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Morphometric Assessment Of Mandibles With Complications Resulting From Temporomandibular Joint Ankylosis In Children From 4 Months To 3 Years Of Age

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Abstract

Purpose: The main aim of this paper was to perform the morphological assessment of children's mandibles of different etiology of dysfunctions within the temporomandibular joint, from isolated idiopathic ankylosis to craniofacial malformations co-existing with genetic disorders.

Patients and Methods: The investigations encompassed seven patients at the age 0-3. Measurements were conducted on the basis of data obtained from computed tomography. In the patients' 3D models 12 characteristic anthropometric points were marked, on the basis of which the researchers calculated 9 linear dimensions, 3 angles and, further on, 5 indices defining the proportions of the mandible.

Results: The comparisons of the measurements showed that the mandibles with malformations were smaller than obtained for healthy children. The values of the mandible angles (alpha) revealed high changeability among the examined children. It was also revealed the significant asymmetry between the right and left side.

Conclusion: This work shows an innovative approach to the evaluation of anatomical malformations, whereas the developed indices enable an objective quantitative assessment of geometry, which can be applied to the analysis of the degree of the deformation severity. An in-depth analysis of the image diagnostics should be a standard procedure in the pre-operative planning of surgical treatment and the preparation of multi-specialist logopaedic, functional and orthodontic rehabilitation.

Keywords: Statistical analysis, mandible geometry, infants and toddlers, 3-dimensional measurements, mandible diseases

Introduction

Ankylosis of temporomandibular joint (TMJ) is a disease which leads to disorders of the joint functions [6]. There are two types of ankylosis, true and false. The true ankylosis occurs when the mandible movement disorders are caused by a fibrous or bony fusion between the surfaces of the joint structures of the condylar process and the joint pit [8]. In the case of false ankylosis, the mandible movement disorder is caused by extracapsular pathology and the imaging tests do not reveal any changes in the joint structures. An example of the extracapsular ankylosis is a rare fibrous or bony fusion between the coronoid process of the mandible and the zygomatic arch [[5]]. Ankylosis may take over the whole joint or a part of the joint; it may be unilateral (approx. 93% of cases) or bilateral (approx. 7% of cases) [28] The disorder is diagnosed and confirmed by means of imaging tests [2].

Depending on its type, the disease may lead to fibrosis, deformations or bony fusions within the masticatory system. It may also lead to micrognathia. It may stop the growth of both jaws [11][16][17] deformation of the dental arch and ankylostoma (lockjaw). Medical complications also include the damage to the hearing organ [20]. Structural changes accompanying TMJ ankylosis clearly disturb the functioning of the masticatory system in patients. Patients have difficulty crushing, chewing and swallowing food as well as breathing, which often results in choking [22][25][27][29].

TMJ ankylosis may occur both in children and adults. According to available data, it most often takes place in children aged 10-15. However, the disorder is very often diagnosed late in spite of clear symptoms.

The precise etiology of TMJ ankylosis is not well known and has not been thoroughly investigated. Pathogenesis, which probably explains the origins of the disorder, includes a post-traumatic haematoma which causes pathological formation of the bone in an incorrect re-building process. The most frequent factors triggering TMJ ankylosis are therefore traumas or inflammatory states [4]. The occurrence of TMJ ankylosis due to trauma fluctuates from 26% to 75% [1]. The trauma may result, among other things, from the forceps delivery which was performed in an incorrect way. Other factors include inflammatory conditions of the TMJ, developmental anomalies of the foetus and improper treatment of the middle ear inflammation [3][12][21][28].

The presence of developmental anomalies formed during intrauterine life constitutes a characteristic feature of selected genetic disorders. The subject literature does not provide much information on the genetic origins of congenital TMJ ankyloses (ORPHA:210576); they are associated with craniofacial malformation developed from the first branchial arch. Ankyloses

may occur as an isolated anomaly or be accompanied by the malformation of other organs in different genetic disorders, including PRS - Pierre Robin Sequence, which may occur as an isolated form (ORPHA:718) or in combination with other syndromes [9]. Patients with PRS show micrognathia, cleft palate and glossoptosis. The literature describes candidate genes that might be associated with PRS. Ankylosis may also occur in SGS - Schinzel-Giedion Syndrome (ORPHA:798). Recent findings describe variants in gene SETBP1 in patients with SGS. The Schinzel-Giedion Syndrome is characterized by multiple developmental anomalies (defects of the urinary and reproductive system, congenital heart disease), considerable mental disability and facial deformations [15]. In CdLS - Cornelia de Lange Syndrome (ORPHA:199) there are numerous developmental anomalies (congenital heart disease and kidneys defects) and dysmorphic facial features. The syndrome is caused by pathogenic variants in the following genes: *NIPBL, SMC1A, SMC3, RAD21* and *HDAC8 [23]*.

Ankyloses may also be observed in Nager syndrome (ORPHA 245; mutations in gene *SF3B4*) and in Axenfeld-Rieger syndrome (ORPHA 782; mutations in genes *FOXC1* (6p25.3) and *PITX2* (4q25). In Axenfeld-Rieger syndrome, apart from congenital TMJ ankylosis, there also occur vision defects and craniofacial malformations, whereas in Nager syndrome there are craniofacial malformations and limbs deformations.

Development of the mandible and changes caused by age

At the moment of birth, the mandible consists of two symmetrical parts joined by means of cartilage, which mineralizes during the first year of life. After birth, the shape of the mandible is subject to constant modifications [17]. Combined with the growth of the whole organism due to an increase in age, an erect body position, the growth of primary dentition, changes in occlusion or methods of nutrition (from liquid nutrition to solid food) – individual parts of the neurocranium and the mandible grow and the mutual relations of their anatomical elements change. Due to the development of permanent teeth buds, the mandible body is subjected to elongation. It also translates into an increase in vertical and transverse dimensions of the mandible. The gonial angle becomes less obtuse and during the first 4 years of the child's life decreases from 150-160 degrees to approx. 140º [17]. The condylar process, which is initially rather small, undergoes re-structuring being in close contact with the pterygoid pit of the temporal bone (temporomandibular joint). The direction of the processes growth is closely connected with the direction of transposition of the mandible and its vertical deviations [12].

