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An Overview of Frequency Malocclusion in Cases of Down Syndrome Children: A Systematic Review

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ABSTRACT

Background: Malocclusion is one of the important problems in the field of dental and oral health in Indonesia. Dental and oral health in children is an important factor that must be considered as early as possible. Therefore, knowledge of children, especially knowledge of malocclusion, must be considered more because if a children experiences malocclusion it can affect tooth grow that a laterage. Down Syndromeis a chromosomal disorder thatresults in mental retardation. Mental retardationis a worldwide problem with major implications, especially for developing countries. It is estimated that the incidence of severe mental retardationisabout 0.3% ofthe total population, andnearly 3% havean IQ below 70%. As human resources, ofcourse they cannot be utilized, because 0.1% of these children need care, guidanceandsupervisionthroughouttheirliving.Objective: To review the frequency of malocclusion in DownSyndromechildren. Methods: Scientific evidence and clinical cases were drawn from the literature to support this review and information on the frequency of malocclusion in childrenwith Down Syndromewascollected.Result/Discussion: There are several types of malocclusion that often occur in children with DownSyndrome. Some of them are associated with class III Anglemalocclusion, crowding, and anterior open bite. Conclusion:Down Syndrome is one of the most studied genetic syndromes because of its frequency in our population and its medical significance. Malocclusion was observed in 92% of subjects with Downsyndrome. The incidence of malocclusion is higher in children/adolescents with Down Syndrome than in individuals without the syndrome. Class III malocclusion is most frequently observed.

Keywords: Malocclusion, DownSyndrome, Angle'sMalocclusion

1. INTRODUCTION

Malocclusion is one of the important problems in the fieldo f dental and oral health in Indonesia. Dental and oral health in children is an important factor that must be considered as early as possible. Therefore, knowledge of children, especially knowledge of malocclusion, must be considered more because if a child experiences malocclusion it can affect tooth grow that a later age. Malocclusionis a condition that deviates from normal occlusion including irregularity of the teeth in thearch of the jaw, such as crowding, protrusion, malposition or an unharmonious relationship with the antagonistic teeth. andmandibular teeth.[1,2]

The occurrence of malocclusion is very much influenced by heredity inherited fromparentsandenvironmentalfactorssuch as bad habitsand diet. Usuallythesetwofactorsmanifest as animbalance in thegrowthanddevelopmentofthedentofacialstructuresresulting in malocclusion. The influenceofthesefactorscandirectlyorindirectlyleadtomalocclusion. Heredity has a majorinfluenceonmalocclusion, forexamplethesize, shape, and number of teeth that grow out of line with the arch of the jaw, causing crowding of teeth. Children still havephysicalandpsychologicallimitations, in accordancewithongoinggrowthanddevelopment. Itis not uncommon for children who are in theirinfancy, have problems with their teeth. Malocclusioniscommonatages 9-12 years, thisisthesecondphaseofthemixedtoothperiod. During this period there was a change in dimensions from primary teeth to permanent teethwhichcausedmanyproblems. Occlusionsometimesbecomesunsuitable, resulting in overcrowdedteeth, crossbites, open bites, deepbites, andpermanenttoothlossdueto caries.[3]

DownSyndromeis a chromosomal disorder thatresults in mental retardation. Mental retardationis a worldwide problem withmajorimplications, especiallyfordevelopingcountries. Itisestimatedthattheincidenceofsevere mental retardationisabout 0.3% ofthe total population, andnearly 3% havean IQ below 70%. As human resources, ofcoursetheycannotbeutilized, because 0.1% ofthesechildrenneedcare, guidanceandsupervisionthroughouttheirliving. The frequencyofDownsyndromeincreaseswithincreasingageofthemother. Whilethe disorder occurred in only 0.04% (4 in 10,000 births) ofchildrenborntowomenunder 30 yearsofage, theriskroseto 0.92% (92 outof 10,000) formothersaged 40 andevenhigherforoldermothers. Downsyndrome'scorrelationwith maternal age has not been explained.[4]

