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Pierre Robin Sequence and Treacher Collins Hypoplastic Mandible Comparison Using Three-Dimensional Morphometric Analysis

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Abstract

Pierre Robin sequence and Treacher Collins syndrome are both associated with mandibular hypoplasia. It has been hypothesized, however, that the mandible may be differentially affected. The purpose of this study was to therefore compare mandibular morphology in children with Pierre Robin sequence to children with Treacher Collins syndrome using three-dimensional analysis of computed tomography (CT) scans. A retrospective analysis was performed identifying children with Pierre Robin sequence and Treacher Collins syndrome receiving CT scans. Three-dimensional reconstruction was performed and ramus height, mandibular body length, and gonial angle were measured. These were then compared to control children with normal mandibles and to clinical norms corrected for age and sex based on previously published measurements. Mandibular body length was found to be significantly shorter for children with Pierre Robin sequence while ramus height was significantly shorter for children with Treacher Collins syndrome. This resulted in distinctly different ramus height/mandibular body length ratios. In addition, the gonial angle was more obtuse in both the Pierre Robin sequence and Treacher Collins syndrome groups compared with the controls. Three-dimensional mandibular morphometric analysis in patients with Pierre Robin sequence and Treacher Collins syndrome thus revealed distinctly different patterns of mandibular hypoplasia relative to normal controls. These findings underscore distinct considerations which must be made in surgical planning for reconstruction.

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Keywords

computed tomography; Pierre Robin sequence; Treacher Collins syndrome; mandibular hypoplasia; morphometric analysis

INTRODUCTION

Mandibular hypoplasia is a frequently encountered anomaly in the pediatric population and can be either congenital or acquired.(1) While mandibular hypoplasia developing from radiation, trauma, or hemifacial atrophy fall into acquired/developmental post-natal etiologies, congenital mandibular hypoplasia can occur in patients with Pierre Robin sequence or Treacher Collins syndrome.(2) In either case, patients with hypoplastic mandibles can suffer from both deficits in form and function.

The triad of micrognathia, glossoptosis, and airway obstruction was first described by Pierre Robin in 1923.(3) In some developing embryos, the tongue occupies a large part of the oronasal cavity, preventing fusion of the palatal shelves leading to a U-shaped cleft palate.(4) Until 1974, this triad was known as Pierre Robin Syndrome. In 1976, Cohen suggested the term “anomalad” to describe the Pierre Robin triad since it was a nonspecific symptom complex that could occur in various situations: 1) isolated, 2) as a component of a syndrome, and 3) associated with other developmental defects that, taken together, do not represent a specific syndrome.(5) Pashayan and Lewis later suggested the name “Robin sequence” to reflect the fact that the anomalies occurred as a developmental sequence.(6–7) The hypothesis was that mandibular hypoplasia in the developing fetus led to a cascade of effects, including U-shaped cleft palate, glossoptosis, and early respiratory problems. Although the triad of anomalies is associated with several eponyms, the condition is now described as “Pierre Robin sequence.”

Treacher Collins syndrome, on the other hand, encompasses a group of closely related defects of the head and neck. In 1900, Edward Treacher Collins first described the essential features to this disorder in his presentation of two cases with notching of the lower lids combined with an underdevelopment of the malar bones.(8) In 1949, Franceschetti and Klein further defined the syndrome and coined the term “mandibulofacial dysostosis.”(9) Treacher Collins syndrome is an autosomal dominant condition with variable expressivity and arises from aberrations in the development of craniofacial structures derived from the first and second branchial arches.(10) Although there is a wide variation in phenotypic expression, the adult patient with fully expressed Treacher Collins syndrome typically has a convex facial profile with a prominent dorsum of the nose, antimongoloid slanting of the eyes, and a retrusive lower jaw and chin.(11) However, the most characteristic finding is malar and mandibular hypoplasia, often with limited formation of the zygomatic complex. The complete form of Treacher Collins syndrome can also be characterized by zygomaticotemporoauromandibular dysplasia and corresponds to a combination of Tessier clefts Nos. 6, 7, and 8.(12) The posteriorly positioned maxilla and steep mandibular plane in Treacher Collins syndrome can lead to poor chin projection and glossoptosis with airway obstruction.(13)