TMJ ankylosis, irrespective of its form (idiopathic, connected with genetic syndromes, caused by trauma), is a difficult issue requiring a complex and multi-phase, surgical and therapeutic approach [7][14][24]. What is essential is the knowledge of the growing process and direction of changes in the pathologically formed mandible in small children [13][16]. To undertake proper treatment it is necessary to adopt radiological diagnostics which enables correct and effective treatment. One of the most basic diagnostic methods in the treatment of ankylosis is computed tomography. The presentation of a three-dimensional image of bones makes it possible to develop realistic and spatially accurate models, which may support the diagnosis and pre-operative planning of particular stages of surgical treatment as well as further rehabilitation in the scope of orthodontics, logopaedics, physiotherapy and dietetics. Taking into consideration the above-mentioned issues, this work's purpose was a morphometric assessment of the mandibles in children, aged 0-3, diagnosed with ankyloses of the temporomandibular joint (TMJ) of different types, characterized by a similar degree of developmental malformations of the head and neck structures. The analysis made use of the computed tomography, which is one of the best diagnostic methods in the evaluation of the mandible morphometry [11][18]. Additionally, the comparison of the obtained measurements with the reference data of healthy mandibles was done.

Materials and methods

The investigations encompassed seven patients at the age 0-3, who were admitted for treatment at the Head and Neck Surgery Clinic for Children and Young Adults, Department of Clinical Pediatrics, University of Warmia and Mazury in the city of Olsztyn. The research are performed with approval of the University Bioethics Committee for Scientific Research at the University of Physical Education in Katowice (Resolution No. 1/2021). All the patients were diagnosed with ankylosis of the temporomandibular joint: one case was idiopathic ankylosis (patient 4), one case was genetically undiagnosed (NGD, patient 2), other cases resulted from genetic disorders (one case of Pierre-Robin Sequence (PRS, patient 6), one case of Schinzel-Giedion Syndrome (SGS, patient 3), one case of Cornelia de Lange Syndrome (CdLS, patient 5), two cases of Hanhart syndrome (characterized by hypoglossia and hypodactylia, patients 1 and 7). **Table 1** presents the comparison of data concerning age, sex and the description of irregularities within the head and neck areas as well as other relevant body parts in the examined children.

173 *Table 1. Clinical description of the tested patients*

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P	TYPE OF ANKYLOSIS	PART OF BODY	DESCRIPTION OF ABNORMALITIES
P1 Male HPG-HPDs 4 months	Bilateral ankylosis including condylar process	Head, neck	Small, stiffly positioned mandible; lack of mouth closure; further opening of oral cavity difficult; visible high palate; during the examination of the oral cavity by means of palpation the finger is slightly pushed back; no reaction to the insertion of a tube (intubation); no optically induced eye movements and no eye fixation; turning the head towards a strong source of light; eyeballs widely positioned, outer canthi pointing down; presence of tracheostomy due to life-threatening breathing inefficiency.
		Other systems	Lack of the left foot; preserved tibial bone and peroneal bone; underdevelopment of phalanges and metacarpus in both hands; malformation of the right foot; weak reaction to the surroundings and nursing care; lack of crying and producing sounds; rare motor activization, increased muscle tension; decreased axial tension; pneumonia.
P2 Female NGD 5 months	Bilateral ankylosis including coronoid process	Head, neck	Hypoplasia of the lower part of the face; lack of abduction of jaws; bilateral lack of TMJ mobility; presence of tracheostomy due to breathing inefficiency.
		Other systems	Presence of PEG; increased left atrium.
P3 Female SGS 9 months	Bilateral ankylosis including condylar process	Head, neck	Hypoplasia of the maxilla and mandible, high-arched palate; primary dentition; vomitory reflex; flat, high forehead; increased vertical measurements of the head; shortening of the anterior-posterior measurements of the head; prominent frontal tubers; short neck; micrognathia and mandibular retroposition; blue sclera; divergent squint; slanting and pointing-down palpebral fissures; hypertelorism; low-set ear conchas; condition after tracheostomy.
		Other systems	Shoemaker's chest; presence of PEG; decreased muscular tension; limited contact, lack of co-operation; arachnodactyly, abnormally long fingers and toes, improper location of thumbs; condition after the correction of talipes equinovarus; congenital pneumonia.
P4 Female IDP 16 months	Bilateral ankylosis including coronoid process	Head, neck	Limitated abduction of jaws (<1cm); suspicion of congenital ankylosis of temporomandibular joints.
P5 Male CdLS 18 months	Bilateral ankylosis including condylar process	Head, neck	Limited abduction of jaws (<1cm), microcephaly, micrognathia, flat nasal base, choanal atresia, ankylostoma (lockjaw), hypertelorism
		Other systems	Clinodactyly of fingers; synodactyly of 2nd and 3rd toe; cryptorchism; widely set nipples; contractures in limb joints; ventricular septal defect; hypertrophy of ventricular muscles; irregular image of brain structures: post-haemorrhagic cysts with irregular flow in cerebral vessels; dysplastic right-hand kidney with the widened pyelocalyceal system.
P6 Male PRS 32 months	Bilateral ankylosis including condylar process	Head, neck	Big head; dental caries, mandible hypoplasia; condition after the installation of an orthopaedic appliance on the palate; cochlear implant on the right-hand side; presence of tracheostomy due to life-threatening breathing inefficiency.
		Other systems	Bilateral cleft feet; rudimentary right-hand thumb; polysyndactyly of the left hand; decrease in axial tension; lack of speech development.
P7 Male HPG-HPDs 32 months	Bilateral ankylosis including condylar process	Head, neck	Limitated abduction of jaws (1 cm); hypodontia; mandible hypoplasia, ankylostoma (lockjaw); presence of tracheostomy.
		Other systems	Lack of the right-hand forearm; underdevelopment of left hand; lack of fingers 2-5, rudimentary thumb; underdevelopment of both feet.

