REVIEW PAPER

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Separate growth charts and cephalometric norms for children with Down syndrome

Joanna Kurpik[1, a], Artur Matthews-Brzozowski[2, b]

[1] Chair and Department of Maxillofacial Orthopaedics and Orthodontics, Poznan University of Medical Sciences, Poland
[2] Department of Oral and Maxillofacial Surgery, Medical Centre Leeuwarden, Leeuwarden, The Netherlands
[a] https://orcid.org/0000-0002-9851-7248
[b] *not available*

ABSTRACT

Different phenotypic features characterizing the body structure of children with Down's syndrome, which include low growth, small head, short limbs, as well as the tendency to obesity and other systemic diseases or congenital malformations, prompted the WHO to develop separate standards including growth charts for children with this syndrome. Selected authors in their studies also compare orthodontic parameters, and more precisely cephalometric parameters, between children with Down's syndrome and healthy individuals. They note a tendency to repeated deviations from the accepted norms, including the skeletal class, antero-posterior dimensions of the jaw, the length of the base of the skull, the cranial base angle, and ANB, SNA, SNB angle. It is related to the occurrence of specific features of the skull skeleton structure, typical for children with Down's syndrome. The described tendency of changes in cephalometric parameters, in correlation with the already developed separate growth charts to assess the growth of children with Down's syndrome, leads to considerations on the need to develop separate standards in the field of orthodontics, adequately defining the skeletal structure of the facial part of the skull of these children.

Keywords: growth charts, Down's syndrome, cephalometric analysis.

Introduction

Properly constructed and appropriate growth charts are necessary to assess the correctness of body growth, and also indicate optimal physical development of the child, health and nutrition [1, 2]. Monitoring and assessment of child development is one of the most important tasks of medical care. These growth charts, presenting graphically developed development norms, are constructed in such a way that successive percentile lines determine the percentage of children in each age group below their level, i.e. if the measurement value of the tested feature is on the 10th percentile, it means that in this calendar age 10% of peers are characterized by a lower value of this feature [3]. Limits of the so-called narrow standard are defined by 25th and 75th centile. The growth chart, also known as percentile or centile chart, gives the opportunity to compare the selected parameter, e.g. weight or height of the child in relation to other children of the same age and sex [4]. The basic method of assessing the physical development of a child is to compare his individual phenotypic image with the developmental norm (reference system), however it should be emphasized that the developmental norm may depend on comorbid diseases, which include genetic syndromes [3].

A specific group is represented by children with various mental disabilities co-occurring with specific genetic syndromes, e.g. children with Down's syndrome. The prevalence of this syndrome is estimated to be 11–16 per 10,000 [5, 6]. There are many characteristics of children with Down's syndrome, which include, among others, low growth, small head, single transverse palmar crease, almond shaped eyes caused by a fold over the eyelid, weakened muscle tone. An increased risk of congenital heart disease, gastroesophageal reflux, recurrent middle ear infections, hyperthyroidism syndrome and thyroid gland diseases are also reported [1, 2, 7, 8]. High risk of occurrence of numerous impairments in the functioning of individual body systems does not remain indifferent to the process of proper growth and development. Separate growth charts for children with Down syndrome

Children with Down's syndrome (Ds) are born with a smaller birth weight, but they develop overweight when they are 3 to 4 years old. The tendency to overweight is quite common — at the age of 19, it occurs in 31% of men with Ds and in 36% of the female with Ds [9]. The tendency towards the specific features of body structure in children with this syndrome, prompted the World Health Organization to construct dedicated growth charts for children with Down's syndrome. Van Gameren-Oosterom et al. describe that the first charts for Dutch children with Down syndrome were published and introduced in 1996 [2].

The growth scheme of children with Down syndrome is distinguished by a significant impairment of their developmental pace, starting from the moment of birth to adolescence, intensified in particular in the age range from 6 months to 3 years and also during puberty. According to the WHO guidelines for the assessment of excessive body mass, the BMI mass index is most commonly used to classify the nutritional status of children, adults and the elderly [9]. It is emphasized that obesity is a common condition among children with Down's syndrome [10]. The tendency to present typical features of body structure is one of the important arguments for the rightness of constructing separate growth chart for children with Down's syndrome.