Children with Pierre Robin sequence or Treacher Collins syndrome can demonstrate similar clinical findings. In both cases, children may present with micrognathia. However, different areas of the mandible may be affected. Although numerous studies have addressed the presentation and treatment of Pierre Robin sequence and Treacher Collins syndrome, there have been, to our knowledge, no studies comparing the morphological differences in their mandibles relative to age-matched norms. It has been noted that with Pierre Robin sequence,

the mandibular body is small and the ramus is relatively normal, whereas in children with Treacher Collins syndrome, the ramus is short and the mandibular body is relatively normal. These findings were frequently observed as we participated in the treatment of countless patients during our craniofacial fellowship with Dr. Henry K. Kawamoto. The purpose of this study was to therefore compare the mandibular morphology in children with Pierre Robin sequence to children with Treacher Collins syndrome using three-dimensional morphometric analysis of computed tomography (CT) scans. This work represents the collaborative efforts of four prior “Kawamoto Fellows” (JPB – Fellow #21, SRB – Fellow #13, MTL – Fellow #17, and DCW – Fellow #31).

MATERIALS AND METHODS

A retrospective analysis was performed in concordance with Institutional Review Board guidelines. CT scans of the craniofacial skeleton were obtained from 16 patients who had not had any prior mandibular surgery. Age and sex were recorded at the time of CT scan for each patient. Patients 18 years of age or older were excluded from the study. The study group consisted of five children with Pierre Robin sequence (ten hemi-mandibles), five children with Treacher Collins syndrome (ten hemi-mandibles), and six control children (12 hemi-mandibles). All CT images originally stored in Digital Imaging and Communications in Medicine (DICOM) format were imported into Mimics Innovation Suite (Materialise; Lueven, Belgium) medical image processing software. Semi-automated thresholding of pixel values was performed in order to achieve differentiation between bone and soft tissue. For bone segmentation, the lower threshold used was +226 Hounsfield Units (HU), and the upper threshold was +3071 HU. Three-dimensional surface reconstructions of the skull were then acquired to evaluate the morphology of the mandible (Figure 1).

Using anatomical landmarks and measurements previously described by Shaw *et al.* and Franklin *et al.*, each individual hemi-mandible was evaluated.(14–16) The ramus height was determined by the distance between the condylion superior and the gonion. For patients with Treacher Collins syndrome, the ramus height was determined by the distance between the most superior portion of along the posterior border of the mandible and the gonion. Mandibular body length was measured by the distance between the pogonion and the gonion. Finally, gonial angle was determined by the angle formed between the plane along the inferior border of the mandible and plane along posterior border of the ramus. All measurements were obtained using the 3D spatial coordinate system (x, y, z). Definition and illustration of each landmark used for measurement are provided in Figure 2.

Left and right hemi-mandibles were analyzed for each CT scan. Each value was then compared with the clinical norm corrected for age based on previously published reports. (17–21) Cephalometric longitudinal analyses have determined the approximate normal ramus height at age nine to be 47.6 ± 2.8 mm and the mandibular body length to be 65.0 ± 3.0 mm.(18, 24–26) Age-specific corrections to these values were made by ± 1.2 mm/year and ± 1.6 mm/year for the ramus height and mandibular body length, respectively.(18, 24–26) Differences between measured results and age-corrected clinical norms for each individual patient were then calculated. Mean difference was derived for comparison and statistical analysis between each group. A one-way analysis of variance was used to analyze results and a **p*-value < 0.05 was considered significant.

RESULTS

The mean age of patients receiving CT scans in the Pierre Robin sequence group was 3.20 ± 4.29 years and the mean age for patients in the Treacher Collins syndrome group was 5.40 ± 4.97 years. For patients in the control group with normal mandibles, the mean age was 5.00

± 2.00 years. Measured values (ramus height, mandibular body length, and gonial angle) for each individual patient can be found in Table 1.

Relative to age-corrected norms, the ramus height among patients with Pierre Robin sequence was not statistically significant ($+1.36 \pm 2.00$ mm) (Figure 3A). In contrast, patients in the Treacher Collins group had a significantly shorter rami with a mean difference of -4.20 ± 0.092 mm when compared to age-corrected norms ($*p < 0.05$). There was no statistically significant difference between our control patient ramus heights ($+1.24 \pm 1.77$ mm) and age-corrected norms.