175 NGD – no genetic diagnosis, HPG-HPDs - Hypoglossia-Hypodactyly Syndrome

Following the admission to hospital and prior to the therapy, each patient underwent a CT (computed tomography) examination of their facial cranium. All diagnostic CT examinations were conducted in the same laboratory of the Clinic, using the same equipment, namely CT Aquilion Prime SP (Canon, Otawara, Tochigi, Japan), and were performed by the same technician. This procedure enabled the obtainment of 2D images of individual cross sections of the body (resolution 512x512 px; pixel spacing: 0.3886/0.3886; slice thickness: 0.625 mm). The data obtained in the examination were saved in the DICOM format (Digital Imaging and Communications in Medicine). On the basis of the Hounsfield scale it was possible to select particular tissues/structures (e.g. bones) and generate 3D models (in the case of bones, the range of the HU scale equalled from 226 to 3071; Figure 1). The segmentation of the mandible made it possible to conduct anthropometric measurements of the bones. To process data obtained from the CT examinations the MIS24 software programme (Materialise, Leuven, Belgium) was used. The applied approach enabled the determination of values of linear and angular parameters of 3D models as well as parameters describing the shape and proportions of the mandible.

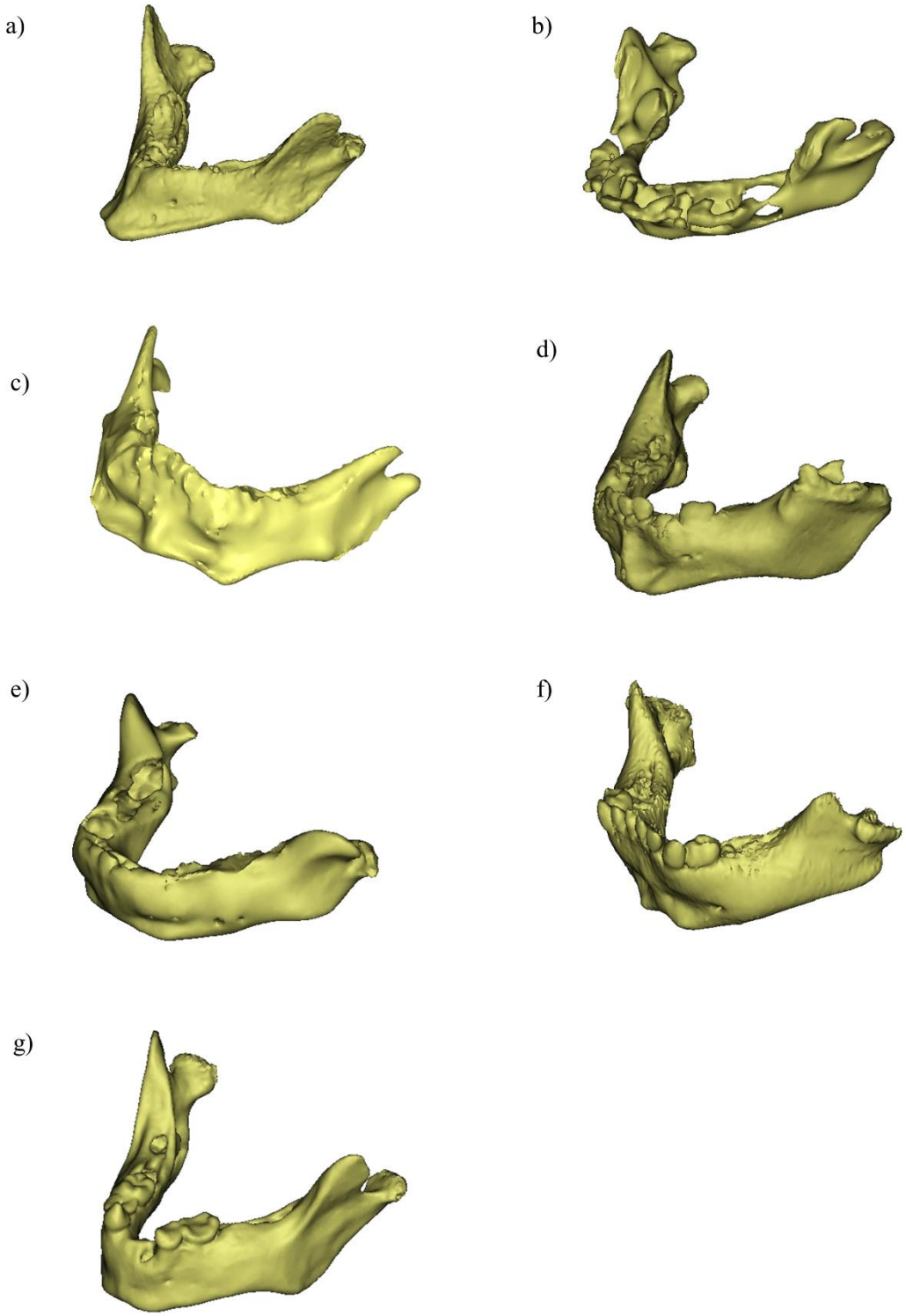

Figure 1. View of the mandible: a) patient #1, male, aged 4 mo. (HPG-HPDs), b) patient #2 female, aged 5 mo. (no genetic diagnosis) c) patient #3, female, aged 9 mo., (SGS - Schinzel-Giedion Syndrome) d) patient #4, female, aged 16 mo., (idiopathic ankylosis) e) patient #5, male, aged 18 mo., (CdLS - Cornelia de Lange Syndrome) f) patient #6, male, aged 32 mo.,(Pierre-Robin Sequence (PRS) g) patient #7 male, aged 32 mo.(HPG-HPDs)

Anthropometric points marked on the obtained mandible models are described and presented in Figure 2:

1. gn – gnathion – an odd point located at the lowest position on the lower edge of the mandible in the median plane;
2. go.l., go.r – gonion left and right – an even point located at the lowest, posterior and lateral, outer point on the mandible angle;
3. id – infradentale – an odd point located at the junction of the median line of the body with the line connecting upper edges of dental alveoli of both lower central incisors;
4. kdl.l., kdl.r. – kondylion laterale left and right – an even point located on the outer, lateral, most protruded edge of the mandible head;
5. kdm.l., kdm.r. – kondylion mediale left and right – an even point located most centrally on the joint process on the mandible head;
6. kr.l., kr.r. – koronion left and right – an even point located at the highest point of the temporal process of the mandible;
7. ml.l., ml.r. – mentale left and right – an even point located at the lowest point on the edge of the mental foramen.

