Lateral facial profile of β-thalassemia major in Javanese children: a photogrammetric analysis

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ABSTRACT

BACKGROUND Impairment of globin chain synthesis in patients with β-thalassemia major causes ineffective erythropoiesis. This condition triggers bone marrow hyperplasia and can lead to craniofacial bone abnormalities. This study aimed to evaluate the lateral facial profile of β-thalassemia major, identify any differences when compared with a control group of similar age and ethnicity, and reveal the facial profile of β-thalassemia major in Javanese children.

METHODS This cross-sectional study included 35 β-thalassemia major children aged 7–15 years. They were divided into 3 groups based on their age. Lateral facial photogrammetry was taken by measuring the forehead protrusion and nasolabial angle. Data were then mapped to the normal group of children in the same age group and descriptively analyzed using SPSS software.

RESULTS The 1 and 2 SD group had a higher prevalence of β-thalassemia major in children aged 7–9 years for both sexes. The older age group had a closer mean value to those of children without thalassemia.

CONCLUSIONS The forehead and maxillary profile of Javanese β-thalassemia major children tended to be protrusive, especially in the 7–9 years age group, while the older age groups had closer mean values to those of children without thalassemia.

KEYWORDS beta-thalassemia, facial bones, photogrammetry

Thalassemia is a common genetic blood disorder in Indonesia. Thalassemia cases have increased from 4,896 in 2012 to 8,761 in 2018.¹-² β-thalassemia major, the most severe thalassemia, causes severe anemia due to impaired hemoglobin synthesis,³ stimulating the bone marrow to increase erythrocyte production due to ineffective red blood cell formation. Subsequently, bone expansion may result in cortical thinning, bone distortion, and fragility, often leading to characteristic skeletal and oral and maxillofacial deformities.¹

The most visible facial characteristics of patients with β-thalassemia major are maxillary protrusion and a prominent forehead or frontal bossing,⁴ which can be identified using facial photography. Photogrammetry is an effective method to evaluate external craniofacial structures, including soft tissue involvement,⁵ and measure the angular facial profile.⁶

Studies regarding the facial profile characteristics of patients with β-thalassemia major, especially in Indonesia, remain unclear. This study aimed to identify the lateral facial profile of Javanese children with β-thalassemia major and elucidate the forehead and maxilla as the clinical predictors in pediatric dentistry for diagnosing β-thalassemia major.
METHODS

This study was approved by the Ethics Committee of the Faculty of Medicine, Public Health, and Nursing, Universitas Gadjah Mada, Dr. Sardjito General Hospital, Yogyakarta (approval number: KE/FK/0697/EC/2021). Informed consent was obtained from the patients’ parents.

This quantitative study included 35 Javanese children (18 males and 17 females, aged 7–15 years) at the Division of Hematology and Oncology, Department of Pediatrics, Dr. Sardjito General Hospital, Yogyakarta, who underwent blood transfusions every 2–3 weeks and were treated during September 2021. The patient’s demographic data, medical history, body weight, and height were recorded. Photographs of the lateral aspect of the face were obtained at a distance of 150 cm from the tip of the patient’s nose for standardization using a mirrorless camera (Sony α7 mark II, Sony, Japan). The patients were seated upright on a chair without a backrest in a resting position, with their eyes looking straight ahead. All of the photographs were obtained by one photographer in the same setting using microlens placed on a tripod at the eye level. The facial midline and interpupillary line were aligned with vertical and horizontal lines on the camera to correct for facial deviations. A cephalostat modification was used to maintain the position of the head so that the Frankfurt horizontal plane was parallel to the floor, and the camera direction was perpendicular to the patient’s face. Other photographic parameters were also standardized (aperture: F/4.5; shutter speed: 1/125; and ISO: 2000). Adequate lighting was used for all photographs.

The forehead protrusion (trichion [highest point of the forehead]–nasion) and nasolabial angle (NLA) were measured on the photographs using CorelDRAW 2021 (Corel Corporation, Canada) (Figure 1). The forehead protrusion angle was measured from the intersection of the trichion–glabella line and the line drawn from the glabella along the edge of the forehead to the trichion. The NLA was formed by the intersection of pronasal with the subnasal line and subnasal with the upper lips line.