The mandibular body length was significantly shorter for children with Pierre Robin sequence (Figure 3B). Compared to age-corrected clinical norm values, the mean difference for this group was -6.89 ± 0.99 mm ($*p < 0.05$). However, mandibular body length in both the Treacher Collins group and control group were not significantly different from age corrected norms (0.77 ± 1.94 mm and 0.29 ± 2.24 mm, respectively).

Given variation in measurements for each group with age, a ramus height to mandibular body length ratio was also calculated. For patients within the Pierre Robin group, the ramus height to mandibular body length ratio was 0.86 ± 0.03 (Figure 3C). A significantly smaller ratio was observed for patients with Treacher Collins syndrome (0.65 ± 0.02) ($*p < 0.05$). And for patients within the control group, the ramus height to mandibular body length ratio was 0.75 ± 0.02 . This value was also significantly different from both patients with Pierre Robin sequence and Treacher Collins syndrome ($\#p < 0.05$).

Finally, gonial angle was measured for each hemi-mandible in our study. Patients with Pierre Robin sequence were found to have an obtuse gonial angle ($136.26 \pm 2.66^\circ$) (Figure 3D). Similarly, an obtuse gonial angle was also noted for patients with Treacher Collins syndrome ($135.03 \pm 4.70^\circ$). For our control patients with normal mandibles, the gonial angle measured was $126.98 \pm 2.07^\circ$. Gonial angles for patients with Pierre Robin sequence and Treacher Collins syndrome were significantly greater than the control group ($*p < 0.05$).

DISCUSSION

Mandibular hypoplasia can not only cause facial disfigurement, but also significant functional impairment with potential life-threatening airway obstruction secondary to malpositioning of the tongue base. When severe, patients with insufficient bone stock may require reconstruction with rib grafting. In other situations, mandibular lengthening by distraction may offer an alternative strategy to correct mandibular hypoplasia.(22) However, given the three-dimensional complexity and variability of mandibular deformities, multiplanar devices with the ability to distract in three planes: linear, angular, and transverse, have been used by some surgeons.(23)

Fundamentally, one of the most important aspects in mandibular reconstruction is to recognize how the mandible is deficient, and in what manner reconstruction should be performed. In the case of distraction, consideration must be made in pre-operative planning as to orientation of the corticotomy and placement of the device. The desired vector of distraction, based on craniofacial skeletal assessment, should thus facilitate restoration of normal mandibular morphology. However, in many patients with craniofacial anomalies, mandibular deficiencies can be found in three dimensions: anteroposterior (sagittal), mediolateral (coronal), and superoinferior (vertical). It has been noted that in children with Pierre Robin sequence, the mandibular deficiency is primarily in projection of the mandibular body whereas in children with Treacher Collins syndrome, the deficiency is primarily in height of the ramus. In the present study, we attempted to quantify this

observation by evaluating mandibular morphology in children with Pierre Robin sequence and Treacher Collins syndrome using three-dimensional morphometric analysis of CT scans.

Multiple studies have assessed the normal mandible using a variety of techniques including analysis of cephalometric radiographs and dry skull samples. Importantly, mandibular morphology has been shown to be an accurate developmental marker with predictable age related changes. By analyzing 53 pediatric skeletons, Norris found the maximal height of the ramus to be the one measurement most capable of accurately predicting age at the time of death to within six months.(20) Similarly, Franklin and colleagues evaluated multiple age-known samples, and using linear regression they found that ramus height could accurately predict age in the pediatric skeleton.(15) Furthermore, three-dimensional multivariate descriptors of the mandible based on geometric morphometric data were found to be suitable not only for children, but could also be applied to young adults.(15–16) Based on multiple cephalometric longitudinal analyses, investigators have thus determined the approximate normal ramus height and mandibular body length with yearly corrections which have been shown to accurately describe age-specific measurements.(18, 24–26)

As previously mentioned, patients with Pierre Robin sequence have been noted to possess deficiency primarily limited to the mandibular body and this observation has been reported in multiple studies. Glander *et al.* performed cephalometric analysis of the craniofacial skeleton in 49 patients with Pierre Robin sequence and discovered that the absolute length of the maxilla and the body of the mandible were shorter than in normal patients.(27) More recently, Rogers and colleagues showed that in both nonsyndromic and syndromic Pierre Robin sequence patients, mandibular body was smaller and the gonial angle was more obtuse.(28) In addition, ramus height was found to be significantly shorter in syndromic patients including those with Stickler syndrome relative to nonsyndromic patients.(28)