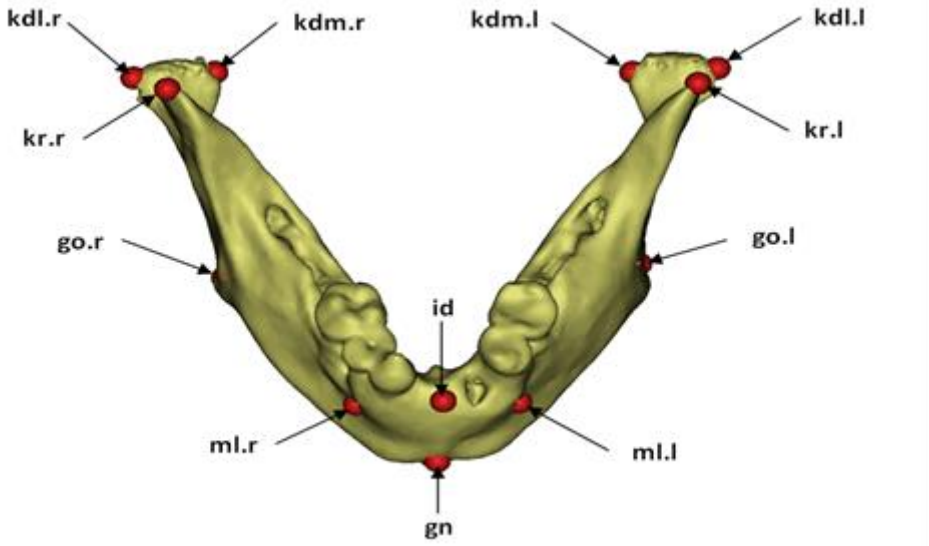

Figure 2. Anthropometric points of the mandible.

On the basis of the above-mentioned points, the linear and angular measurements were conducted **(Table 2)**. As a result, the indices describing shape and proportions of the mandible were calculated:

$i_1 = \frac{go.l-go.r}{kdl.l-kdl.r} \times 100$ the lower the value, the narrower the lower part of the mandible;

$i_2 = \frac{go.l-go.r}{gn-kdl} \times 100$ the higher the value, the smaller the length of the mandible;

$i_3 = \frac{gn-go}{gn-kdl} \times 100$ the higher the value, the shorter the mandible body,

$i_4 = \frac{gn-id}{gn-go} \times 100$ the higher the value, the greater the height of the mandible symphysis;

$i_5 = \frac{go-kdl}{go-gn} \times 100$ the lower the value, the shorter the mandible ramus.

Table 2. Anthropometric measurements and angles describing mandibles.

LENGTH	DIMENSION	DESCRIPTION
Breadth measurements		
go_l–go_r	Bigonial breadth	Odd measurement; distance between points on the mandible angles on the right and left side
kdl_l–kdl_r	Bicondylar breadth external	Odd measurement; distance between the most posterior points on the right and left condylar process
kdm_l–kdm_r	Bicondylar breadth internal	Odd measurement; distance between the most interior points on the right and left condylar process
kr_l–kr_r	Bicoronial breadth	Odd measurement; distance between points at the tip of the coronoid process of the mandible
ml_l–ml_r	Bimental breadth	Odd measurement; distance between the two mental foramina
Length measurements		
gn-go	Length of the mandibular body	Even measurement; line of the mandible base, the most distant point of the mandible in the median line and the mandibular angle point
gn-kdl	Total length of the mandible	Distance between the lowest point of the mandible in the median line and the most lateral point on the joint process
Height measurements		
id-gn	Height of the mandibular symphysis	Odd measurement; distance between the lowest point of the mandible symphysis and the point located centrally between lower incisors
go-kdl	Height of the ramus	Even measurement; distance between the point on the mandible angle and the most posterior point on the condylar process
Angles		
gonial (alpha)	Gonial angle	Even measurement; obtuse angle at the back of the mandible formed by the intersection of the vertical and horizontal portions of the jaw
go-gn-go	Angle of the mandible	Odd measurement; angle between lines defining the mandibular body length, on the right and left side
beta	Mental angle	Odd measurement; angle between the mandible base and the line linking the mental protuberance with the id point.

To compare measurements and indices of the analyzed mandibles with the data of healthy mandibles, the measurements performed on skeletal remains were used. The mandibles of newborns come from the collection of the Chair of Anatomy, Histology and Embryology of the Medical Institute in Płowdiw (Bulgaria) – 77 male mandibles and 58 female ones. The mandibles of infants come from the collection of the Museum of the Chair of Normal Anatomy of the Medical Anatomy in Poznań and other various research centres in Poland. The

measurements were performed by Malinowski and were presented in a monograph in the form of mean values divided into age groups encompassing the first three years of life [19]

Results

All measurements were performed on the basis of the data obtained from computed tomography. The linear and angular values as well as indices were presented in comparison with available peer data from the same age groups [20]. The comparisons of the measurements showed that the mandibles with malformations in children in the first year of age (patients 1-3) were smaller (narrower, lower and shorter) against a background of the mean values obtained for healthy children in this age group (**Table 3**). The breadth of the mandibles in two children in the first half year of their life was even smaller than the width of the newborn's mandibles. Also, the distortion of the mandible proportions was clearly visible (**Table 4**). It revealed itself first of all in the ratio of the breadth to the length of the mandible (i2=go-go/gn-kdl) and the ratio of the height of the mandibular ramus to the length of the mandible body (i5=go-kdl/go-gn and i3=gn-go/gn-kdl). The mandibles in children (patients 1-3) with malformations (in the first year of age) were short in relation to their length and were characterized by the low mandible ramus in relation to the length of the mandible body.

