

Description of upper intermolar dental arch size in thalassemia beta mayor aged 9-14 years old based on gender (The study is performed at the Thalassemia Pediatric Clinic of Dr. Hasan Sadikin Bandung General Hospital in 2007)

Nana Anggraini, Eriska Riyanti, Eka Chemiawan

Department of Pediatric Dentistry Faculty of Dentistry Universitas Padjadjaran

ABSTRACT

Beta major thalassemia was characterized by severe hereditary hemolytic anemia and suffered from retardation in growth, and facial skeletal alteration also narrower arches. The purpose of this research was to obtain data on intermolar width in the 9-14 years old beta major thalassemia patients by gender at the Thalassemia Clinic of Pediatric Department Dr. Hasan Sadikin General Hospital Bandung. This research used the description method with survey technique. Sample collecting was by consecutive sampling resulting in a number of 57 samples and measurement in model were allowed. Research result showed that the average of the intermolar width of beta major thalassemia patients age 9-10 (intermolar width 5,52 cm), 10-11 (5,10), 11-12 (5,28), 11-12 (5,46), 12-13 (5,52), and 13-14 (5,52). In 25 girls patient 9-10 (5,01), 10-11 (5,02), 11-12 (4,93), 12-13 (5,04), and 13-14 (5,13). The conclusion of this research was that intermolar width varied in every age range.

Key words: beta major thalassemia, intermolar width

INTRODUCTION

Thalassemia is the most frequent genetic disorder in the world which is due to damages in the synthesis of hemoglobin subunit. Around 100,000 babies in the world are born with a severe form of this disease each year.^{1,2} According to the Ministry of Health it is estimated that one in 1,600 newborns in Indonesia suffer from severe thalassemia mayor in 2002. it is estimated that in every 4 million babies born each year 2,500 one born with thalassemia major.³

Dr. Hasan Sadikin General hospital is the central referral for thalassemia disease in West Java because it has a special clinic for thalassemia patients and disferal pump that is only accessible at this Thalassemia clinic and with the availability of poor patient insurance scheme (Askeskin). In December 2006, there were 380 patients treated in the Thalassemia clinic of Dr. Hasan Sadikin General Hospital not all of them were thalassemil as some of them suffered from other blood-related disorders such as hemophilia and leukemia.

Based on the disorder in the globin chain

synthesis, thalassemia is categorized into two types: thalassemia alpha and thalassemia beta. Based on the number of globin chains subjected to the synthesis disorder, thalassemia can be categorized into Thalassemia mayor and thalassemia minor.⁴ Thalassemia minor occurs in carrier and is not dangerous. Parents with thalassemia minor may inherit the blood disorder to their children which may suffer from thalassemia mayor.

Thalassemia beta mayor is a serious blood disorder that starts since childhood. A child suffering from thalassemia beta mayor cannot produce enough hemoglobin in their blood. Therefore, they need regular blood transfusion and medical treatment. The most common thalassemia is thalassemia beta mayor which is also referred as Cooley's Anemia.⁵ Thalassemia beta mayor is also called Mediterranean Cooley's Anemia or homozygous beta thalassemia.⁶

The oral manifestation of thalassemia beta mayor occurs in the form of bimaxillary or occlusal abnormalities. Dental and facial abnormalities seen include interdental space, malar bone protrusion, and saddle nose. These several skeletal changes lead to upper lip retraction and a chipmunk facies description.⁷ According to a study performed by Alhaija et al.⁸ All thalassemia beta mayor patients have class II skeletal relationship.