Similar to Pierre Robin sequence, several reports have also described the abnormal morphology of the mandible associated with Treacher Collins syndrome. Steinbacher *et al.* noted a clockwise rotation of the mandibular body and objectively documented the obtuse ramus to body relationship in these patients.(13) Chong *et al.* performed cephalometric analysis of the craniofacial skeleton in 24 Treacher Collins patients and found mandibular dysplasia primarily in ramus height. Interestingly, he also described hypoplasia in other components of mandible including body length relative to age- matched controls, however this was less prominent compared to deficiencies in the ramus.(29)

In the present study, CT analysis of five children with Pierre Robin sequence revealed relatively normal ramus height but significantly shorter mandibular body lengths when compared to age-corrected normal values. Similarly, gonial angle was found to be significantly more obtuse than control patients in this study. These findings thus parallel previously published reports.(27–28) Of particular interest, it has also been debated whether patients with Pierre Robin sequence undergo “catch up” growth in their jaw. Vegter *et al.* evaluated newborns in their first year of life and found mandibles in patients with Pierre Robin sequence to grow but maintain a proportionately smaller size relative to normal mandibles.(30) Daskalogiannakis and colleagues came to the same conclusion, as they found patients with Pierre Robin sequence to have significantly smaller mandibles with differences that did not change after the age of 5 years.(31) In our study, while we did note a trend toward decreased difference in mandibular body length from age-corrected norms in our older patients, the oldest at 11 years of age was still more than 5 mm deficient. An additional patient identified at age 19 not included in the study was also found to have a deficient body length relative to normal adult mandibles. Collectively, these findings thus suggest that smaller mandibles in patients with Pierre Robin sequence do not undergo “catch up” growth and continue to remain smaller than similarly aged mandibles.

In regards to the five patients with Treacher Collins syndrome, ramus height was found to be significantly less than age-corrected normal values while mandibular body length was relatively normal. Gonial angle, like in patients with Pierre Robin sequence, was similarly more obtuse than in our control patients. Again, these findings are in agreement with previously published reports.(13, 29) And akin to that noted in patients with Pierre Robin sequence, no “catch up” growth was appreciated in ramus height deficiency with age for patients with Treacher Collins syndrome.

It may be argued that use of calculated age-corrected normal values to standardize ramus height and mandibular body length for each of our patients can be limited, particularly at extreme ages. Mandibular growth likely occurs in a non-linear fashion which can introduce error into our calculations. Nonetheless, among the six control patients with normal mandibles in our study, calculated age-corrected values for ramus height and mandibular body length did not significantly vary from that measured. Furthermore, individual ramus height to mandibular body length ratios were performed for each patient, and for those with Pierre Robin sequence, these values were consistently greater than published norms.(15–16, 24–26) In contrast, each patient with Treacher Collins syndrome had ramus height to mandibular body length ratios well below the norm. And as expected, this ratio for our control group patients did not significantly vary from published normal values.(15–16, 24–26) These data therefore concur with those conclusions drawn from calculated age-corrected standards.

Importantly, interpretation of our findings must be made keeping in mind the number of patients with Pierre Robin sequence and Treacher Collins syndrome undergoing CT scanning. As not all patients with either condition routinely undergo this form of imaging, this necessarily limits our sample size for each group. Furthermore, it may be suggested that our results may be somewhat biased by more severe cases necessitating surgical intervention who thus receive CT scans. In light of this, a larger multi-center effort combining craniofacial surgery programs throughout the country may be necessary to allow for development of a more comprehensive patient database. Such a resource would facilitate greater patient numbers for analysis to truly evaluate growth patterns for many craniofacial malformations. Nonetheless, our findings in the present manuscript are congruent with multiple observations and published reports. To our knowledge, this is also the first study to quantify in three dimensions the mandibular deficiencies seen in both Pierre Robin sequence and Treacher Collins syndrome. The data in this study thus lay the groundwork for subsequent, more detailed morphometric studies and highlight important potential pre-surgical considerations which must be made prior to reconstruction of mandibles for patients with either Pierre Robin sequence or Treacher Collins syndrome.

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Figure 1.