Ankylotic mandibles in two ill children at 16 and 18 months of age, in comparison with mean reference values for two-year-old children, were clearly shorter and lower, whereas the breadth of the mandibles was smaller (patient 4) or comparable (patient 5) with reference values of the mandibles in healthy children (**Table 3**). The mandibles of the two-year-old children with genetic defects still showed distorted proportions; they were shorter in relation to the breadth and had the low mandible ramus in relation to the length of the mandible body **(Table 4).**

The mandible in a three-year-old boy with PRS (patient 6) was characterized by greater breadth in comparison with model values, however, it was shorter. A three-year-old patient with HPG-HPDs was characterized by the mandible of smaller dimensions of breadth, length and height **(Table 3).** Proportions of the mandibles in both three-year-olds were incorrect, namely they were shorter in relation to their breadth and had low ramus in relation to their body **(Table 4).**

The comparison of the values of the mandible angles (alpha) revealed high changeability among the examined children. In four cases out of seven, the alpha angle was at least on one side greater than the given mean value of the angle in the peer group.

Apart from generally smaller dimensions of the ankylotic mandibles in examined children, both in the case of linear and angular measurements, there was asymmetry between the right and left side reaching even 5.31 mm in the length of the mandible body (gn-go) in patient # 3 with SGS at the age of 9 months and the 8.07 degree for the alpha angle in patient #4 with idiopathic ankylosis at the age of 16 months. The occurring asymmetry of the measurements is reflected in the indices calculated on their grounds, for the left and right side respectively **(Table 4).**

Table 3. Individual measurements of mandibles in children with temporomandibular joint ankylosis against a background of available reference values [20].

Measurement	Malinowski (2003)	Patient #1	Patient #2	Patient #3	Malinowski (2003)	Patient #4	Patient #5	Malinowski (2003)	Patient #6	Patient #7	Malinowski (2003)
Sex		male	female	female		female	male		male	male	
Diagnosis[a]		HPG-HPDs	NGD	SGS		IDP ank.	CdLS		PRS	HPG-HPDs	
Age [months]	Newborns[b]	4 months	5 months	9 months	1-12 months	16 months	18 months	13-24 months	32 months	32 months	25-36 months
Age [years]	1st year of age					2nd year of age			3rd year of age		
Breadth measurements											
go_l–go_r	49.0-49.5	44.81	44.76	61.14	66.3	65.39	70.54	71.1	77.18	60.68	72.3
kdl_l–kdl_r	62.9-64.4	61.28	54.29	70.70	79.7	81.52	85.00	84.6	91.10	85.39	88.8
kdm_l–kdm_r	-	40.86	40.65	56.92	-	65.16	60.46	63.0	67.44	61.05	65.3
kr_l–kr_r	-	58.31	49.53	65.52	-	80.78	82.88	-	73.54	77.68	-
ml_l–ml_r	-	14.93	15.66	27.24	-	35.42	36.00	-	33.66	23.74	-
Length measurements											
gn–go	36.7-37.5	L: 36.28 P: 34.19	L: 33.26 P: 29.94	L: 41.27 P: 46.57	50.0	L: 47.78 P: 50.45	L: 51.61 P: 47.93	57.2	L: 51.03 P: 49.97	L: 48.97 P: 46.68	57.8
gn–kdl	-	L: 51.73 P: 49.17	L: 41.61 P: 40.79	L: 59.65 P: 60.45	-	L: 65.73 P: 66.20	L: 61.72 P: 58.71	-	L: 61.17 P: 61.77	L: 79.58 P: 78.90	-
Height measurements											
id–gn		14.64	10.14	15.76	19.3	20.65	19.30	20.3	27.21	17.80	20.7
go–kdl	18.6-18.7	L: 18.59 P: 18.14	L: 11.92 P: 14.31	L: 23.23 P: 17.54	32.7	L: 24.68 P: 25.07	L: 13.05 P: 13.24	36.1	L: 18.03 P: 17.17	L: 36.43 P: 37.89	38.6
Angles											
Alfa (gonial)	147.33	L: 140.77 P: 142.62	L: 133.42 P: 134.34	L: 133.21 P: 138.47	137.2	L: 136.01 P: 127.94	L: 135.55 P: 143.15	139.0	L: 130.61 P: 133.92	L: 136.29 P: 138.76	135.1
Con (go-gn-go)		77.79	90.30	91.13		81.89	89.78		99.89	78.83	
Beta	80.67	56.83	100.93	80.87	93.5	80.06	100.55	88.4	92.15	71.93	74.1

[a] Diagnosis:PRS - ……; SGS - ……; CdLS - …..

[b] Values have been provided separately for males and females (lower values are characteristic of females)

Table 4. Individual indices describing proportions of mandibles in children with temporomandibular joint ankylosis

