

Prevalence of Macroglossia in Children with Down Syndrome in Dental and Oral Hospital of Unpad FKG

Rinda Prawidiastuti¹, Inne S. Sasmita^{2*}, Yetty Herdiyati², Ratna Indriyanti², Willyanti Soewondo²

1. Student of Dentistry, Faculty of Dentistry, University Padjadjaran, Bandung, Indonesia.

2. Department of Pedodontics Dentistry, Faculty of Dentistry, University Padjadjaran, Bandung, Indonesia.

Abstract

Macroglossia is a condition in which large tongues can interfere with the quality of life of the sufferer, such as difficulty in eating, drinking, speaking, respiratory disorders, and dental problems. Down syndrome is a genetic disorder caused by the trisomy of chromosome 21 which results in various abnormalities in hard tissue and soft tissue. The purpose of this study is to determine the prevalence of macroglossia in children.

This research was conducted using the descriptive method with selective prevalence technique. A sample of 30 children with Down syndrome was obtained by total sampling technique. The study was conducted by looking at the size of the tongue at rest position using glass mouth.

This study obtained the prevalence of macroglossia in children with Down syndrome at Dental and Oral Hospital of FKG Unpad 53,3%.

The prevalence of macroglossia in children with Down syndrome at Dental and Oral Hospital of FKG Unpad is 53.3%, where more than half of the population of children with Down syndrome in Dental and Oral Hospital of FKG Unpad have macroglossia.

Clinical article (J Int Dent Med Res 2018; 11(3): 911-915)

Keywords: Macroglossia, Down Syndrome, Craniofacial.

Received date: 08 October 2017

Accept date: 12 January 2018

Introduction

Down syndrome is a genetic disorder resulting from chromosomal abnormalities caused by trisomy of all or most of the chromosome 21.¹ Down syndrome children have extra chromosomes in all body cells with a total of 47 rather than 46 chromosomes.²

Down syndrome is the most common chromosomal disorder and causes mental retardation.^{4,5} Down syndrome events are estimated to occur between 1 in 1000 live births.⁶ In the United States, there are 400,000 children with Down syndrome.⁷

According to Riskesdas in 2013, the down syndrome rate has a value of 0.12% in 2010 and an increase of 0.13% in 2013. The Association of Parents of Children with Down Syndrome

(POTADS) reported there were more than 300,000 cases of syndrome down in Indonesia.^{8,9}

Children with Down syndrome show various findings of physical characteristics, including abnormalities in the craniofacial and oral cavity.⁵ One of the most common disorders of the oral cavity is macroglossia.¹⁰ According to Gullikson's research in 1973, it was reported that the finding of macroglossia in childhood syndrome down about 61%. In Chennai India, found 52 children from 70 children with Down syndrome had a macroglossia whose percentage was about 83.6%.^{11,12}

Macroglossia can occur congenitally and can be. Congenital macroglossia can be due to muscle hypertrophy, vascular malformations, congenital hypothyroidism, and chromosomal abnormalities such as down syndrome, whereas macroglossia can be caused by passive tongue enlargement if the lower teeth are lost, metabolic disorders, inflammatory conditions, and allergic reactions.¹³⁻¹⁵

Macroglossia can cause difficulties in speech, mastication, swallowing, tongue movements and respiratory disturbances, so in children with special needs such as Down

*Corresponding author:

Inne S. Sasmita
Department of Pedodontics Dentistry,
Faculty of Dentistry,
University Padjajaran,
Bandung, Indonesia.
E-mail innesuhernasasmita@yahoo.co.id

syndrome, this cause will affect the quality of life of the child.^{5,14} According to Proffit in 1978 states that macroglossia gives a great force in the dental arch, causing the shifting of teeth which will cause dental problems. Such dental problems include malocclusion, anterior open bite, upper and lower incisor proclination, diastema, temporomandibular and maxillofacial joint disorders.^{14,15,16}

A prominent tongue due to macroglossia may affect bone growth and may lead to class III skeletal disorders. Research conducted by Harvold in 1972 proves that the tongue affects the growth and development of the jaw and teeth.^{14,15,17}

Based on the description above, this study was conducted with the aim to determine the frequency of macroglossia in children with Down syndrome at Dental and Oral Hospital Faculty of Dentistry Padjadjaran University.