Downsyndromewasfirstdescribedby a doctorfromEnglandnamedLangdonDown in 1866.[1,2] Thissyndromeiscausedby a congenitalanomalyofautosomalchromosomes (non-sex chromosome) onchromosome 21.[1-3]Downsyndromeis a chromosomal disorder atbirththatisquitecommon in theworld. WHO reports, theestimatedincidenceisbetween 1 in 1,000 to 1 in 1,100 births. There are currently 8 millionsufferers in theworld. Eachyearanestimated 3,000 to 5,000 children are bornwithDownsyndromeandthere are about 250,000 families in the United States whereonememberofthefamily has this syndrome.[3]The latest data fromthe Basic HealthResearch(2013) statesthat until nowtherehavebeen 300,000 cases of children with DownSyndrome in Indonesia.[5]

2. MATERIALS AND METHODS

Scientific evidence and clinical cases were drawn from the literature to support this review and information on the frequency of malocclusion in Down Syndrome children was collected.

LITERATURE SEARCH

A systematic review of the literature was carried out looking for all publishedarticlesontherelationshipof dental conditionstotheincidenceofstunting in children. On December 21st, 2020, a literaturesearchwascarriedoutusingthefollowingkeywords: "*Malocclusion in children, Downsyndrome, MalocclusionandDownsyndrome, Dental and oral healthforchildrenwithDownsyndrome*." The followingdatabases were searched: PubMedandGoogleScholar.

3. DISCUSSION

1. Definition of Malocclusion

There are various diseases and disorders that affect tooth function, one of which is malocclusion. Malocclusionis a formofocclusionthatdeviatesfromthestandardformthatisaccepted as a normal form. Malocclusionisalso a conditionthatdeviatesfrom normal occlusionincludingirregularityoftheteeth in thearchofthejaw, such as overcrowded, protrusive, malpositionordisharmoniousrelationshipswiththeantagonistteeth. Occlusionissaidtobe normal iftheteeth are arranged in an orderly arch and there is a harmonious relationship between the maxillary and mandibular teeth.[3,6]

2. EtiologyofMalocclusion

KusyolaConstantyalsoexplainsthecausesoftoothdeformities in children, which are as follows:

a. Bad habits

A habitis a certainactionthatisrepeated over and over again, while oral habitis a habitthatcancausechanges in occlusalrelationshipssuch as suckingand biting thelips, pushingthejawforward, pushingthetongue, or biting nails.

b. Genetics

For example, mothers who have small teeth and fathers who have large jaws, tend to have children with small jaws and large teeth, automatically causing the teeth to crowd.

c. Trauma

Hard impact on the mouth and injuring the jaw and teeth is also a cause of malocclusion. Environmental factors that can cause malocclusion include disease, nutritional status, and oral habits.[7]

3. Definition of Down Syndrome

Understanding Down Syndrome, syndrome is defined as a symptom or sign that appears together (Alwi, 2002:1069). Meanwhile, the word down used in this case is a term taken from doctor from England, namely John Langdon Down. Kosasih (2012:79) states that Down syndrome is a condition of underdevelopment in the physical and mental development of children caused by abnormalities in chromosomal development. Chromosomes are special fibers that are present in every cell in the human body, where there is genetic material that determines a person's characteristics. Wiyani (2014: 113-114) complements the explanation that Down syndrome occurs because of an abnormality in the 21st chromosome arrangement of 23 human chromosomes. In normal humans, 23 chromosomes pair up to 46. In people with Down syndrome, the number 21 chromosome is three (trisomy), bringing the total to 47 chromosomes. This excessive amount results in shocks in the cellular metabolic system, which eventually leads to Down syndrome.[8]