Measurement	Malinowski (2003)	Patient #1	Patient #2	Patient #3	Malinowski (2003)	Patient #4	Patient #5	Malinowski (2003)	Patient #6	Patient #7	Malinowski (2003)
Sex		male	female	female		female	male		male	male	
Diagnosis[a]		HPG-HPDs	NGD	SGS		IDP ank.	CdLS		PRS	HPG-HPDs	
Age [months]	Newborns[b]	4 months	5 months	9 months	1-12 months	16 months	18 months	13-24 months	32 months	32 months	25-36 months
Age [years]	1st year of age					2nd year of age			3rd year of age		
i_1 go-go / kdl-kdl	76.9-77.9	73.13	82.46	86.48	83.2	80.21	82.98	84.0	84.73	71.06	81.4
i_2 go-go / gn-kdl	49.6-50.9	L: 86.64 P: 91.13	L: 107.57 P: 109.75	L: 102.50 P: 101.14	65.4	L: 99.48 P: 98.77	L: 114.28 P: 120.15	63.1	L: 126.18 P: 124.96	L: 76.24 P: 76.90	66.8
i_3 gn-go / gn-kdl	71.1-73.6	L: 70.14 P: 69.52	L: 79.94 P: 73.41	L: 69.19 P: 77.04	59.0	L: 72.70 P: 76.20	L: 83.61 P: 81.65	56.2	L: 83.43 P: 80.90	L: 61.54 P: 59.16	53.6
i_4 gn-id / gn-go	36.5-36.2	L: 40.34 P: 42.81	L: 30.49 P: 33.87	L: 38.19 P: 33.84	38.6	L: 43.22 P: 40.93	L: 37.39 P: 40.25	35.5	L: 53.32 P: 54.46	L: 36.35 P: 38.13	35.8
i_5 go-kdl / go-gn	58.2-58.3	L: 51.23 P: 53.05	L: 35.84 P: 47.79	L: 56.29 P: 37.66	62.7	L: 51.65 P: 49.69	L: 25.30 P: 27.62	67.7	L: 35.33 P: 34.36	L: 74.40 P: 81.17	65.1

Discussion

The work presents the dimensions of mandibles in children with different etiology of pathology within the temporomandibular joint, from isolated idiopathic ankylosis to craniofacial malformations in genetic syndromes, such as PRS - Pierre Robin Sequence, SGS - Schinzel-Giedion Syndrome, CdLS - Cornelia de Lange Syndrome and Hanhart Syndrome [23]. Morphometric data obtained thanks to the CT were compared with available dimensions of children's regular mandibles. The presented ankylotic mandibles, irrespective of the cause of the adhesion and degree of the mandible immobility, show some characteristic features, such as smaller direct measurements and distorted proportions. Early (prenatal) occurrence of developmental and growing disorders is confirmed by smaller (than in healthy newborns) dimensions of the mandibles in the first half year of the children's lives. Smaller dimensions and distorted proportions (short and low mandible) were also found in older children, irrespective of the syndrome. Moreover, the occurrence of idiopathic ankylosis in one child (patient #4) without other developmental anomalies was also connected with smaller dimensions and irregular proportions of the mandible, which lasted in further years [5][6][16][26]. No genetic syndrome was diagnosed in patient #4. Nevertheless, the occurrence of congenital ankylosis not co-existing with a genetic syndrome does not prove its lack or the mutation of the gene. Due to this fact, a genetic test should be still carried out in order to confirm or eliminate the possibility of occurrence of mutations in such a child. A possible discovery will make it possible to indicate directions of the diagnosis of the remaining anatomical structures of the craniofacial area / cranium and enable thus an objective assessment of the mandible with ankylotic complications.

This work involved a morphometric assessment of the mandible in children with temporomandibular joint ankylosis by means of computed tomography. The analysis was based on the data obtained from CT imaging, which was an indispensable tool in the patients' diagnosis. A clear advantage of the procedure was an easy access to information. The obtained results were compared to the data presented by Malinowski (2003), who made use of the bone material from several Polish collections. This fact is a certain limitation to the evaluation of morphometric features as Malinowski's research was based on the measurements performed directly on the mandibles subjected beforehand to preparatory proceedings. In addition, such a solution makes it difficult to obtain comparative material; its quantity and accessibility is limited. Due to this fact, the method based on the analysis of the information obtained from CT imaging enables the creation of a greater database with a larger amount of information on the mandible morphometry (e.g. surface area, volume, etc.). This will allow better understanding

of anatomical changes occurring in the population and enable the determination of growth standards.

The presented morphometric assessment of the mandibles with TMJ ankylosis was conducted at a certain age of the children. Moreover, the morphometric measurements were expressed using absolute parameters. In the future, it is planned to connect the mandible morphometry with other parameters describing the growth and development of the whole body (body height and mass, fat deposition). Such indices would make it possible to evaluate the child's features during the development and compare them to the features of other children being in the same developmental phase.

In order to properly conduct the morphometric measurements it was essential to know the anatomical structure of the mandible. The Mimics software provided all necessary tools for the performance of a reliable analysis. that one of the main objectives of our study is to standardize the diagnostic procedure in assessing mandibular geometry deformities. the main intention of this study was to initiate a process that could, in the future, lead to the development of more defined and useful assessment indicators. At this stage, based on the limited number of cases, the goal was to present a novel approach and evaluate its potential value in the diagnostic context. Authors are aware of the limitations posed by the small number of cases analysed in this study, but the presented procedure, innovative diagnostic approach and obtained results may be useful in the assessment of an anomaly and provide guidelines for physicians who decide about further treatment. The computed tomography and other advanced imaging examinations have been used so far by physicians to diagnose patients and then plan mainly surgical treatment. The data and knowledge of possible structural disorders are directly linked to the predicted dysfunctions. This may refer to the mobility of the mandible itself, but also to breathing and ingestion. The correlation of the mandible structure disorders with the occurring dysfunctions is the key information for the team of specialists treating the disorder. Also, full understanding of the deformity enables the surgeon to precisely plan the surgical treatment. The possibility of determining osteotomy or resection sites of the ankylotic mass (particularly in the case of frequent re-ankyloses requiring new resections of much larger bone masses) enables effective rehabilitation and decreases the risk of post-surgical complications [9].

