

# Distraction Osteogenesis Normalizes Mandibular Body-Symphysis Morphology in Infants With Robin Sequence



Srinivas M. Susarla, DMD, MD, MPH, \*Kelly N. Evans, MD, †Hitesh Kapadia, DDS, PhD, ‡Nefeli Vasilakou, DDS, §Mark A. Egbert, DDS, || and Richard A. Hopper, MD, MS ¶

**Purpose:** To evaluate changes in mandibular morphology in infants with Robin sequence (RS) after mandibular distraction osteogenesis (MDO) and compare the post-distraction morphology with that in infants without RS and infants with RS who had not undergone MDO.

**Materials and Methods:** Infants with RS treated with MDO were retrospectively evaluated over a 12-year period. All patients had pre-distraction and end-consolidation maxillofacial computed tomograms. Morphologic features of the mandible were divided into ramus and condyle, body and symphysis, and composite measurements. Post-distraction RS mandibular morphology was compared with pre-distraction morphology, as well as to age-matched infants without RS and age-matched infants with RS who had not undergone MDO. Comparisons were done using nonparametric paired-samples analyses.

**Results:** During the study period, 17 patients with RS treated with MDO met the inclusion criteria for the study. The mean ages at distraction and end-consolidation were  $1.95 \pm 3.24$  and  $8.46 \pm 5.99$  months, respectively. The post-MDO mandible was significantly different from the pre-MDO mandible with regard to the ramps-condyle unit and body-symphysis measurements, including development of a more parabolic mandibular arch form ( $P \leq .001$ ). Compared with age-matched non-RS infant mandibles, the post-distraction RS mandibles had similar morphologies. Compared with age-matched non-MDO RS mandibles, the post-distraction mandibles had significantly different morphologies anterior to the gonial angle, including a more parabolic arch form ( $P \leq .006$ ).

**Conclusions:** MDO normalized mandibular morphology in infants with RS, with the greatest effect on measurements anterior to the gonial angle.

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Robin sequence (RS), most commonly defined as the triad of micrognathia, glossoptosis, and airway obstruction, has been a well-known clinical entity since 1934.<sup>1-4</sup> However, there remain several controversies related to the diagnosis and

management of the condition.<sup>5-9</sup> Although there are no uniformly accepted criteria for diagnosis or staging severity, there is consensus about micrognathia as a defining-feature of the condition.<sup>7,8</sup> Historically, treatment was designed to alter the

Received from the Craniofacial Center, Seattle Children's Hospital, Seattle, WA.

\*Assistant Professor, Divisions of Oral and Maxillofacial Surgery and Craniofacial and Plastic Surgery.

†Assistant Professor, Division of Craniofacial Medicine.

‡Assistant Professor, Division of Craniofacial Orthodontics.

§Instructor, Division of Craniofacial Orthodontics.

||Associate Professor, Division of Oral and Maxillofacial Surgery.

¶Professor and Chief, Division of Craniofacial and Plastic Surgery.

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Address correspondence and reprint requests to Dr Susarla: Craniofacial Center, Seattle Children's Hospital, 4800 Sand Point Way NE, Seattle, WA 98105; e-mail: [Srinivas.susarla@seattlechildrens.org](mailto:Srinivas.susarla@seattlechildrens.org)

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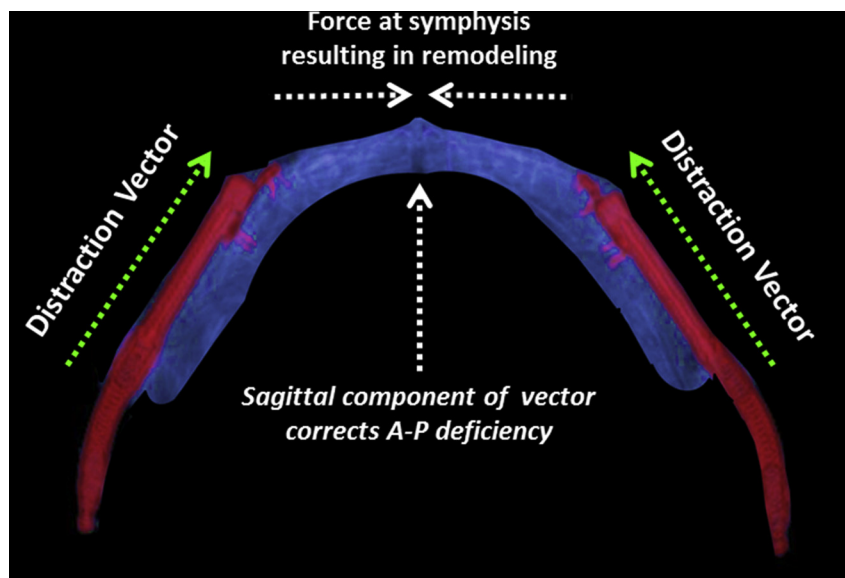
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position of the mandible to open the airway (eg, prone positioning, tongue-to-lip adhesion) or bypass the collapsible upper airway altogether (eg, tracheostomy).<sup>10-12</sup> During the past 15 years, mandibular distraction osteogenesis (MDO) has become an increasingly accepted alternative for management, including in neonates, as a strategy to avoid tracheostomy placement.<sup>13-34</sup> To date, there are several reports in the literature that suggest that treatment with MDO improves mandibular bony volume, decreases the need for tracheostomy and gastrostomy placement, improves sleep architecture and oxygen saturation, favorably alters soft tissue morphology in patients with associated cleft palate, and decreases the apnea-hypopnea index.<sup>15,18,26,27</sup> In addition, the cumulative cost for MDO may be lower than for alternative treatments.<sup>19,20</sup> As such, many centers are using MDO as a first-line treatment for infants with RS who are symptomatic or fail to respond to less invasive maneuvers.