4. Epidemiology of Down Syndrome

Epidemiologyisthe study ofthepatternsandcausesofhealth-relatedtraits in a givenpopulation. The results of these studies form the basis forinterventionaltreatment. For Downsyndrome (DS), suchepidemiologicalstudiesbegan in the mid-1800s when several physicians described a group of patients, who had mental retardation and short stature alongwithcertainfacialcharacteristics, includingslantedeyeslits, epicanthalfolds, flat nosebridges, andprotrudingtongue. J. LangdonDown, forwhomDown syndromeisnamed, makes a significantcontributiontotheepidemiologyofthesyndromebyemphasizingthatthis set of clinical findingsis a distinctentity, and that affected individuals can be distinguished from a

heterogeneousgroupofallpersonswithintellectualdisabilities. In an excellent review of Down syndrome's history, Rynders and Pueschel continued the story of Down syndrome's recognition in to the late 1800s andearly 1900s.

Now, Down syndrome ortrisomy 21 is one of the most studied human aneuploid conditions. This review will focus on the current literature relating to two distinct, butrelated, epidemiological fields of Down syndrome. First, we will review the prevalence of Down syndrome and its associated risk factors. We will focus on meioticnond is junction of chromosome 21, the most common cause of Down syndrome. Second, we will review studies on the prevalence of birth defects and abnormalities associated with Down syndrome. As each topic deservesanentirejournal, wewillconcentrateon a fewexemplarystudiestoillustratetheprogressmade in the field and tooutline a potential genetic epidemiological study design capable of uncovering thebiologicalmechanismsunderlyingchromosome 21 nondisjunctionanditsclinical consequences.[9]

5. Malocclusion in DownSyndromeChildren

Table 1. Malocclusion in DownSyndromechildrenbysomeresearch

No.	Author and Titles	Year	Result and Conclusion
1.	Siti Salmiah, Nurul Sukma Mustafa DescriptionofMalocclusiona nd Bad Habitsof 6-18 YearsofDownSyndromePati ents in Medan City. Journal: Dentika Dental Journal, Vol 19, No. 1, 2016: 42-46	20 16	Result: The resultsofthis study foundthattheprevalenceofmalocclusionbasedonAn gle'sclassification in Downsyndromechildrenaged 6-18 years in specialschools in Medan City was 31.71% ofthechildren had Class I molar relationships, 3.66% Class II, and 48.78% Class III. The mostcommonformsofmalocclusion were anterior crossbites, namely 42.68%, followedbycrowding 39.02%, anterior open bites 23.17%, and posterior crossbites 21.95%. The highest bad habits were tonguethrusting, namely 41.46%, mouthbreathing 40.24%, bruxism 37.8%, fingersucking 36.58%, andnailorfinger biting 21.95%. Conclusion: The prevalenceofmalocclusionand bad habits in Downsyndromechildrenisquitehigh. Thisneedstobe a concernofparents/guardians/caregiverstominimize these bad habitssothatchildren'schewingandphoneticscanfun ctionoptimally. This study showsthatthehighprevalenceofthese bad habitsleadsto a highprevalenceofmalocclusion in peoplewithDownsyndrome.
2.	Bauer Danielle, etal.	20 12	Result: The DownSyndrome (DS) group had