The patients were divided into three dental age groups: 7–9, 10–12, and 13–15 years. The patient characteristics were compared with children without thalassemia in the same age group who were of Javanese ethnicity, lived in Yogyakarta or its surrounding areas, had a normal body mass index, had no history of orthodontic treatment, and had an externally normal jaw. The mean and standard deviation (SD) of the control group’s measurements were calculated by SPSS software version 26 (IBM Corp., USA) and classified into four categories: mean, -1 SD, -2 SD, and -3 SD value groups. The individual score of thalassemia patient was mapped into control group’s graphs.

The photographs were processed using Adobe Photoshop (Adobe Systems Incorporated, USA). Facial profile photos were evaluated using CorelDRAW 2021 (Corel Corporation) at a 1:1 magnification. All photographs were re-evaluated by the examiner after 1 week. The data were analyzed using SPSS software version 26 (IBM Corp). Statistical significance was set at $p<0.05$.

RESULTS

Of 35 children with thalassemia major, 17 were in the 7–9 age group, 13 in the 10–12 age group, and 8 in the 13–15 age group. The NLA and forehead protrusion angle were distributed across the three age groups, and the mean value for each group can be seen in Figure 2. The highest mean value of the forehead protrusion angle for males and females was observed in the 12–15 age group.

The mean value for each control group and the mapping results showing the distribution of each
β-thalassemia major individual in the control group are shown in Figure 3. The 1 and 2 SD group had a higher prevalence of β-thalassemia major in children aged 7–9 years for both sexes. The older age group (10–12 and 13–15 years) had closer mean values to those of children without β-thalassemia major.

**DISCUSSION**

Based on the mapping chart of the forehead protrusion and NLAs, patients aged 7–9 years with β-thalassemia had protrusive features compared with those without thalassemia. Forehead protrusion or frontal bossing is a common characteristic of patients with β-thalassemia major. Ineffective erythropoiesis leads to bone marrow hyperplasia, resulting in wall thinning, bone expansion, and protruding frontal bones in the forehead and maxilla. These protrusions typically appear between 6 and 24 months of age. Therefore, differences in bone structure can be observed in children aged 7–9 years. Abnormal bone structure affects the development of the paranasal sinuses and mastoids and often affects the maxillary, sphenoid, and frontal sinuses, which are distinctive radiographic findings in patients with β-thalassemia major that are not observed in healthy individuals.
Furthermore, a previous cephalometric photographic analysis study demonstrated a more prominent forehead in children with β-thalassemia major.

In this study, patients with β-thalassemia major aged 10–12 or 13–15 years had less prominent facial features; their measurements were closer to those of children without thalassemia. This may be due to the growth of the neurocranium being nearly complete, allowing the facial complex to develop in a more typical pattern by this age, similar to the growth pattern of children without thalassemia. No previous photogrammetry studies regarding the forehead protrusion angle in children without thalassemia have been reported. While some studies have reported the NLA, the patient population in those studies does not align with that of the current study. In addition, some previous studies used the pronasal point of the nose, as in this study, while others used the columella when determining the NLA. Varying NLAs were reported for 12-year-old children in southern China, though the NLAs were based on the columella, and the SD was wide (maximum NLA: 120–130°).

Several factors, such as blood transfusions, may influence the condition of patients with thalassemia. Blood transfusion is the main treatment for severe anemia in patients with β-thalassemia major as it suppresses ineffective erythropoiesis. The variations observed in the current study may have been caused by the duration, frequency, and regimen of blood transfusions in each patient. Notably, 55% of patients with thalassemia who received adequate transfusions did not develop facial bone deformities, indicating that adequate treatment can prevent such deformities.

The variations in the findings of the current study may also be attributed to the unequal distribution of age and sex in each group. Other factors such as poor oral habits and soft tissue thickness, which were not observed in this study, may also have contributed to these variations.

The forehead and maxilla of patients with β-thalassemia major are valuable indicators for recognizing the patient’s condition and determining the appropriate treatment options in pediatric dentistry. These clinical predictors are important for diagnosing β-thalassemia major. In addition, extraoral photographs are suitable for assessing patients with thalassemia as they eliminate the need for invasive and lengthy procedures.

In conclusion, the forehead and maxillary profile of Javanese children with β-thalassemia major was protrusive, especially in patients aged 7–9 years. More research with a larger patient population in each age group is needed to complete a statistical analysis of the facial features in this patient population.

REFERENCES