Although there are studies reporting improvements in mandibular bony volume and sagittal position after distraction, there is a paucity of information about MDO-induced changes in RS mandibular morphology compared with age-matched unaffected (normal) infants and age-matched infants with RS who have not undergone MDO.<sup>27-33</sup> Recently, our group defined the differences in mandibular morphology, as assessed by linear, angular, and composite measurements, between infants with RS and age-matched normal infants.<sup>33</sup> The morphology of the RS

mandible was most markedly different anterior to the gonial angle. Specifically, RS mandibles anterior to the gonion had shorter sagittal lengths, shorter inferior border arc lengths, steeper gonial angles, narrower symphyseal angles, were more elliptical in shape, and had a smaller submental cross-sectional area.<sup>33</sup> In contrast, structures posterior to the gonion (ie, ramus and condyle units) were morphologically similar between the RS and unaffected control groups.

The purpose of this study was to assess changes in RS mandibular morphology that occur as a result of MDO with a goal of determining whether the induced change approached or deviated from normal shape. The primary hypothesis was that, after MDO, the RS mandible would achieve a more normal morphology. Digital analysis of linear, angular, and composite measurements of mandibular shape on computed tomographic (CT) scans was used to test this hypothesis. The specific aims were to 1) identify a cohort of patients with RS who were treated with MDO and compare their post-MDO (end-consolidation) measurements with their pre-MDO scans to quantify the change in shape that occurred during the MDO treatment period, 2) compare the post-MDO RS mandible with age-matched control mandibles to determine whether the observed changes in morphology approached or deviated from normal, and 3) compare the post-MDO RS mandible with age-matched RS mandibles that had not undergone MDO to determine where the observed change during MDO treatment was present in untreated RS mandibles.



**FIGURE 1.** Internal distraction device placement, submental 3-dimensional view. The internal devices were fixated at the inferior border of the mandible and therefore had a convergent vector toward the symphysis (green arrows). Metrics before and after distraction are consistent with this convergence effecting changes in the mandibular arch anterior to the gonion that approach normal, without any evidence of morphologic changes posterior to the gonion toward the condyles. A-P, anteroposterior.

**Table 1. DESCRIPTION OF VARIABLES**

|                                        | Definition                                                                                                                                                                     |
|----------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Landmark</b>                        |                                                                                                                                                                                |
| Condylion (Co)                         | Most posterosuperior point on the mandibular condyle                                                                                                                           |
| Articulare (Ar)                        | Junction between the inferior surface of the cranial base and the posterior border of the ascending ramus of the mandible                                                      |
| Gonion (Go)                            | Most posteroinferior point on the angle of the mandible                                                                                                                        |
| Menton (Me)                            | Lowest point on the mandibular symphysis                                                                                                                                       |
| B Point (B)                            | Most concave point on the mandibular symphysis                                                                                                                                 |
| <b>Ramus and condyle measurements</b>  |                                                                                                                                                                                |
| Intercondylar distance (Co-Co)         | Distance between right and left condylion points                                                                                                                               |
| Intergonial distance (Go-Go)           | Distance between right and left gonion points                                                                                                                                  |
| Ramus height (Co-Go)                   | Linear distance between the most superior point on the condyle and gonion                                                                                                      |
| Ramus angle                            | Angulation of the long axis of the ramus relative to the intergonial plane                                                                                                     |
| Gonial angle (Ar-Go-Me)                | Angle formed by connecting the articulare, gonion, and menton                                                                                                                  |
| <b>Body and symphysis measurements</b> |                                                                                                                                                                                |
| Mandibular sagittal length             | Distance between the midpoint of the mandible at the inferior border to the intergonial line                                                                                   |
| Mandibular radial length               | Length of a line drawn from the point of intersection of the intergonial line and midsagittal plane to the inferior border of the mandible at an angle of 45° relative to both |
| Inferior border arc length             | Distance along an arc drawn at the inferior border from gonion to gonion                                                                                                       |
| Intergonial angle (Go-Me-Go)           | Angle formed by right gonion to menton to left gonion, as viewed from inferiorly                                                                                               |
| Symphyseal angle (Go-Me-B)             | Angle formed by connecting the gonion, menton, and B point                                                                                                                     |
| <b>Composite measurement</b>           |                                                                                                                                                                                |
| First linear eccentricity              | Ratio of the mandibular oblique length to the mandibular sagittal length                                                                                                       |
| Mandibular radial-to-sagittal ratio    | Composite measurement of shape, calculated as the ratio of the intermediate axis distance to the mandibular sagittal length                                                    |
| Submental cross-sectional area         | Area bounded by the intergonial plane and an arc drawn along the inferior border of the mandible from gonion to gonion                                                         |

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## Materials and Methods

### STUDY DESIGN AND SAMPLE

This was a retrospective study of infants treated at the Seattle Children's Hospital (Seattle, WA) over a 12-year period. The project was approved by the institutional review board for human studies. The Declaration of Helsinki was followed at all times during this study. The authors identified a subset of 20 patients with RS, diagnosed clinically as the triad of micrognathia, glossoptosis, and airway obstruction, who were treated with MDO and who had maxillofacial CT (mCT; slice thickness, 0.625 to 1.0 mm) scans with multiplanar reformations (axial, sagittal, coronal, and 3-dimensional) in anticipation of surgery. Of these, 17 (85%) had preoperative imaging completed immediately before distraction ( $T_0$ ) and postoperative imaging at end consolidation ( $T_1$ ) and were included as study subjects. The study sample included a heterogeneous group of patients who met the institutional criteria for RS; the authors included patients with isolated findings consistent with RS and those with Stickler syndrome and multisystem anomalies

with associated major chromosomal anomalies without a syndromic diagnosis. The authors excluded patients with diagnoses of Treacher-Collins, Nager, and auriculo-condylar syndrome.

