periodontol 2005;76:1061-5.
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5) Martins F, Simões A, Oliveira M, et.al. Efficacy of antimicrobial photodynamic therapy as an adjuvant in periodontal treatment in Down syndrome patients. Lasers Med Sci 2016; 31;1977-81.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search results
40,351 (Pubmed), 21 (Cochrane Reviews)
#1. down syndrome
#2. periodontitis OR gingivitis
#3. cleaning OR scaling
#4. #1 AND #2 AND #3
Papers considered useful: 5

Medical Journal Search Results
9581 cases (medical journal)
#1. down syndrome /TH or down syndrome /AL
#2. periodontal therapy /TH or basic periodontal therapy /AL and (except PT= meeting minutes)
#3. #1 AND #2
Papers considered useful: 0

Resolution Decision

Acceptable 13 Not acceptable 0 Correction required 0

(Periodontal Disease Management Team)

CQ16

Is Prosthetic treatment recommended for individuals with Down syndrome who have difficulty masticating due to tooth loss?

[Perspective]
Although prosthetic treatment has general utility, evidence to support a recommendation specific to Down syndrome is unclear. (3D)

Background and Objectives

Down syndrome is often characterized by earlier tooth loss primarily due to periodontal disease rather than dental caries, and anatomical/functional factors may affect eating and swallowing from early childhood. This clinical question evaluates whether prosthetic treatment of missing teeth is recommended.

Explanation

Individuals with Down syndrome may have difficulty self-managing oral hygiene because of delays in fine and gross motor development and challenges with coordinated movements. Caregiver-assisted oral care, including family support, is therefore important to maintain hygiene. Without adequate oral hygiene, periodontal disease may develop and lead to tooth loss. When prosthetic treatment is required for missing teeth, adaptation to the prothesis may be challenging.

Articles retrieved by the search strategy were case reports only, and no criteria for prosthetic treatment were identified.[1-6] Likewise, no clear criteria for implant surgery in Down syndrome were found, and no study verified whether implant placement and subsequent prosthetic restoration effectively restores masticatory function.

Prosthetic treatment in Down syndrome should follow evaluation of feeding/masticatory function and oral hygiene, weighing risks and benefits. Although there are reports of effectiveness in patients with intellectual developmental disorder (IDD),[7-11] device types and levels of treatment cooperation vary. Further research is needed to establish indications/criteria for prosthetic treatment in Down syndrome and to account for the degree of patient cooperation.

Literature

1) Alqahtani NM, Alsayed HD, Levon JA, et al. Prosthodontic rehabilitation for a patient with Down syndrome: A Clinical Report. J Prosthodont 2018;27:681- 7.
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3) Lin JD, Lin LP, Hsu SW, et al. Are early onset aging conditions correlated to daily activity functions in youth and adults with Down syndrome? Res Dev Disabil 2015;36C:532-536.
4) Yoshinari M, Shiiba S, Iwamoto M, et al. Application of dental implants

in patients with Down syndrome. Journal of Dentistry 2002;23:545-8.
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7) Ogasawara M, Izawa M, Watanabe K, et al. A Study on Removable Dentures for the Handicapped. Part 1: Possibility of utilizing dentures for the handicapped. Journal of Disability Dentistry 1986;7:42-53.
8) Ogasawara T, . Kasahara H, Fukuzawa Y, et al. A clinical Study on Removable Dentures for Handicapped. Part2: Concerning the Personality. Journal of Disability Dentistry. 1987;8:33-43.
9) Ogasawara T, Kasahara H, Hiraide Y, et al. A Study on Removable Dentures for the Handicapped. Part 3: Survey of Removable dentures to be utilized by the Handicapped . Journal of Disability Dentistry 1988; 9:25-34.
10) Shigehisa Akiyama, Takaaki Murayama, Taketoshi Morita, et al. Survey on Dentures in Institutionalized Mentally Retarded Patients: Actual Conditions of Denture Holding and Use. Journal of Disability Dentistry 1999;20:292-7.
11) Mori T, Takeda N, Egusa M, et al. Retrospective Study on Retention of Cast Crown Restorations in Dentistry for the Disabled. Journal of Disability Dentistry 2011;30:17-23.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane Library Search result
25,433 (Pubmed), 21 (Cochrane Reviews)
#1. “Down Syndrome”[MeSH]
#2. “Tooth Loss”[MeSH]
#3. “Prosthodontics”[MeSH]
#4. “Mastication”
#5. “Denture”
#6. “chewing”
#7. #4 OR #6
#8. #3 OR #5
#9. #7 OR #8
#10. #1 AND #2 AND #9
Papers considered useful: 3