Materials and methods

The research was conducted at RSGM FKG Unpad in November 2016 by using the descriptive method with selective prevalence approach in children with Down syndrome. The data used are primary data by checking the size of the tongue of the child syndrome down at rest position using glass mouth. The criteria of this research are children with Down syndrome in RSGM FKG Unpad and willing to be examined or have got permission from their parents. The method used in sampling is total sampling. Research is done by first making a letter of ethics and research permit. After that, the researcher chose the samples to be studied, gave informed consent, prepared the tools and materials, and the examination sheet, then the data collected, recorded, processed and analyzed using descriptive data analysis and presented in tabular form.

Results

Research on the prevalence of macroglossia in down syndrome children at Dental and Oral Hospital Faculty of Dentistry Padjadjaran University was conducted in November 2016. The sample obtained as many as 30 children; consisting of 16 boys and 14 girls.

Macroglossia	Frequency	Percentage
Yes	16	53,3 %
No	14	46,7 %
Total	30	100 %

Table 1. Frequency of Children with Down Syndrome who have Macroglossia in RSGM FKG Unpad

Table 1 shows that of 30 study subjects there were 16 children (53.3%) who had macroglossia and 14 children (46.7%) who did not have macroglossia, which means 8 out of 15 down syndrome children in RSGM FKG Unpad had macroglossia with a percentage of 53.3%.

Age (Year)	MACROGLOSSIA			
	Yes	%	No	%
<1	0	0	1	7.14
1 - <5	5	31.25	8	57.14
5 - <10	4	25	4	28.57
10 - <15	4	25	1	7.14
15 - 17	3	18.75	0	0
Total	16		14	

Table 2. Frequency of Children with Down Syndrome who Have Macroglossia by Age at RSGM FKG Unpad

Table 2 shows that of 16 children with Down syndrome at RSGM FKG Unpad who have macroglossia, there are 5 children (31.25%) aged 1- <5 years, 4 children (25%) aged 5- <10 years, 4 children (25 %) aged 10- <15 years, and 3 children (18.75%) aged 15-17 years. Based on the above data, it can be seen that the child syndrome down at RSGM Unpad FKG which has the most macroglossia are children aged 1- <5 years with a percentage of 31.25%.

MACROGLOSSIA	Sex			
	L	%	P	%
Yes	9	56.25	7	50
No	7	43.75	7	50
Total	16		14	

Table 3. Frequency Distribution of Children with Down Syndrome who have Macroglossia by Sex in RSGM Faculty of Dentistry Padjadjaran University.

Table 3 shows that 56.25% of children with male sex down syndrome have macroglossia in their oral cavity, whereas children with female down syndrome who have 50% of macroglossia, it can be concluded that the boy's syndrome down men has more macroglossia than women.

Discussion

Based on the results of research in Table 4.1 can be seen that of 30 children with syndrome down at RSGM Faculty of Dentistry Padjadjaran University there are 16 children (53.3%) who have macroglossia. The results of this study are similar when compared with research conducted in 2015 by Rahul et al. in a special school for mental retardation in Chennai India, in 70 children found 52 children had macroglossia with a score of 83.6%. Another study by Gullikson in 1973 reported that there were about 61% of children with Down syndrome who had macroglossia. Results from both Gullikson and Rahul et al. similar to this study, that of the more observed syndrome children who had more macroglossia than those who did not have macroglossia.^{11,12}