### DISTRACTION SURGICAL TECHNIQUE AND PROTOCOL

The mandible was approached through bilateral submandibular (Risdon) incisions. Mandibular corticotomies were performed bilaterally in an inverted-L fashion with the vertical limb immediately posterior to the lingula and the horizontal limb superior to the inferior alveolar nerve interface with the mandibular foramen. Then, linear uni-vector distraction devices (uni-directional horizontal mandibular distractor, KLS Martin Group, Tuttlingen, Germany; titanium single-vector mandibular distractor, Depuy Synthes CME, West Chester, PA) were fixated bilaterally across the corticotomy gaps at the inferior border with percutaneous activation arms. The distraction appliances were applied with monocortical screws such that the footplates were flush with the lateral

**Table 2. COMPARATIVE DATA FOR STUDY GROUPS**

| Variable                                          | T <sub>1</sub> (n = 17 Infants) |        | T <sub>0</sub> (n = 17 Infants) |        |                      | Unaffected Control (n = 9 Infants)* |        |                      | Robin Sequence Control (n = 11 Infants)* |        |                      |
|---------------------------------------------------|---------------------------------|--------|---------------------------------|--------|----------------------|-------------------------------------|--------|----------------------|------------------------------------------|--------|----------------------|
|                                                   | Mean                            | SD     | Mean                            | SD     | P Value <sup>†</sup> | Mean                                | SD     | P Value <sup>†</sup> | Mean                                     | SD     | P Value <sup>†</sup> |
| Age (mo)                                          | 8.46                            | 5.99   | 1.95                            | 3.24   | <.001 <sup>‡</sup>   | 8.35                                | 6.37   | .62                  | 8.86                                     | 6.12   | .09                  |
| Ramus and condyle measurements                    |                                 |        |                                 |        |                      |                                     |        |                      |                                          |        |                      |
| Intercondylar distance (mm)                       | 70.60                           | 4.23   | 58.72                           | 3.92   | <.001 <sup>‡</sup>   | 69.96                               | 4.27   | .98                  | 66.89                                    | 6.44   | .03 <sup>‡</sup>     |
| Intergonial distance (mm)                         | 61.28                           | 7.55   | 52.00                           | 5.33   | <.001 <sup>‡</sup>   | 61.75                               | 2.75   | .69                  | 58.54                                    | 6.13   | .33                  |
| Ramus length (mm)                                 | 28.51                           | 3.76   | 21.59                           | 3.15   | <.001 <sup>‡</sup>   | 32.03                               | 4.49   | <.001 <sup>‡</sup>   | 26.96                                    | 4.12   | .08                  |
| Ramus angle (°)                                   | 102.99                          | 4.66   | 104.53                          | 12.15  | .492                 | 103.12                              | 3.68   | .94                  | 102.70                                   | 3.61   | .59                  |
| Gonial angle (°)                                  | 145.78                          | 9.49   | 150.82                          | 6.55   | .028 <sup>‡</sup>    | 130.79                              | 4.34   | <.001 <sup>‡</sup>   | 146.80                                   | 4.55   | .46                  |
| Body and symphysis measurements                   |                                 |        |                                 |        |                      |                                     |        |                      |                                          |        |                      |
| Mandibular oblique length (mm)                    | 46.04                           | 7.06   | 35.09                           | 3.66   | <.001 <sup>‡</sup>   | 49.91                               | 4.94   | .02 <sup>‡</sup>     | 41.71                                    | 4.48   | .013 <sup>‡</sup>    |
| Mandibular sagittal length (mm)                   | 38.79                           | 6.94   | 23.32                           | 3.37   | <.001 <sup>‡</sup>   | 40.52                               | 5.62   | .16                  | 29.13                                    | 4.48   | <.001 <sup>‡</sup>   |
| Mandibular radial length (mm)                     | 28.14                           | 4.46   | 22.37                           | 3.00   | .001 <sup>‡</sup>    | 28.79                               | 2.82   | .55                  | 27.32                                    | 3.79   | .23                  |
| Inferior border arc length (mm)                   | 100.18                          | 14.97  | 75.13                           | 7.95   | <.001 <sup>‡</sup>   | 103.65                              | 8.89   | .21                  | 88.57                                    | 10.97  | .004 <sup>‡</sup>    |
| Intergonial angle (°)                             | 68.44                           | 10.45  | 96.70                           | 7.40   | <.001 <sup>‡</sup>   | 71.71                               | 6.79   | .12                  | 91.38                                    | 8.91   | <.001 <sup>‡</sup>   |
| Symphyseal angle (°)                              | 87.67                           | 3.64   | 77.96                           | 5.67   | <.001 <sup>‡</sup>   | 87.72                               | 7.79   | .91                  | 81.25                                    | 3.20   | .001 <sup>‡</sup>    |
| Composite measurements                            |                                 |        |                                 |        |                      |                                     |        |                      |                                          |        |                      |
| Mandibular first eccentricity                     | 0.84                            | 0.05   | 0.66                            | 0.05   | <.001 <sup>‡</sup>   | 0.81                                | 0.04   | .03 <sup>‡</sup>     | 0.70                                     | 0.06   | <.001 <sup>‡</sup>   |
| Radial-to-sagittal length ratio                   | 0.73                            | 0.04   | 0.96                            | 0.09   | <.001 <sup>‡</sup>   | 0.72                                | 0.06   | .31                  | 0.94                                     | 0.03   | <.001 <sup>‡</sup>   |
| Submental cross-sectional area (mm <sup>2</sup> ) | 1,882.86                        | 467.74 | 958.79                          | 201.94 | <.001 <sup>‡</sup>   | 1,973.17                            | 345.63 | .52                  | 1,347.49                                 | 278.36 | .001 <sup>‡</sup>    |

Abbreviations: SD, standard deviation; T<sub>0</sub>, before mandible distraction osteogenesis; T<sub>1</sub>, after mandible distraction osteogenesis.

\* Unaffected and Robin sequence controls were matched by age to the T<sub>1</sub> infants asymmetrically (1:1.9 and 1:1.5, respectively) to ensure full capture of the mandible distraction osteogenesis sample.

<sup>†</sup> P values are for comparisons with the reference group (infants with Robin sequence after mandible distraction osteogenesis).

<sup>‡</sup> Statistically significant ( $P \leq .05$ ) associations.

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border of the mandible and the longitudinal axis of the distraction was parallel to the inferior border, with a resultant horizontal vector that was convergent anteriorly (Fig 1). Activation was completed at a rate of 2.0 mm per day to the end of the activation length of the distractor (15 to 25 mm). The consolidation period was at least twice as long (in days) as the number of days of activation.