Such planning includes preparation of adequate orthopaedic and surgical equipment, such as prostheses, implants and other restoration or prosthetic appliances. The planning process also involves specialists in the scope of biomedical engineering, who develop necessary mathematical models and materials for the surgical procedures. In the authors' opinion, further treatment and, in particular, broadly-understood rehabilitation should also make use of the

imaging methods, which should become the standard procedure [10]. Such a procedure will make it possible to indicate possibilities and, what is even more important, the limitations to be overcome in the course of physiotherapeutic, orthodontic, logopedic, dietary and the like activities. The interpretation of the CT, MRI, USG and X-ray imaging of the tissues with developmental anomalies and changes caused by disease or trauma should be included as a standard element in the education of several medical and therapeutic specializations. It should be noted that the above-described disorders belong to rare diseases and each description encompassing a larger number of patients is invaluable for the teams of specialists treating such patients. The understanding of craniofacial anomalies involves the knowledge of a basic wide spectrum of both regular craniofacial morphology and dysmorphology. The anomalies should be interpreted from a perspective of anatomy and pathology, in comparison with regular structures, taking into consideration their development as well as the growth rate and processes. It is worth defining which of the anomalies represent the earliest or the primary defect in morphogenesis and whether all of them can be ascribed to single problems in morphogenesis. The awareness of these dependences plays the key role in the determination of the etiology of craniofacial anomalies, understanding of the pathogenesis of such conditions, assessment of a risk of recurrence, development of the therapy and preventive methods in order to successfully treat such cases and achieve desired therapeutic results.

Declarations

1. **Ethics approval and consent to participate**

- The case study was conducted according to the guidelines of the Declaration of Helsinki and approved by the Institutional Review Board of Head and Neck Surgery Clinic for Children and Young Adults (KB 430-15/17-02.03.2017).
- The research were performed with approval of the University Bioethics Committee for Scientific Research at the University of Physical Education in Katowice (Resolution No. 1/2021)
- Informed consent was obtained from all parents and/or legal guardians of patients involved in the case study.
- The authors confirm that the patient data used in the study are anonymous and do not enable identification.

2. **Consent for publication:** The Authors hereby consents to publication of this manuscript in any and all Springer Nature journals. The patients' parents and legal guardians consented to the publication of their identifiable data in an online, open-access journal.
3. **Data Availability Statement:** Data available within the article.
4. **Funding:** This study received no external funding.
5. **Conflicts of Interest:** The authors declare no conflict of interests.

References

[1] Al-Hudaid, A., Aldialami, A., Helmi, J., Al-Wesabi, M., & Madfa, A. (2017). Management of temporomandibular joint ankylosis in Yemeni children by metatarsal bone grafts. Journal Of Oral Research, 6(8), 216-221. doi:10.17126/joralres.2017.060

[2] Commander, S. J., Chamata, E., Cox, J., Dickey, R. M., & Lee, E. I. (2016). Update on Postsurgical Scar Management. Seminars in plastic surgery, 30(3), 122–128. https://doi.org/10.1055/s-0036-1584824

[3] Corso, P.F., Machado, R.A., Gerber, J.T., Deliberador, T.M., Costa, D.L., Klüppel, L.E., Rebellato, N.L.B., Brancher, J.A., Küchler, E.C., Coletta, R.D., Scariot, R. (2021) Mutations in the osteoprotegerin−encoding gene are associated with temporomandibular joint ankylosis. Oral and Maxillofacial Pathology, 133(3), 308-314. https://doi.org/10.1016/j.oooo.2021.08.017

[4] Dhupar, V., Akkara, F., Khandelwal, P., & Louis, A. (2018). Zygomatico-coronoid Ankylosis as Sequel of Inadequate Treatment. Annals of maxillofacial surgery, 8(1), 158–161. https://doi.org/10.4103/ams.ams_107_15

[5] Dowgierd, K., Kawlewska, E., Joszko, K., Kropiwnicki, J., & Wolanski, W. (2023). Biomechanical Evaluation of Temporomandibular Joint Reconstruction Using Individual TMJ Prosthesis Combined with a Fibular Free Flap in a Pediatric Patient. Bioengineering (Basel, Switzerland), 10(5), 541. https://doi.org/10.3390/bioengineering10050541

[6] Dowgierd, K., Lipowicz, A., Kulesa-Mrowiecka, M., Wolański, W., Linek, P., & Myśliwiec, A. (2022). Efficacy of immediate physiotherapy after surgical release of zygomatico-coronoid ankylosis in a young child: A case report. Physiotherapy theory and practice, 38(13), 3187–3193. https://doi.org/10.1080/09593985.2021.1952672

[7] Dowgierd, K., Pokrowiecki, R., Kulesa Mrowiecka, M., Dowgierd, M., Woś, J., Szymor, P., Kozakiewicz, M., Lipowicz, A., Roman, M., & Myśliwiec, A. (2022). Protocol for Multi-Stage Treatment of Temporomandibular Joint Ankylosis in

Children and Adolescents. Journal of clinical medicine, 11(2), 428. https://doi.org/10.3390/jcm11020428

[8] Gil-da-Silva-Lopes, V. L., & Luquetti, D. V. (2005). Congenital temporomandibular joint ankylosis: clinical characterization and natural history of four unrelated affected individuals. The Cleft palate-craniofacial journal : official publication of the American Cleft Palate-Craniofacial Association, 42(6), 694–698. https://doi.org/10.1597/04-027.1

[9] Giudice, A., Barone, S., Belhous, K., Morice, A., Soupre, V., Bennardo, F., Boddaert, N., Vazquez, M. P., Abadie, V., & Picard, A. (2018). Pierre Robin sequence: A comprehensive narrative review of the literature over time. Journal of stomatology, oral and maxillofacial surgery, 119(5), 419–428. https://doi.org/10.1016/j.jormas.2018.05.002

[10] Gomes K., Sousa B., Melo K., Macedo P., Resende Z., Gonçalves E., Resende J., Urbano E. (2019). Virtual planning and simulation in facial asymmetry cases: case report and literature review. International Journal of Oral and Maxillofacial Surgery, 48(1), 12. DOI|:10.1016/j.ijom.2019.03.032

[11] Gulsen, A., Sibar, S., & Ozmen, S. (2018). Orthognathic treatment of facial asymmetry due to temporomandibular joint ankylosis. Archives of plastic surgery, 45(1), 74–79. https://doi.org/10.5999/aps.2016.01774