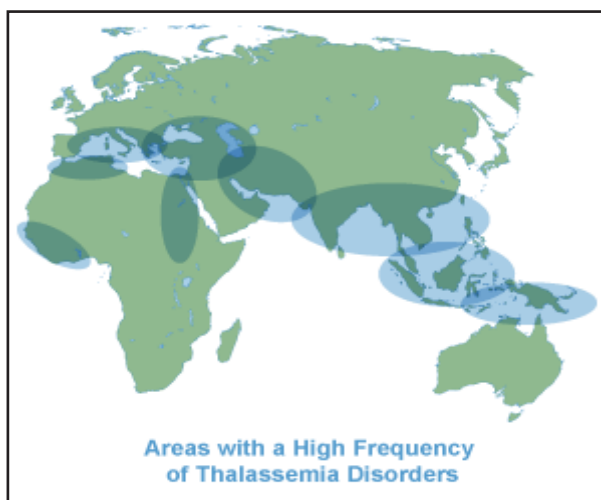


Figure 1. Area with High Frequency of Thalassemic Disorder.

Patients with thalassemia beta mayor show narrower maxilla and shorter mandible with incisive dental space smaller in maxillary and mandibular arches. The genetic variation and environmental factors such as endocrine dysfunction and somatomedin deficiency will affect the dental size of thalassemia beta mayor patient as the broad effect of delayed growth.⁹ A longer maxillary dental arch length is found in thalassemia beta mayor patients due to anterior dental protrusion that increases overjet and overbite leading to lower lip trapping between upper and lower incisors. The consequences include delayed mandible growth and more apparent maxillary protrusion with narrower maxilla from sagittal view.⁶

The different position of permanent first molar, inclination or bucolingual size of teeth may read to narrower intermolar width in thalassemia beta mayor patients compared to normal people. In addition, delayed growth and development is also found in these patients.⁶

MATERIALS AND METHOD

The population sample of thalassemia beta mayor patients in Thalassemia Pediatric Clinic of Dr. Hasan Sadikin General Hospital Bandung are patients that are willing to participate in the study after the parents are briefed and give proper informed consent. The subjects are selected using consecutive sampling method. The materials used for this research are impression materials of alginate and with impression plaster in the form of dental stone. The instruments used in this research include study sheet, gloves, mask, glass for mouth rinsing, basic instruments, perforated impression tray, rubber bowl, spatula, compass with two sharp points, and ruler.

The study is a descriptive study using survey technique. The population sample are children who suffer from thalassemia beta mayor who visited Thalassemia Pediatric Clinic of Dr. Hasan Sadikin General Hospital, Bandung. The results of the study is presented in the form of table and diagram.

RESULTS

Table 1. Patient distribution based on age and gender.

Usia	Laki2		Perempuan		Total	Persentase Total
	Total (n)	Persentase (%)	Total (n)	Persentase (%)		
9-10	5	8.77	7	12.28	12	21.05
10-11	6	10.53	6	10.53	12	21.06
11-12	10	17.53	4	7.02	14	24.55
12-13	7	12.28	4	7.02	11	19.30
13-14	4	7.02	4	7.02	8	14.04
Total	32	56.13	25	43.87	57	100

Table 2. Intermolar dental arch size. Boys thalassemia patients based on the age group.

Usia	Total (n)	Average
9-10	5	5.52
10-11	6	5.10
11-12	10	5.28
12-13	7	5.46
13-14	4	5.33

Table 3. Intermolar dental arch size. Female thalassemia patients based on the age group.

Usia	Total (n)	Average
9-10	7	5.01
10-11	6	5.02
11-12	4	4.93
12-13	4	5.04
13-14	4	5.13

DISCUSSION

Thalassemia beta mayor is a severe hemolytic anemia disease with patients experiencing a very slow growth and development and even obstructed growth and development.^{6,10} This is also seen in thalassemia beta mayor patients in Pediatric Thalassemia Clinic of Dr. Hasan Sadikin General Hospital who participated as sample in this study. The physical appearance of beta mayor thalassemia patients resembles a child (under 10 years old) when they are actually in their teenage phase indicating delayed growth and development. The intermolar dental arch size also does not show much difference between the youngest age group (9-10 years old) and the oldest age group (13-14 years old) in both gender.

The maxillary growth in thalassemia beta mayor is more towards the anterior due to bone marrow hyperplasia such that the maxilla seems bigger than normal.⁷ The dental arch of these patients are narrower and the growth is delayed.⁶ The intermolar dental arch width in boys are bigger than girls in all age group.