#### CONTROL SAMPLES

In addition to paired comparisons between pre- and post-distraction time points (T<sub>0</sub> and T<sub>1</sub>, respec-

tively), the authors compared the post-distraction RS mandible with age-matched mandibles in infants without known craniofacial anomalies or skeletal dysmorphism and age-matched RS mandibles that had not been treated with MDO. An age-matched set of infants was used to assess similarity between the post-distraction mandible and the non-RS mandible. These normative controls had mCT scans for evaluation of clinically evident soft tissue masses (no skeletal dysmorphism was identified at imaging), skull or upper midface trauma (no displaced fractures were identified at imaging), evaluation of head shape

(no evidence of craniosynostosis), or feeding difficulty (no evidence of clefting or skeletal dysmorphology was found). Age-matched infants with RS who had not undergone MDO were included for comparison with control infants for the effects of age on mandibular growth in patients with RS. This older cohort of patients with RS was initially treated with noninvasive methods (eg, nasopharyngeal airway, positioning, etc), but subsequently had CT scans in anticipation of MDO because of persistent airway obstruction. As such, the age of the older cohort was matched to the end-consolidation age of the MDO cohort, all of whom had earlier distraction for severe obstruction. Unaffected controls and RS controls were matched to MDO cases at ratios of 1:1.5 and 1:1.9, respectively, to ensure full capture of the MDO RS group.

#### STUDY VARIABLES

The primary study measurements were radiographic digital metrics of mandibular shape and position and were classified as ramus and condyle, body and symphysis, and composite measurements (Table 1). Ramus and condyle measurements were intercondylar distance (millimeters), intergonial distance (millimeters), ramus height (millimeters), ramus inclination (degrees), and gonial angle (degrees). Symphysis and body measurements were mandibular sagittal length (millimeters), mandibular oblique length (millimeters), mandibular radius (millimeters), inferior border arc length (millimeters), intergonial angle (degrees), and symphyseal angle (degrees). Composite measurements were used to quantify shape and included mandibular first eccentricity (range, 0 [circle] to 1 [parabola]), radius-to-sagittal length ratio, and submental cross-sectional area (square millimeters). These measurements were previously validated by the authors, with a high degree of interobserver agreement between 2 independent observers who made serial measurements over several time points (intraclass correlation for absolute agreement, 0.95 to 0.99;  $P < .001$ ).<sup>33</sup> Measurements were made using 3-dimensional cephalometric imaging software (Dolphin 3D, Dolphin Imaging, Patterson Dental Supply, Redmond, WA). For this study, a single observer performed all measurements.

#### STATISTICAL ANALYSES

De-identified patient data were entered into a database (SPSS 24.0, IBM Corp, Armonk, NY). Nonparametric paired-samples analyses (Wilcoxon signed ranks test) were used to compare the  $T_0$  (before distraction) and  $T_1$  (end-consolidation) time points and to compare the  $T_1$  group with the RS and unaffected control groups. For all analyses, a  $P$  value less

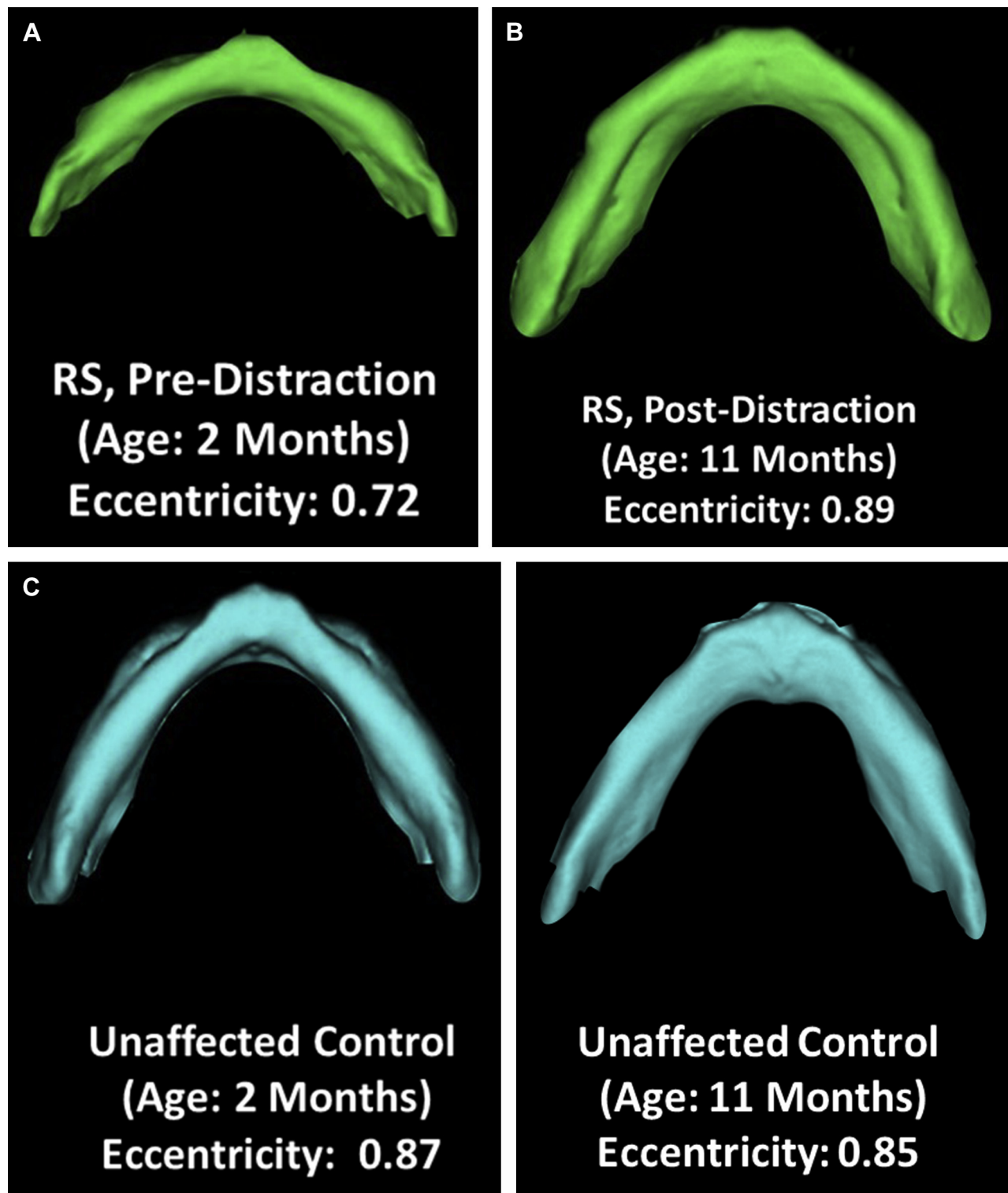
than or equal to .05 was considered statistically significant.