Lateral view of reconstructed mandibles. (Top) Mandible from 3 month-old patient with Pierre Robin sequence. Note the relatively short mandibular body length. (Middle) Mandible from 1 month-old patient with Treacher Collins syndrome. Note the nearly absent ramus. (Bottom) Mandible from 3 year-old normal control patient.

NUMBER	LANDMARK	DEFINITION
1	Condylion Superior	The most superior point on the mandibular condyle
2	Gonion	The most lateral external point of the junction between the horizontal body and the ascending rami of the lower jaw
3	Pogonion	The lowest point on the inferior margin of the mandibular body

LETTER	MEASUREMENT	DEFINITION
A	Ramus Height	Distance between Condylion Superior (Point 1) and Gonion (Point 2)
B	Mandibular Body Length	Distance between gonion (Point 2) and pogonion (Point 3)
C	Gonial Angle	The angle formed by the inferior border of the mandibular body and posterior border of the ramus

Figure 2. Schematic showing landmarks employed for measurements. Lateral view (top left) and frontal view (top right) of normal mandible with overlay of landmarks used. (Bottom) Definition of each landmark.

Figure 3.

Mandibular measurements from three-dimensional reconstruction of CT scans. (A) Ramus height for Pierre Robin (red) and normal control (green) groups were not significantly different from age-corrected norms. Patients in the Treacher Collins group (blue) had significantly shorter ramus heights ($*p < 0.05$). (B) Mandibular body length was significantly less in the Pierre Robin group ($*p < 0.05$) while the Treacher Collins and normal control groups were not significantly different from age-corrected norms. (C) Ramus Height to Mandibular Body Length ratio in the Pierre Robin group was significantly greater than the Treacher Collins group ($*p < 0.05$). Both of these groups, however, were significantly different from the normal control group ($\#p < 0.05$). (D) Gonial angle for the

Pierre Robin and Treacher Collins groups were significantly greater than the control group (* $p < 0.05$).

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TABLE 1

Three-Dimensional Measurements of the Mandible from CT Scans

Pierre Robin	Age	Side	Ramus Height (mm)	Mandibular Body Length (mm)	Gonial Angle (degrees)
Patient #1	3 mo	R	37.95	43.55	137.13
	3 mo	L	38.13	43.18	136.96
Patient #2	4 mo	R	35.95	42.66	138.59
	4 mo	L	37.03	42.41	139.86
Patient #3	2 yr	R	38.53	46.24	136.87
	2 yr	L	38.16	46.59	136.04
Patient #4	3 yr	R	44.25	49.50	136.10
	3 yr	L	45.07	48.67	137.55
Patient #5	11 yr	R	52.92	62.94	130.91
	11 yr	L	52.02	62.53	132.63

Treacher Collins	Age	Side	Ramus Height (mm)	Mandibular Body Length (mm)	Gonial Angle
Patient #1	1 mo	R	33.78	49.79	128.57
	1 mo	L	33.74	48.73	128.29
Patient #2	2 yr	R	35.32	52.78	137.94
	2 yr	L	35.49	53.26	136.68
Patient #3	3 yr	R	36.55	59.41	139.06
	3 yr	L	36.71	56.56	139.15
Patient #4	10 yr	R	43.85	69.27	130.05
	10 yr	L	43.91	69.50	132.42
Patient #5	12 yr	R	45.79	70.89	140.17
	12 yr	L	45.67	69.88	138.01

Control	Age	Side	Ramus Height (mm)	Mandibular Body Length (mm)	Gonial Angle
Patient #1	2 yr	R	41.36	54.95	130.74
	2 yr	L	41.28	55.13	129.89

Control	Age	Side	Ramus Height (mm)	Mandibular Body Length (mm)	Gonial Angle
Patient #2	3 yr	R	38.72	54.87	128.45
	3 yr	L	39.28	54.33	128.13
Patient #3	5 yr	R	46.88	58.04	127.33
	5 yr	L	45.42	59.73	127.96
Patient #4	6 yr	R	46.03	61.83	124.97
	6 yr	L	45.77	61.39	125.17
Patient #5	7 yr	R	44.66	60.82	125.89
	7 yr	L	445.87	60.95	125.67
Patient #6	7 yr	R	49.15	65.67	124.63
	7 yr	L	48.87	65.31	124.89