Medical Journal Search Results
3,964 (Medical Journal)
#1. (Down's syndrome /TH or Down's syndrome /AL) and (PT= except conference proceedings)
#2. (tooth loss /TH or tooth loss /AL) and (PT= except meeting minutes)
#3. (chewing /TH or chewing /AL) and (PT= except meeting minutes)
#4. (*Organ Transplant /TH or Prosthetic /AL) and (PT= except meeting minutes)

#5. #2 or #3 or #4
#6. #1 and #5
Papers considered useful: 3

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Early Aging Prevention Team)

CQ17

Is nutritional guidance recommended to prevent weight gain or weight loss in individuals with Down syndrome?

[Perspective]
Although behavior change techniques can be incorporated into nutritional guidance, the evidence to support a recommendation specific to Down syndrome is unclear.
(3D)

Background and Objectives

Overweight and obesity are common in Down syndrome and pose health risks; conversely, with aging there is a risk of malnutrition and weight loss. This clinical question asks whether nutritional guidance is recommended to prevent weight gain or to address weight loss, and how such guidance should be tailored to the underlying cause.

Explanation

In a survey of families of infants with Down syndrome receiving nutritional guidance, 84% reported that their concerns were resolved after meeting with a dietitian. The content of consultations varied by age: at 5–6 months, weight was a common concern; at 7–8 months, feeding volumes and eating patterns; at 9–11 months, food ingredients; and at 12–18 months, weight and food seasoning/flavoring.[1)]

There are reports that weight control for children with disabilities, including Down syndrome, can be improved through nutritional management and exercise.[2)] In particular, nutritional assessment of obesity in schoolchildren has indicated that the energy consumption and micronutrient intake of children with Down syndrome are low.[3)] Therefore, nutritional guidance for obesity in Down syndrome should include limiting energy consumption, promoting well-balanced intake of vitamins and trace elements, and providing exercise guidance.[3)]

A case report describes a successful case of dietary control for diabetes in an adult patient. Behavioral change methods were used in small steps, starting at an acceptable level with those who were able to communicate with the patient. The program included dietary and exercise counseling, with advice to discourage impulsive food purchases.[4)]

Overall, nutritional guidance appears to be necessary from infancy. In adulthood, the nutritional guidance may be effective if the patient cooperates with the guidance, suggesting the importance of behavior modification methods.

Literature

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of a multidisciplinary weight management intervention for youth with disabilities. . Child Obes 2017;13:455-61.
3) Luke A, Sutton M, Schoeller DA, et al. Nutrient intake and obesity in prepubescent children with Down syndrome. J Am Diet Assoc 1996;12:1262-7.
4) Imai S, Matsuda M, Higashikawa C, et al. A case of Down syndrome type 2 diabetes mellitus with good glycemic control for 4 years by medical guidance considering mild intellectual developmental disorder. Pathophys Nutr 2010;13:239- 45.

Search expressions

Period: ~ August 31, 2021

Pubmed, Cochrane LibrarySearch results
25,433 (Pubmed), 21 (Cochrane Reviews)
#1. "Down Syndrome"[MeSH]
#2. "Body Weight"[MeSH]
#3. "Obesity"[MeSH]
#4. "Metabolic Syndrome"[MeSH]
#5. "Nutrition Therapy"[MeSH]
#6. "Nutritional care"[MeSH]
#7. #2 OR #3 OR #4 OR #5 OR #6
#8. #1 AND #7
#9. "prevention and control" [Subheading]
#10. #8 AND #9
Papers considered useful: 2