## Results

Comparative data among the various time points and groups are presented in Table 2. The primary study sample was composed of 17 infants with RS who underwent treatment with MDO. Of these infants, 7 had a syndromic association (4 with Stickler syndrome and 3 with other major chromosomal anomalies). The mean age at the time of distraction ( $T_0$ ) was  $1.95 \pm 3.24$  months (median, 1.0 month; range, 6 days to 14 months); the mean age at end consolidation ( $T_1$ ) was  $8.46 \pm 5.99$  months (median, 5.6 months; range, 1.3 to 21.6 months). A comparison of the post-MDO RS mandible with the pre-MDO RS mandible showed significant changes ( $P \leq .03$ ) in all morphologic measurements, except ramus angulation ( $P = .49$ ). Patients in the MDO group had a mean follow-up time of  $5.0 \pm 3.1$  years (median, 4.9 yr). During the follow-up period, 1 patient who had a tracheostomy placed before distraction was successfully decannulated after MDO; a second patient had a tracheostomy placed after distraction for persistent obstruction related to subglottic stenosis. The remaining patients did not require a tracheostomy.

Post-MDO RS mandibles were compared with those of age-matched control infants to assess whether distraction normalized mandibular morphology. Compared with age-matched control mandibles, there were few relevant differences posterior to the gonion. The intercondylar distance ( $P = .98$ ), intergonial distance ( $P = .69$ ), and ramus angulation ( $P = .94$ ) were not significantly different. Ramus length was significantly shorter in the post-distraction RS mandibles ( $28.51 \pm 3.76$  vs  $32.03 \pm 4.49$  mm;  $P < .001$ ) and the gonial angle was steeper ( $145.78 \pm 9.49^\circ$  vs  $130.79 \pm 4.34^\circ$ ;  $P < .001$ ). For body and symphysis measurements, the oblique length remained shorter after MDO ( $46.0 \pm 7.1$  vs  $49.9 \pm 4.9$  mm;  $P = 0.02$ ). The remaining body and symphysis measurements (mandibular sagittal length, mandibular radius, inferior border arc length, intergonial angle, and symphyseal angle) were statistically equivalent between the post-MDO RS mandibles and the age-matched control mandibles ( $P \geq .12$ ). The shape of the post-distraction mandibular arch was comparable to that of the normal mandibular arch and perhaps even overcorrected (eccentricity,  $0.84 \pm 0.05$  vs  $0.81 \pm 0.04$ ;  $P = .03$ ; radial-to-sagittal ratio,  $0.73 \pm 0.04$  vs  $0.72 \pm 0.06$ ;  $P = .31$ ). The submental cross-sectional areas were comparable ( $1,882.86 \pm 467.74$  vs  $1,973.17 \pm 345.63$  mm<sup>2</sup>;  $P = .52$ ).

To assess the effects of distraction versus natural early growth of the RS mandible within the first year

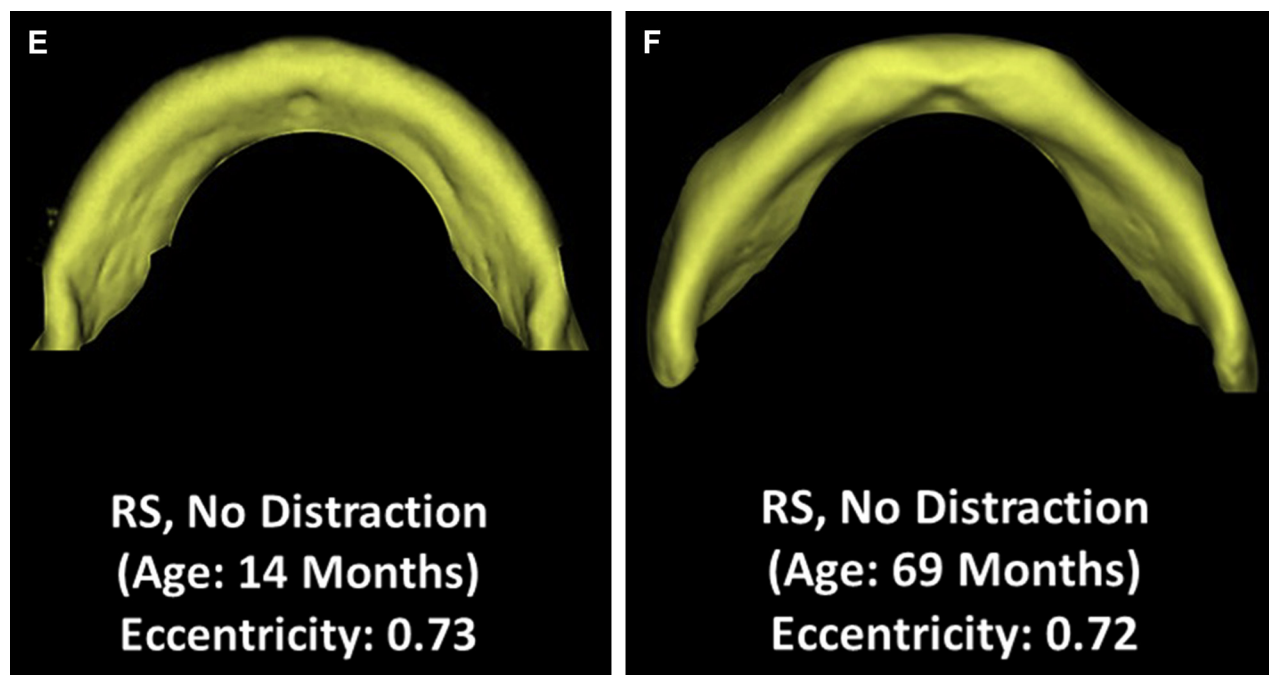


**FIGURE 2.** Differences in body and symphysis mandibular morphology, inferior view. *A*, Infant with RS at 2 months of age. *B*, The same patient after mandibular distrac-tion osteogenesis at 11 months of age. *C*, Control infant at 2 months of age. *D*, Control infant at 11 months of age. (Fig 2 continued on next page.)