Zemel et al. describe that the characteristic features of the phenotypic image of children with Down's syndrome in relation to healthy children include, among others, shorter limbs, which undoubtedly affects a different distribution of body mass in relation to weight [10]. As shown by Bertapelli et al., the uninterrupted scheme of BMI growth observed in children with Down's

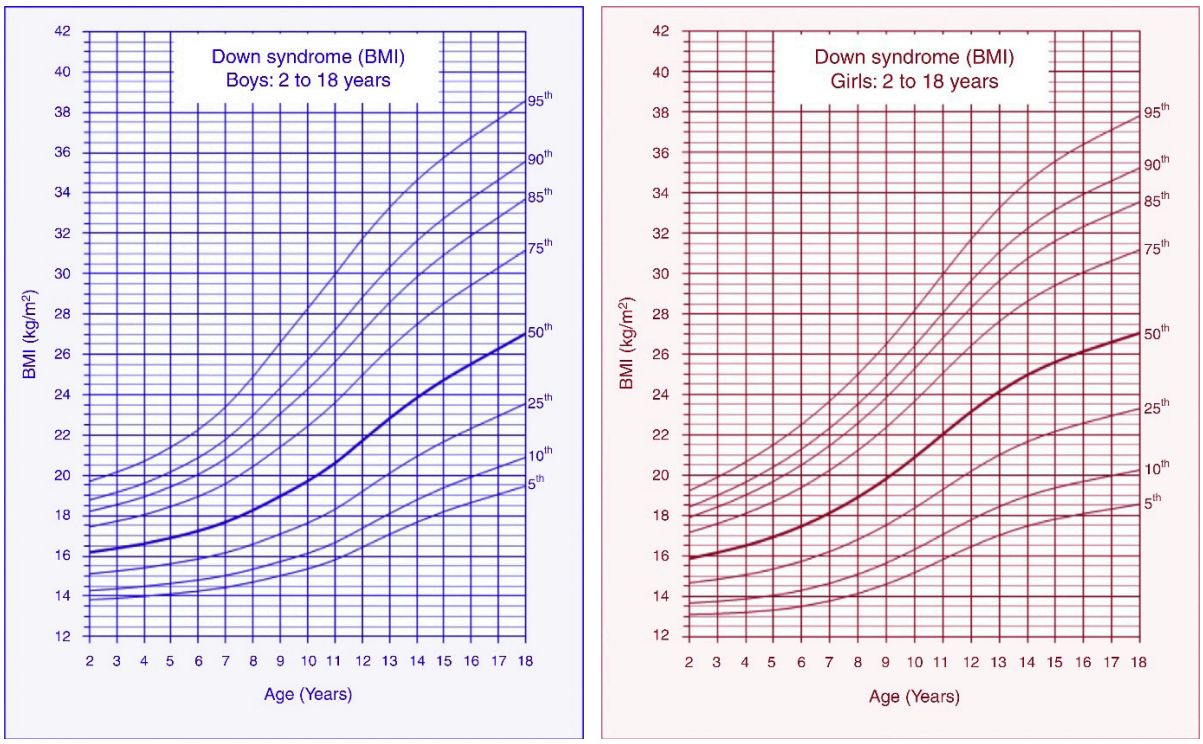

Figure 1. Growth charts expressing BMI values for boys and girls with Down's syndrome aged 2–18 years [11]

Figure 2. CDC Growth Charts – growth charts expressing BMI values for healthy boys and girls, without Down's syndrome, aged 2 to 20 years [12]

syndrome is definitely different compared to BMI standards defined for the general population without this syndrome (**Figures 1, 2**). International growth references indicate a rapid acceleration of the central percentile of BMI in the first year of life, followed by a decline by the age of 5 years, and a subsequent acceleration in later life. These age-specific BMI curves are descriptive growth standards in children and adolescents with Down syndrome. However, these curves do not indicate the optimal standard of weight to which all children with this syndrome should grow. The use of these curves, however, allows us to compare and monitor the status of body mass [11].

Cephalometric parameters in children with Down's syndrome

The literature describes a number of characteristic features of skull structure in children with Down's syndrome, including hypoplasia of the middle part of the face, flattening of the skull base, skeletal class III together with the co-occuring open frontal bite.

Quintanilla et al. [13] assessed the morphology of the facial part of the skull of patients with Down's syndrome based on the results of cephalometric analysis, the study group included children from 7 to 18 years of age. The average size of the overbite and overjet parameters was -1.01 and 1.73 respectively, with respect to the Ricketts standards (overbite = overjet = 2.5 mm) adopted by the authors, which are therefore lower values, and the negative overbite confirms the reverse overjet. The inter-incisal angle, whose mean value in the patients tested was 126.4°, was underestimated with respect to the Ricketts standard of 130°. The average length of the anterior segment of the skull base of children with Down's syndrome was 52.12 mm, which was slightly lower than the accepted standard of 55 mm. The authors did not include the control group corresponding to the age of the respondents, hence all the values referred only to the adopted standards developed by Ricketts. In addition, the analysis does not include the parameters describing the base angle of the skull, the anterolateral relation of the mandible to the jaw, as well as the relationship of the mandible and jaw to the base of the skull. They describe

that, unlike other authors, they did not obtain classification results for the III skeletal class, which was explained by the fact that this group included people in the period of the growth of the skull.