The results of this study indicate that more than half the population of children with Down syndrome at Dental and Oral Hospital of FKG Unpad has macroglossia, where this research is done by looking at the size of the tongue clinically using the glass mouth at rest position, if the lateral edge of the tongue cover \geq 1/3 surface occlusal of the mandibular teeth, then the tongue includes macroglossia.¹³

Down syndrome patients have impaired growth and physical development that lies in the craniofacial, orofacial muscles and tongue muscles.^{5,18,19} Craniofacial disorders include brachycephaly, maxillary hypoplasia, short mandibular bodies, and ramus, as well as low space of the palate causing the oral cavity in small down syndrome children, whereas disorders of the orofacial and lumbar muscles include the hypotonic orofacial muscles and muscles of the tongue. Orofacial muscle lies in the muscles of the orbicularis, zygomatic, masseter, temporalis, whereas the muscles of the tongue lie in the intrinsic muscles and extrinsic muscles of transverse, longitudinal, vertical, palatoglossus, styloglossus, genioglossus, and hyoglossus. Orofacial muscle

and hypotonic tongue muscles with small oral cavities cause the tongue in children with large down syndrome or macroglossia.^{5,18,20,21}

Macroglossia cannot be possessed by a child with Down syndrome, as most of the childhood macroglossia belonging to the Down syndrome includes relative macroglossia or pseudo macroglossia, which is a normal-sized tongue but because of the delay of growth of the jaw causing the cavity of the smallmouth that shows the tongue like macroglossia. The severity of the hypotonic muscle affects the enlargement of the tongue in the child's down syndrome, as the weaker, the muscle becomes, the less it prevents the movement of the tongue so that the tongue may become larger.^{22,23}

The results of the research contained in Table 4.2 shows that children with Down syndrome who have the most macroglossia in RSGM FKG Unpad are children aged 1- <5 years. This is because at this age children's syndrome down does not do much social interaction such as speech, and the existence of interference in the language ability of the child's down syndrome causes the child's syndrome down more using nonverbal communication, so that muscle hypotonus found in children with Down syndrome trained that causes the tongue in a child's syndrome down macroglossia.²⁴

Dieguez's research in 2008 explained that children with Down syndrome at mealtimes tend to swallow food without chewing for too long, and the food consumed by children at the age of 1- <5 years is generally soft-textured and the portion of food consumed is small, this will affect the hypotonus orofacial muscles and tongue muscles that belong to the child's Down syndrome. This is in accordance with the research of Van Trotsenburg et al. in 2005 which explains that in children with 2 years of age the syndrome is found muscle hypotonus and poor muscle stiffness so that the muscles cannot control the movement.^{25,26,27}

The results of the research contained in Table 4.3 shows that children with Down syndrome in RSGM Faculty of Dentistry Padjadjaran University who have the most macroglossia are children of the male sex. This happens because the incidence of the syndrome is more prevalent in men than in women, this is due to meiotic disorders in fathers affecting the sex chromosomes or at the conception of accessibility of sperm carrying a larer Y

chromosome to the ovaries so that the child's syndrome down is gender male.^{28,29}

The prevalence of sex-based down syndrome has been studied in several countries, as in China studied by Huang et al. in 1967, in Japan investigated by Tonomura et al. in 1966 and Slavin et al. in 1967, in France investigated by Bernheim et al. in 1979 and Nielsen et al. in 1981, in Sweden studied by Lindsten et al. in 1981, in Canada studied by Uchida in 1970, and the United States investigated by Chitham and MacIver in 1965, Huether in 1990, Petersen et al. in 1993, as well as by Griffin et al. in 1996, all of these studies showed that the prevalence of sex-based down syndrome was more prevalent in males than in females and because the down syndrome showed various abnormalities in hard tissue and soft tissue so that from this disorder will affect the tongue in the child's syndrome down male sex.³⁰⁻⁴¹