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of life, RS MDO mandibles were compared with RS mandibles that did not have MDO (non-MDO RS controls). The mean age in the distrac-tion group at

end consolida-tion was  $8.46 \pm 5.99$  months (median, 5.6 months; range, 1.3 to 21.6 months) compared with  $8.86 \pm 6.12$  months (median, 6.8 months; range,



**FIGURE 2 (cont'd).** E, Patient with RS without mandibular distraction osteogenesis at 14 months of age. F, Patient with RS without mandibular distraction osteogenesis at 69 months of age. RS, Robin sequence.

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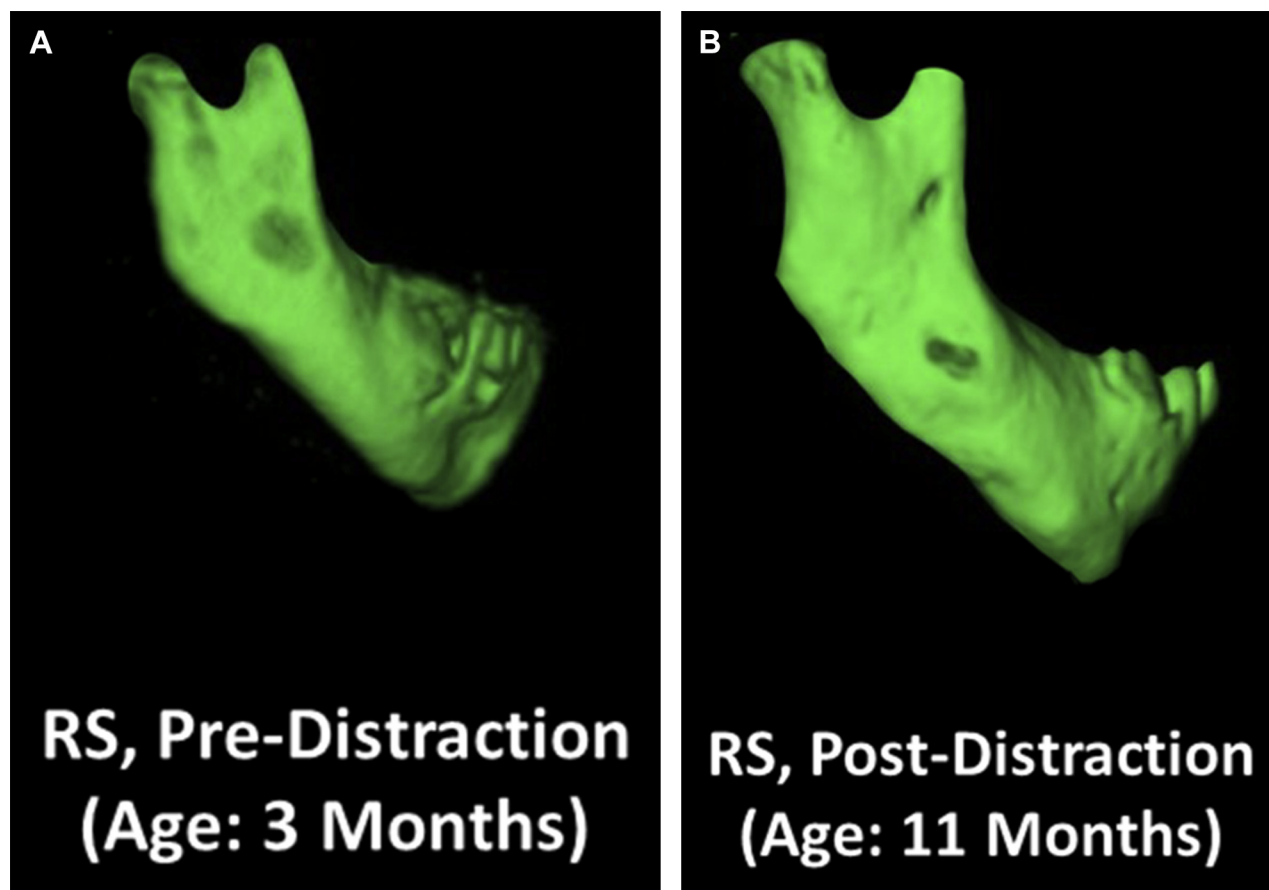
1.3 to 21.9 months) in the non-MDO RS controls ( $P = .09$ ). Of the non-MDO RS controls, 6 had a syndromic association (3 with Stickler syndrome and 3 with major chromosomal abnormalities). Compared with non-MDO RS mandibles, the post-MDO RS mandibles had several notable changes, primarily in the body and symphysis. For ramus and condyle measurements, post-MDO RS mandibles had a wider intercondylar distance ( $70.60 \pm 4.23$  vs  $66.89 \pm 6.44$  mm;  $P = .03$ ). Anterior to the gonion, the post-MDO RS mandibles had increased oblique length ( $46.04 \pm 7.06$  vs  $41.71 \pm 4.48$  mm;  $P = 0.01$ ), sagittal length ( $38.79 \pm 6.94$  vs  $29.13 \pm 4.48$  mm;  $P < 0.001$ ), and inferior border arc length ( $100.18 \pm 14.97$  vs  $88.57 \pm 10.97$  mm;  $P = 0.004$ ). The post-distraction mandible had a narrower intergonial angle ( $68.44 \pm 10.45$  degrees vs  $91.38 \pm 8.91$  degrees;  $P < 0.001$ ) and a more orthogonal symphyseal angle ( $87.67 \pm 3.64$  degrees versus  $81.25 \pm 3.2$ ;  $P = 0.001$ ). The post-MDO RS mandible had a more parabolic shape (eccentricity,  $0.84 \pm 0.05$  vs  $0.70 \pm 0.06$ ;  $P < .001$ ) and became less round (radial-to-sagittal ratio,  $0.73 \pm 0.04$  vs  $0.94 \pm 0.03$ ;  $P < .001$ ). The submental cross-sectional area increased after distraction ( $1,882.86 \pm 467.74$  vs  $1,341.0 \pm 276.41$  mm<sup>2</sup>;  $P < .001$ ).