[12] Hutchinson, E. F., L'Abbé, E. N., & Oettlé, A. C. (2012). An assessment of early mandibular growth. Forensic science international, 217(1-3), 233.e1–233.e2336. https://doi.org/10.1016/j.forsciint.2011.11.014

[13] Kaban, L. B., Bouchard, C., & Troulis, M. J. (2009). A protocol for management of temporomandibular joint ankylosis in children. Journal of oral and maxillofacial surgery : official journal of the American Association of Oral and Maxillofacial Surgeons, 67(9), 1966–1978. https://doi.org/10.1016/j.joms.2009.03.071

[14] Kulesa-Mrowiecka, M., Piech, J., Dowgierd, K., & Myśliwiec, A. (2021). Physical therapy of temporomandibular disorder in a child with arthrogryposis multiplex congenita: A case report and literature review. CRANIO®, 42(1), 25–32. https://doi.org/10.1080/08869634.2021.1890453

[15] Leone, M. P., Palumbo, P., Palumbo, O., Di Muro, E., Chetta, M., Laforgia, N., Resta, N., Stella, A., Castellana, S., Mazza, T., Castori, M., Carella, M., & Bukvic, N. (2020). The recurrent SETBP1 c.2608G > A, p.(Gly870Ser) variant in a patient with Schinzel-Giedion syndrome: an illustrative case of the utility of whole exome

sequencing in a critically ill neonate. Italian journal of pediatrics, 46(1), 74. https://doi.org/10.1186/s13052-020-00839-y

[16] Lipowicz, A., Kawlewska, E., Larysz, D., Kostyra, P., Graja, K., Joszko, K., Gzik-Zroska, B. & Wolański, W. (2023). Evaluation of mandibular geometry in healthy children aged 0–1 year – a pilot study. Acta of Bioengineering and Biomechanics, 25(3) 93-102. https://doi.org/10.37190/abb-02312-2023-03

[17] Lipowicz, A., Wolański, W., Kawlewska, E., Zwolska, P., Kulesa−Mrowiecka, M., Dowgierd, K., Linek, P., Myśliwiec, A. (2021). Evaluation of mandibular growth and symmetry in child with congenital zygomatic−coronoid ankylosis. Symmetry, 13(9), 1634. https://doi.org/10.3390/sym13091634

[18] Lipski, M., Tomaszewska, I. M., Lipska, W., Lis, G. J., & Tomaszewski, K. A. (2013). The mandible and its foramen: anatomy, anthropology, embryology and resulting clinical implications. Folia morphologica, 72(4), 285–292. https://doi.org/10.5603/fm.2013.0048

[19] Lum, V., Goonewardene, M. S., Mian, A., & Eastwood, P. (2020). Three-dimensional assessment of facial asymmetry using dense correspondence, symmetry, and midline analysis. American journal of orthodontics and dentofacial orthopedics : official publication of the American Association of Orthodontists, its constituent societies, and the American Board of Orthodontics, 158(1), 134–146. https://doi.org/10.1016/j.ajodo.2019.12.014

[20] Malinowski A. [in Polish] Ontogeneza Żuchwy Ludzkiej, Wydawnictwo Uniwersytetu Zielonogórskiego, Zielona Góra 2003

[21] Marszałek-Kruk, B. A., Wójcicki, P., Dowgierd, K., & Śmigiel, R. (2021). Treacher Collins Syndrome: Genetics, Clinical Features and Management. Genes, 12(9), 1392. https://doi.org/10.3390/genes12091392

[22] Martins, D. L. L., Lima, L. F. S. C., Sales, V. S. de F., Demeda, V. F., Silva, A. L. O. da, de Oliveira, Ângela R. S., de Oliveira, F. M., & Lima, S. B. F. (2014). The Mouth Breathing Syndrome: prevalence, causes, consequences and treatments. A Literature Review. JOURNAL OF SURGICAL AND CLINICAL RESEARCH, 5(1), 47–55. https://doi.org/10.20398/jscr.v5i1.5560

[23] Selicorni, A., Mariani, M., Lettieri, A., & Massa, V. (2021). Cornelia de Lange Syndrome: From a Disease to a Broader Spectrum. Genes, 12(7), 1075. https://doi.org/10.3390/genes12071075

[24] Shivakotee, S., Menon, C. S., Sham, M. E., Kumar, V., & Archana, S. (2020). TMJ Ankylosis Management: Our Experience. Journal of maxillofacial and oral surgery, 19(4), 579–584. https://doi.org/10.1007/s12663-019-01293-y

[25] Souki, B. Q., Lopes, B. P., Pereira, T. B. J., Franco, L. P., Becker, H. M. G., Oliveira, D.D. (2012). Mouth breathing children and cephalometric pattern: does the stage of dental development matter?. International Journal of Pediatric Otorhinolaryngology, 76(6), 837-841. https://doi.org/10.1016/j.ijporl.2012.02.054

[26] Thiesen, G., Gribel, B. F., & Freitas, M. P. (2015). Facial asymmetry: a current review. Dental press journal of orthodontics, 20(6), 110–125. https://doi.org/10.1590/2177-6709.20.6.110-125.sar

[27] Triana, B. E. G., Ali, A. H., León, I. B. (2016). Mouth breathing and its relationship to some oral and medical conditions: physiopathological mechanisms involved. Revista Habanera de Ciencias Médicas, 15(2), 200-212.

[28] Ulański, Ł., Dowgierd, K., & Kozakiewicz, M. (2014). Ankylosis of the Temporomandibular Joint and Its Surgical Reconstruction - Case Reports. Modern Methods of Treatment. Dental and Medical Problems, 51, 519–530.

[29] Yano, T., Taniguchi, M., & Tsuneyoshi, I. (2017). Food Choking in a Patient with Congenital Temporomandibular Joint Ankylosis. Turkish Journal of Anaesthesiology and Reanimation, 45(6), 380–381. https://doi.org/10.5152/TJAR.2017.45143