Medical Journal Search Results
3,964 (Medical Journal)
#1. (Down syndrome /TH or Down syndrome /AL) and (PT= except conference proceedings)
#2. (weight /TH or weight /AL) and (PT= except meeting minutes)
#3. (obesity /TH or obesity /AL) and (PT= except meeting minutes)
#4. (Metabolic Syndrome /TH or Metabolic Syndrome /AL) and (PT= except meeting minutes)
#5. (Nutritional guidance /TH or Nutritional guidance /AL) and (PT= except meeting minutes)
#6. (Nutrition Management /TH or Nutrition Management /AL) and (PT= except meeting minutes)
#7. #2 or #3 or #4 or #5 or #6
#8. #1 and #6
#9. #1 and #7
#10. (Preventive Management /AL) and (PT= except meeting minutes) #11. #8 and #10
#12. #9 and #10
Papers considered useful: 2

Voting decision

Acceptable 13 Not acceptable 0 Correction required 0

(Early Aging Prevention Team)

Column 2 Oral adverse effects of medications for age-related conditions in adults with Down syndrome

Direct evidence linking medication use in adulthood to oral health outcomes in people with Down syndrome is limited. However, systemic comorbidities tend to increase with age,[1-3] and polypharmacy may have secondary effects on oral cavity. Age-associated comorbidities commonly seen in older adults with Down syndrome include the following:

(A) Endocrine and metabolic diseases (e.g., thyroid dysfunction, hyperuricemia)
(B) Cardiovascular disease
(C) Respiratory disease (e.g., obstructive sleep apnea syndrome)
(D) Lifestyle-related disease (e.g., obesity, hyperlipidemia)
(E) Gastrointestinal disease
(F) Neurologic disease (e.g., epilepsy, Alzheimer's disease)
(G) Neurodevelopmental or psychiatric disorders (e.g., autism spectrum disorder, obsessive–compulsive disorder, depressive disorders)
(H) Muskuloskelatal disorders (e.g., cervical spine instability, osteoarthritis)
(I) Ophthalmologic disease
(J) Otorhinolaryngologic disease

Thyroid dysfunction is common in adults with Down syndrome—most often hypothyroidism. In the subset with hyperthyroidism, antithyroid drugs can (rarely) cause agranulocytosis, which increases susceptibility to severe infections of the oropharynx and neck.[4-6] In regard to other age-associated conditions, myelosuppression (e.g., pancotypenia, aplastic anemia) and agranulocytosis have been reported with certain medications for hyperuricemia; agranulocytosis has also been described with some antirheumatic sulfonomides and antithrombotic agents. Agranulocytosis may present with recurrent oral aphthae and bacterial infection, and pancytopenia can exacerbate periodontal disease, contributing to deterioration of oral health.

In adults with Down syndrome, neurodevelopmental and psychiatric comorbidities are common (e.g., autism spectrum disorder, obsessive–compulsive disorder, and depressive disorders),[7,8] and related medications are frequently prescribed. Antidepressants often cause xerostomia; antipsychotics can produce movement disorders that impair oral hygiene; and sedative effects across these drug classes may reduce brushing effectiveness. Epilepsy affects an estimated 5-10% of individuals; antiepileptic drugs may lead to gingival overgrowth, xerostomia, dyskinesia, and feeding and swallowing difficulties. Although uncommon, Down syndrome–associated arthritis[9] may require long-term corticosteroids, immunosuppressants, or biologic agents, which can increase the risk of stomatitis and periodontal deterioration. As adults with Down syndrome age, medication exposure typically increases. Dental teams should monitor for medication-related oral adverse effects (e.g., xerostomia, gingival overgrowth, oral mucosal ulceration), reinforce daily oral hygiene, optimize fluoride use, adjust recall intervals as needed, and coordinate with physicians—particularly if neutropenia is suspected or before invasive procedures.

Literature

1) Takeuchi C, Tamai H, Ueda K, et al. A Guide to Health Care Transition Support for People with Down Syndrome in Japan. https://japandownsyndromeassociation.org/wp-content/uploads/2021/04/jdsa-transition healthcare-guide.pdf （ref. 2022-06-06）
2) The Manuals for Management of Individual Serious Adverse Drug Reactions(The Ministry of Health, Labour and Welfare). https://www.mhlw.go.jp/stf/seisakunitsuite/bunya/kenkou_iryou/iyakuhin/topics/tp061122-1.html （ref. 2022-06-06）)
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(Early Aging Prevention Team)

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