## Discussion

The advent of DO in the early 1990s allowed for an additional option for treatment of the patient with RS

specifically geared toward correcting mandibular bony deficiency.<sup>34-36</sup> Since that time, an understanding of the morphology of the RS mandible has improved and, in severe cases, distraction appears to be a promising modality to avoid tracheotomy placement and allow for decannulation in patients who have tracheostomies. Data from the literature suggest that distraction advancement of the RS mandible results in increased bony volume and improved sagittal position.<sup>27</sup> However, it remains unclear whether these post-MDO changes normalize mandible morphology to achieve the beneficial effect on the upper airway or whether they distort morphology, resulting in a mandible that deviates from normal. As such, the purpose of this study was to assess the effects of MDO on the morphology of the RS infant mandible compared with the mandibular morphology in infants without known craniofacial syndromes or skeletal dysmorphism and age-matched RS mandibles that had not undergone distraction.

The changes identified as a result of distraction were generally more profound anterior to the gonial angle (Figs 2, 3). Failure of mandibular growth is postulated as the primary pathogenic problem in RS, with several groups hypothesizing that the primary deficiency might be at the site of initial ossification of the mandible, which is located anterior to the gonial angle.<sup>32,37-40</sup> Proximal and distal growth from this site gives rise to the ramus and condyle unit and the body and symphysis, respectively. A putative



**FIGURE 3.** Differences in mandibular morphology, right lateral 3-dimensional view. A, Infant with RS at 2 months of age. B, Same patient after mandibular distraction osteogenesis at 11 months of age. (**Fig 3 continued on next page.**)

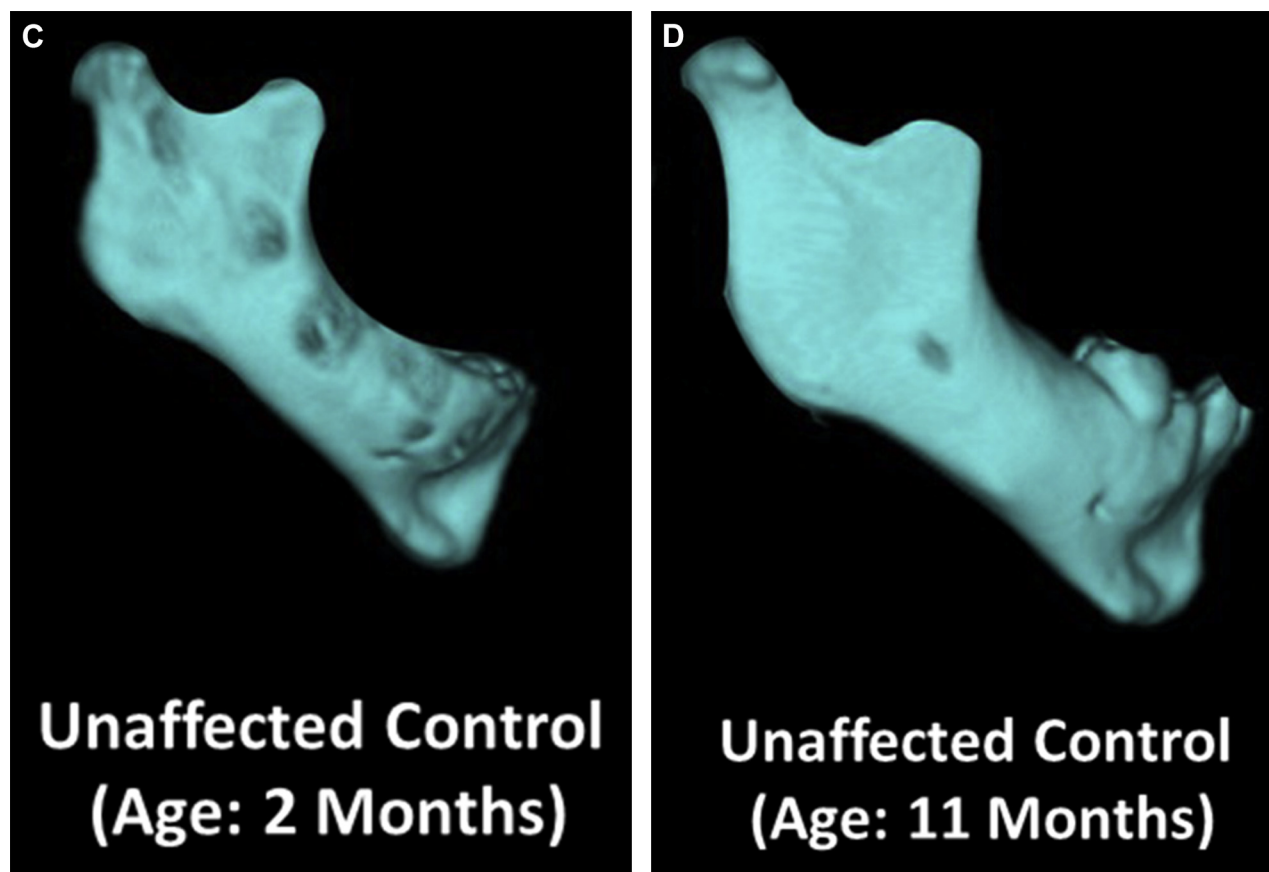
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explanation of the changes seen in this study is that distraction corrects mandibular morphology precisely because the site of the osteotomy and subsequent osteogenesis is where the mandible is deficient. This would explain the dramatic changes seen in the body and symphysis measurements, with minimal changes in the ramus and condyle measurements. DO not only improves the sagittal position of the mandible, as described previously, but also alters the shape of the mandible from elliptical to parabolic, as found in the present study, and ostensibly normalizes morphology.<sup>27</sup>