Macroglossia causes articulation of speech disorders, difficulty eating and swallowing, moving the tongue, respiratory disorders, and dental problems, where this cause will affect the quality of life of children down syndrome, so parents, teachers/faculty, and health practitioners need to pay attention to children with Down syndrome has a macroglossia in its survival from daily activities to the planned treatment to cope with the macroglossia.^{5,14}

Conclusions

Based on the research that has been done on the down syndrome children at Dental and Oral Hospital of Faculty of Dentistry Padjadjaran University, it can be concluded that the prevalence of macroglossia in children with Down syndrome in Dental and Oral Hospital of Faculty of Dentistry Padjadjaran University is 53,3%, where more than half of population of child syndrome down in Dental and Oral Hospital Faculty of Dentistry Padjadjaran University has macroglossia.

Declaration of Interest

The authors report no conflict of interest and the article is not funded or supported by any research grant.

References

1. Al-shawaf R, Al-faleh W. Craniofacial characteristics in Saudi down Syndrome. King Saud University J Dent Sci. 2011;2(1-2):17-22.
2. Moeckel E, Mitha N. Textbook of Pediatric Osteopathy. 1st Edition. English: Churchill Livingstone Elsevier. 2008:43.
3. Alper Avci, Refik Ulku, Serdar Orat. Chylothorax with Down Syndrome: Unusual Case Report. J Int Dent Med Res. 2009;2(1):25-7.
4. Soetjiningsih. Tumbuh Kembang Anak. Cetakan I. Jakarta: EGC. 2000:211.
5. Mubayrik AB. The Dental Needs and Treatment of Patients With Down Syndrome Oral Health Dental Management Systemic Considerations. Dent Clin North Am 2016;60(3):613-26.
6. World Health Organization. www.who.int. (Late access date 9/18/2016).
7. National Down Syndrome Society. www.ndss.org. (Late access date 9/18/2016).
8. Basic Health Research. Agency for Health Research and Development Ministry of Health RI. www.depkes.go.id. (Late access date 9/18/2016).
9. Parents' Association of Down with Syndrome (POTADS). www.potads.or.id. (Late access date 9/18/2016).
10. Diéguez AM. Editorial Macroglossia and Down syndrome. Int Med Rev Down Syndrome. 2008;12(2):17.
11. Gullikson J. Oral findings in children with Down's syndrome. J Dent Child. 1973;40(4):41-50.
12. Rahul VK, Matthew C, Jose S, Thomas G, Noushad MC, Feroz MTP. Oral Manifestation in Mentally Challenged Children. J Intl Oral Health. 2015;7(2):37-41.
13. Langlais RP, Miller CS. Atlas Berwarna Kelainan Rongga Mulut yang Lazim (Color Atlas of Common Oral Diseases). Cetakan I. Jakarta: Hipokrates. 2000:42-44.
14. Farronato G, Salvadori S, Giannini L, Maspero C. Congenital macroglossia: surgical and orthodontic management. Prog Orthodont. 2011;13(1):92-8.
15. Topouzelis N, Iliopoulos C, Kolokitha OE. Macroglossia. Int Dent J. 2011;61:63-9.
16. Proffit WR. Equilibrium theory revisited: factors influencing the position of the teeth. Angle Orthod. 1978;48:175-86.
17. Harvold EP, Chierici G, Vargervik K. Experiments on the development of dental malocclusions. Am J Orthod. 1972; 61:38-44.
18. Southern Association of Institutional Dentists. Down Syndrome: A review for the dental professional. In Self-Study Course Module 3: 1-9. www.saiddent.org. (Late access date 11/19/2016).
19. Margulies P. Genetic Diseases and Disorders Down Syndrome. 1st ed. New York: The Rosen Publishing Group, Inc. 2007:4.
20. Berkovitz BKB, Holland GR, Moxham BJ. Oral Anatomy, Histology, and Embryology. 3rd ed. London: Mosby. 2002:5-67.
21. Shukla D, Bablani D, Chowdhry A, Thapar R, Gupta P, Mishra, S. Dentofacial and cranial changes in down syndrome. Osong Public Health and Research Perspectives. 2014;5(6):339-44.
22. Garcia de Guilarte RF, Fronher BB, Urcelay PR, Najera RC, Meli BG, de Salamanca Celada JE. An idiopathic case of macroglossia. Journal of Plastic, Reconstructive & Aesthetic Surgery. 2009;62:41-3.
23. Caban-holt A, Head E, Schmitt F. Chapter 15 - Down Syndrome Fifth Edition. Rosenberg's Molecular and Genetic Basis of Neurological and Psychiatric Disease. 2015;163-70.
24. Dimitriou D, Leonard HC, Kamiloff-Smith A, Johnson MH, Thomas MSC. Atypical development of configural face recognition in children with autism, down syndrome and Williams syndrome. J Intellectual Disability Res. 2015;59:422-38.