Around 22.80% of patients visit the dentist for dental extraction. The dental extraction is performed on persistent primary teeth or teeth

with severe caries. There is a difference between the dental arch size during the mixed dental period due to the existing Leeway space. Leeway space is a space that occurs when primary canines, first molars and second molars are replaced by permanent canines, first premolars and second premolars that have smaller size.¹¹ This will affect the intermolar dental arch size especially when a premature primary tooth extraction is performed before the eruption of the permanent tooth that will lead to mesial drifting of the posterior teeth.¹²

The intermolar upper dental arch in thalassemia beta mayor patients varies in different age group (Table 2 and 3). This probably depends on the frequency of patients who receive transfusion therapy. It is already acknowledged that when patients received periodic transfusion, the skeletal change due to anemia can be reduced.^{7,10} 43.86% of the patients receive two transfusion a month (once every 2 weeks) and mostly come from Soreang, Ciamis, and Garut. The Hb level test before the patients receive transfusion therapy show an Hb level below 10 g/dL. The main objective of the transfusion therapy is to maintain the Hb level of the patients in the range of 9-10 g/dL, to suppress erythropoiesis increase

so that extramedullary hematopoiesis and skeletal change can be reduced. thirteen Around 85.29% of patients were late/could not receive transfusion due to distance and financial barriers. Delayed transfusion may reduce its benefit and will not prevent skeletal changes.¹⁴

In normal people, the period of is characterized by growth spurt with the peach? peak height velocity (PHV) at the age of 12.5 years in girls and 14 years in males with 4 year range, the growth rate is decreased after the PHV. The PHV of girls comes before the boys with a lower value and shorter period. The period is characterized by secondary sexual characteristics, i.e. menarche (menstruation function development process) one to two years after PHV and changes in voice or similar to adult voice in boys.¹²

Based on the results of the questionnaires and physical examination, there seems to be no growth spurt. The physical appearance resembles a ten year old child and is almost difficult to determine their age except to ask the patient or the patients family directly. Based on the questionnaire result, there are no secondary sexual characteristics in all patients both boys or girls by the age of 14 years old. Sixty four point nine one percent of these patients still face eating difficulty, especially if they have not received the transfusion therapy. This may lead to malnutrition which is one of the factors that delay growth and affect the body size and proportion, body chemicals and the quality and texture of tissues such as teeth and bone.¹²

Patients with siblings who have similar disease contribute 33.33% of the population showing that thalassemia beta mayor is a hereditary disease. About 10.53% thalassemia beta mayor patients have nail biting habit and burxism and 5.26% have thumb and lip sucking habit. 68.42% patients show other bad habit. This indicate that the maxillary protrusion seen is not caused by bad habit but due to bone marrow expansion caused by anemia.⁵

Most patients brush their teeth twice to three times a day, i.e. in the morning and afternoon. Toothbrushing before going to sleep at night is rarely done. Several patients only brush their teeth once a day because of pain during toothbrushing. Dental visits only done once or twice and mostly done during childhood. Most of

thalassemnia patients have severe caries with only part of the crown or root left. This may be caused by the inability to close the mouth that makes the patients prone to caries.⁶

Based on the results from the questionnaire, it is revealed that 56.14% of children with thalassemia beta mayor are diagnosed by the age of less than a year and all patients directly receive transfusion after being diagnosed. Children were usually brought to the doctor after blood examination is done if the child does not recover after receiving medication. This shows that parents awareness towards their childrens development is quite high and children with thalassemia can receive early treatment.

CONCLUSION

Based on the description study of the intermolar dental arch size in thalassemia patients aged 9-14 years old based on gender in the Pediatric Thalassemia Clinic of Dr. Hasan Sadikin General Hospital Bandung it is concluded that the intermolar dental arch varies in each age group. The male intermolar dental arch compared to the females which are narrower than normal.

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