Similar research was undertaken by Suri et al. [14], who compared the results of the analysis of 25 cephalometric X-rays of children with Down's syndrome in the age range from 11 to 18 years. The results obtained were referred to the control group of healthy children of similar age, with I skeletal class. The results showed a reduction in the linear dimension of the anterior length of the skull base and a slight increase in the skull base angle value in children with Down's syndrome, in relation to children without this syndrome. It should be emphasized that the norm of skull base angle according to Segner and Hasund is within 128–136°, while the value of this parameter for children with Down's syndrome was 140.31°, being an inflated value, for children from the control group it was within the normal range. All dimensions concerning the jaw were interpreted by the authors as significantly smaller in the group of children with Down's syndrome, its length was reduced by 17.4% in relation to the control group, amounting to 47.8 mm. The SNA angle for children with Down syndrome was on average 82.47° and showed no significant difference in values relative to the control group. The limit of the standards according to Segner and Hasund is 79–85°. The average value of the SNA angle for both children with Down syndrome and without this syndrome is within this limit. The dimensions of the SNB angle were higher in the test group relative to the control group, but the results of both groups were within the normal range of 77–83° according to Segner and Hasund. Co-occurring anterior mandibular rotation has been recognized by the authors as a factor favoring the occurrence of its prognathism. In the group with Down's syndrome, 48% patients had anterior cross-bite.

In another paper, Melo de Matos et al., [15] analyzed cephalometric X-rays of 15 patients with Down's syndrome in the age range from 21 to 34 years, and the results were referred to a control group of 15 healthy people, appropriately assigned by age, of the Brazilian population. On the basis of own observations they assessed that in Down's syndrome the values of the length of the anterior and posterior base of the skull are reduced, while the value of the base angle of the skull is increased. For people with Down's syndrome the mean value of this angle was 151.5°, in relation to the standards of Segner and Hasund developed for Europeans amounting to 128–136° for the NSBa angle, which is definitely above the upper limit of the norm, also for the control group it was 140.3°, being in the Brazilian population higher than in the norms adopted for Europeans. They also obtained lower values of SNA and SNB angles in people with Down's syndrome compared to the control group, which they estimated as a distal position of the maxilla and mandible relative to the base of the skull. With regard to standards developed by Segner and Hasund, the SNA angle is 79–85°, and the SNB angle is 77–83°, the mean values of these angles for people with Down's syndrome are lower, while for people without this syndrome they fall within the reference values. The authors' analysis of the relation of the mandible to the maxilla based on the ANB angle showed a significant reduction of this angle in relation to the group of healthy people, which was interpreted as a tendency of III skeletal class occurrence. The inter-incisal angle in the group of subjects was lower in relation to the control group, which was caused by protrusion and proclination of the upper central incisors. The norm of values of the inter-incisal angle according to Segner and Hasund is 125–141°, while the average value of this parameter for people with Down's syndrome is below the lower limit of the norm and amounts to 119.3°. For the control group, it is 125.5°, i.e. within the reference values. The authors qualified people with Down's syndrome in their adulthood, hence it is impossible to refer and compare the results to previously reported studies, including children in developmental age. The obtained results were compared only between the test and control groups, omitting a reference to valid cephalometric standards, e.g. in the analysis of Segner and Hasund.

Development of norms of cephalometric parameters for children with Down syndrome

A clear tendency to deviations of certain cephalometric parameters, resulting from a different skeletal structure of the facial part of the skull, which was described in the literature, may suggest the need to develop separate norms of cephalometric parameters for children with Down's syndrome, as well as growth charts. The values that show a tendency to deviate seem to include the length of the anterior cranial base, cranial base angle, antero-pos-

terior dimensions of the jaw, ANB angle, SNA angle and SNB angle, as well as the inter-incisal angle. It is worth noting that the cited studies of different authors demonstrate no unification of the age group of the persons with Down's syndrome, as well as no systematic reference of the obtained results, as some authors refer them to adopted and generally known norms, e.g. developed by Ricketts, and others compare them only between the test and control group, omitting the adopted standards. It would be noteworthy to conduct a study on children with Down's syndrome in the appropriate age range, including the growth period, and allowing proper cooperation with the child to obtain reliable lateral cephalometric X-rays. A similar issue concerns the physical development of the body of children with Down's syndrome, which, showing typical phenotypic traits, should not be compared with the values developed for healthy children, hence separate percentiles were created for them, taking into account the typical body structure and the tendency for a different growth scheme. Based on the above, the question arises whether, due to the tendency for a different skeletal structure of the facial part of the skull described by many authors, it is not worth considering and directing attention to the desirability of developing separate values of cephalometric parameters that would be considered a norm for children with Down's syndrome. This is an open question that requires proper research and, above all, the gathering of a sufficiently large group of subjects, a control group and a comparison of values between them and references to generally accepted norms.

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Correspondence address:
Joanna Kurpik
Chair and Department of Maxillofacial Orthopaedics and Orthodontics, Poznan University of Medical Sciences, Poland
email: joanna@kurpik.pl