25. Van Trotsenburg, et al. The effect of thyroxine treatment started in the neonatal period on development and growth of two-year-old Down Syndrome children: A randomized clinical trial. *J Clin Endocrinol Metabolism*. 2005;90(6):3304-11.
26. More J. *Infant, Child and Adolescent Nutrition: A Practical Handbook*. 1st ed. New York: CRC Press. 2013:134-9.
27. Marchal, et al. Growing up with Down Syndrome: Development from 6 months to 10.7 years. *Res Dev Disabilities*. 2016;59:437-50.
28. Jacobs PA, Hassold TJ. The Origin of Numerical Chromosome Abnormalities. *Advances in Genetics*. 1995;33:101-33.
29. Kovaleva NV. Gender affects clinical suspicion of down syndrome. *Russian Journal of Genetics*. 2011;13:210.
30. Huang SW, Emanuel I, Lo J, Liao SK, Hsu CC. A cytogenetic study of 77 Chinese children with Down's syndrome. *J Mental Deficiency Res*. 1967;11:147-52.
31. Tonomura A, Oishi H, Matsunaga E, Kurita T. Down's syndrome: A cytogenetic and statistical survey of 127 Japanese patients. *Japanese J Hum Gen*. 1966;11:1-16.
32. Slavin RE, Kamada N, Hamilton HB. A cytogenetic study of Down's syndrome in Hiroshima and Nagasaki. *Japanese J Hum Gen*. 1967;12:17-28.
33. Bernheim A, Chastang C, de Heaulme M, de Grouchy J. Excès de garçons dans la trisomie 21. *Ann Genet (Paris)*. 1979;22:112-4.
34. Nielsen J, Jacobsen P, Mikkelsen M, Niebuhr E, Sorensen K. Sex ratio in Down syndrome. *Ann Genet (Paris)*. 1981;24:212-5.
35. Lindsten et.al. The incidence of Down's Syndrome in Sweden During the Years 1968-1977. *Human Genetics Supplement*. 1981;2:95-210.
36. Uchida IA. Epidemiology of mongolism; the Manitoba study. *Annals of the New York Academy of Sciences*. 1970;171:361-9.
37. Chitham RG, Maclver E. A cytogenetic and statistical survey of 105 cases of mongolism. *Ann Hum Genet*. 1965;28:309-15.
38. Huether CA. Epidemiological Aspects of Down Syndrome: Sex Ratio, Incidence, and Recent Aspects of Prenatal Testing. *Issues and Reviews in Teratology*. 1990;5:263.
39. Petersen, et.al. Paternal nondisjunction in trisomy 21-excess of male patients. *Human Molecular Genetics*. 1993;2:1691-5.
40. Griffin DK, Abruzzo MA, Millie EA, Feingold E, Hassold TJ. Sex ratio in normal and disomic sperm: evidence that the extra chromosome 21 preferentially segregates with the Y chromosome. *Am J Hum Genet*. 1996;59:1108-13.
41. Ellis, et.al. *Sex Differences Summarizing More Than A Century of Scientific Research*. 1st ed. New York: Psychology Press. 2008:152.