	SeverityofOcclusalDisharmonies in DownSyndrome Journal : International JournalofDentistry Volume 2012, Article ID 872367, 6 pages		significantlyhigher PAR and ICON scores, as well as 10 times. moreteeth were lostthan in the non-Down syndromegroup. The Down syndromegroupwaspredominantlyClass III malocclusion, withthepresenceofboth anterior and posterior crossbites in themajorityofpatients. The non-Down syndromegroupmostlyexperiencedClass I or II malocclusionwithmissingteethandfewercrossbites. The Down syndromegroupalso had moreseveremalocclusionbasedonocclusaltraitssuch asopen biteandmalocclusiontypes. Conclusion: The Down syndromegroup had veryseveremalocclusion, whilethecontrolgroupfromtheuniversityclinic had moreseveremalocclusionthanthecontrolgroupfrom privatepractice.
3.	Victor Paulo, etal. Malocclusion in childrenandadolescentswith Downsyndrome: A systematicreviewand meta-analysis Journal: Int J PaediatrDent. 2019 Jul;29(4):524-541.	2019	Result: Elevenpublications were included in thesystematicreviewandeight were meta-analyzed. Meta-analysisshowedthatmalocclusionwasmoreprevalent in children/adolescentswithDown syndromeforClassIII Angle malocclusion (riskdifference [RD]=0.40; confidence interval [CI]=0.33, 0.46), posterior crossbites. (riskratio [RR]=3.09; CI=2.02, 4.73), anterior crossbite(RR=2.18; CI=1.41, 3.39), and anterior open bite (RD = 0.21; CI = 0.06, 0.36). Conclusion: The incidenceofmalocclusionishigher in children/adolescentswithDown syndromethan in individualswithoutthesyndrome. The strengthoftheevidencefromtheanalyzedstudies, however, isconsideredmoderateandlow.
4.	MestrovicSenka, etal. PrevalenceofMalocclusion in PatientswithDown'sSyndrome	2002	Result: Malocclusionwasfound in 92% ofsubjects. Class III malocclusionwasobservedmostfrequently (43.8%). Unilateral crowdingandcrossbite were found in 15% ofsubjects. Bilateral crossbites were found in 5.4% ofsubjects. Prematuretoothlosswasobserved in only 1%

	Journal: Acta StomatolCroat, Vol. 36, br. 2, 2002.		ofsubjectswhereasClass II division 2 malocclusionwas not recorded in theexaminedsubjects. Conclusion: Malocclusionwasobserved in 92% ofsubjectswithDownsyndrome. Class III malocclusionwasmostfrequentlyobserved, namely in 43.8% ofcases. Unilateral crowdingandcrossbite were found in 15% ofsubjects. Bilateral crossbites were found in 5.4% ofsubjects. Prematuretoothlosswasobserved in only 1% ofsubjects.
5.	Marques Leandro Silva, etal. Downsyndrome: a riskfactorformalocclusionseverity? Journal : Braz Oral Res [online]. 2015;29(1):1-7	2015	Result: As shown in Table 1, individualthecontrolgroup had a greaterincidenceofdiastema (≥ 2 mm) andoverjet (≥ 4 mm). Differences were alsofoundwithrespecttofacetypeandAngleclassification betweenthetwogroups. The shortfacepatternandClass III Anglemalocclusion were morefrequentamongindividualswithDown syndrome, whereasthe long facepatternandClass II Anglemalocclusion were morefrequentamongindividualsin thecontrolgroup (Table 1).Malocclusionseveritywasgreateramongindividualswith Down syndromecomparedwiththose in thecontrolgroup (p=0.028). Takingintoaccount individual, social, andbehavioralfactors, therewas a greaterfrequencyofseveremalocclusionamongindividualsaged>10 yearsandamongthosewith a historyofpretermbirth, lipdisability, mouthbreathingpattern, andfacialpatternlength (Table 2). Conclusion: Verticalandtransversechanges in occlusion,such as mandibularprotrusions, anterior open bites, and posterior crossbites were significantlymorefrequentamongindividualswithDown syndromethan in thecontrolgroup. Relateddeterminantfactorstheseverityofthemalocclusionwas Down syndrome, historyprematurebirth, and long facialpatterns.

Based on the results of the studies included in this review it seems that there is a relationshipbetweenthestateoftheteethandtheincidenceofstunting in children. Thisseemstobeshown in theresultsofotherstudiesby Siti Salmiah and Nurul Sukma Mustafa in 2016. In this study, themalocclusiontobestudiedincludesmalocclusionof molar relationshipsbasedonAngle'sclassificationandvariousgeneralformsthatcanbeobservedclinicallysuch as anterior and posterior crossbites, bites. anterior opening, andcrowding, andthetypesof bad habitscommonlyexperiencedbychildrensuch as mouthbreathing, tonguethrusting, nailorfinger biting, fingersucking, andbruxism. The mostcommonformofmalocclusion in Down'ssyndromechildrenafter anterior crossbitewascrowding, whichwas 39.02%. Theseresults are consistentwiththeresultsofresearchconductedbyMestrovicetal.whichplacescrowding as thesecondhighestformofmalocclusion in Down'ssyndrome children.[10]

The samethingseemstobeshown in thesecond study byBauer Danielle etalin 2012. In this study, discussingthepossibilityofmalocclusionexperiencedbyDownsyndromesufferersdueto a different palatal shape in individualswith DS than in the normal populationduetoitsnarrowarchand V-shape. A narrowmaxillaand normal transversedimensionsofthemandible are possibleetiologiesof posterior crossbite, eitherunilaterallyorbilaterally. In a study byUongetal.They foundthatthesizeofsofttissuessuch as thetongueandsoftpalate in Down syndromewascomparable in sizeto normal childrenofthesameage, butthatthewidthanddepthofthehardpalate were reduced. Therefore, thegeneralunderdevelopmentofthemaxillaandpalatineappearstopushagainstthetongue, andmakesitimpossibletodevelopthemaxilla as occurs in normal tongue posture.[11]

In a third study by Victor Paulo, etal. in 2019, italsoshowedthatDown syndromeofchildren/adolescentsshowed a higherprevalenceofclass III Anglemalocclusion, posterior, and anterior crossbiteand anterior open bite. This study showednodifferencesbetweengroupsregardingAngleclass III malocclusionandcrowding. Severalfactorsreported in theliterature are associatedwith a higherprevalenceofmalocclusionamongDown syndromeindividuals. Anterior crossbiteandClass III Anglemalocclusion are commonocclusalfeatures in Down syndromechildren/adolescents. Thesecharacteristicsmaybeassociatedwithmaxillaryhypoplasticityandmandibularprognathism, thecraniofacialfeaturescommonlyobserved in Down syndromeindividuals. Indeed, theprimarybone disorder affectingtheorofacialstructures in Down syndromeisunderdevelopmentorhypoplasiaofthemidface area.[12]

In thefourth study byMestrovicSenka, etal. in 2002. This study alsodiscussestherelationshipbetweenmalocclusion in peoplewithDownsyndromebasedon gender. In this study Class III malocclusionwasfoundtooccurtwice as often in male subjectsthan in womenwithDownsyndrome. In contrast, thefrequencyof unilateral and bilateral crossbites in womenwasthreetimesthatofmen. The frequencyofothermalocclusionswasthesame in bothsexes. In the total numberofmalocclusions, Class III malocclusionwasobservedmostfrequently, namely in 47.6% ofcases (in 36.9% and 10.7% in boysandgirls, respectively).[13]

And in a recent study byMarques Leandro Silva, etal. in 2015. In contrasttoseveralpreviousstudieswhichsuggesttherelationshipbetweentheincidenceofstuntingandthepresenceofcariesanddescribethe oral healthofchildren. In this study, theauthorswrotethatthemostfrequentmalocclusionsamongindividualswithDown syndrome were mandibularprotrusion, anterior open bite, and posterior crossbite. This finding confirms the results of another study that reported high prevalence values formal occlusions stemming from transverse and verticalocclusalchanges. Such changes are associated with in sufficientdevelopmentofthebones, hypotoniaoftheorofacialmusclesandthepositionofthetongue.

Hypotoniamuscles, which are associatedwithdecreased oral volume and are characterizedby a deepatresicpalate, canleadto a habitualtendencytostickthetongue over theteethoroutsidethemouth. DS sufferers generally have a short facial patternand reduced development of the middle third of the face, resulting in an Class IIIAnglemalocclusion relationship.[14,15,16,17,18,19,20]

4. CONCLUSION

Down Syndrome is one of the most studied genetic syndromes because of its frequency in our population and its medical significance. Malocclusion was observed in 92% of subjects with Downsyndrome. The incidence of malocclusion is higher in children/adolescents with Down syndrome than in individuals without the syndrome. Class III malocclusion is most frequently observed.

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