One explanation for these findings is related to the forces generated by the distraction process, which are transmitted anteriorly and posteriorly. The internal distraction devices used in this study were placed along the inferior border of the mandible and therefore had converging vectors (Fig 1). In theory this would apply a compressive anterior force at the symphysis and a divergent posterior force at the condyles. After initial activation, the hard stop of the ramus and condyle unit at the glenoid fossa could result in deflection of any additional force to the body and symphysis,

which contributes to the changes seen as a result of distraction. Although the sagittal component of the vector results in correction of the anteroposterior deficiency, the transverse component of the vector results in remodeling of the symphysis, forming a mandible that is more parabolic in shape, with a narrower intergonial angle and an orthogonal symphyseal angle. The posterior force toward the condyles had little or no effect on morphology distal to the gonion, with no change in intercondylar distance, intergonial distance, or ramus inclination. Of note, the vector used in these patients was a horizontal movement composed of sagittal and transverse components, without a vertical component. Although there are data that suggest that horizontal and vertical vector movements are equivalent in resultant airway expansion, the authors cannot comment on whether the observed changes would occur with a vertical vector.<sup>41,42</sup>

A second important finding in this study is the difference between the end-consolidation RS mandible and the age-matched RS control mandible that did not undergo MDO. A previously published comparison of pretreatment RS mandibles with age-matched controls



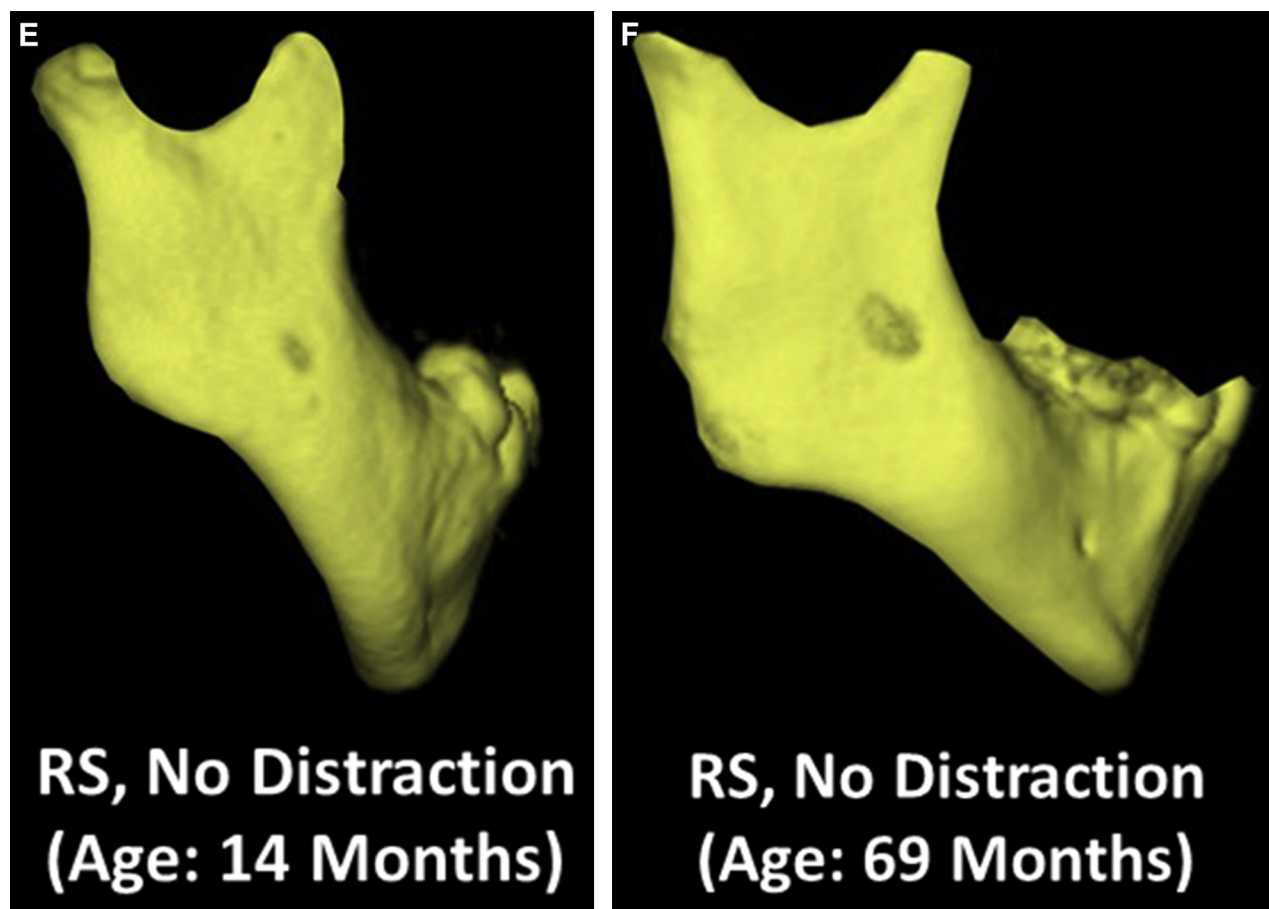
**FIGURE 3 (cont'd).** C, Control infant at 2 months of age. D, Control infant at 11 months of age. (Fig 3 continued on next page.)

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by the authors showed that the main morphologic differences occur anterior to the gonial angle.<sup>33</sup> In infants with mild to moderate RS phenotypes, nonsurgical maneuvers or minimally invasive therapies (eg, tongue-to-lip adhesion) could have a beneficial effect because they allow for stabilization of the airway until growth of the unaffected portion of the mandible (ramus and condyle unit) takes place.<sup>9,32,34,39</sup> Infants in the present study who underwent MDO according to the authors' institution's tongue-based airway obstruction protocol had severe airway obstruction that had not been alleviated with less invasive therapies (eg, nasopharyngeal airways, positioning, supplemental oxygen). In these more severe phenotypes, the mandibular bony deficiency might need to be addressed at a young age and distraction might be the best option for these patients because it normalizes mandibular shape.

As with any retrospective assessment, there are limitations to the design and analysis that merit consideration. First, these data are for a subset of infants with micrognathia who met the institutional criteria for treatment with MDO; these patients underwent MDO to treat severe airway obstruction, defined by respiratory failure with hypoxia and carbon dioxide

retention or severe obstructive sleep apnea. As such, the pre- and post-distraction measurements are representative of a more severe RS phenotype and might not be applicable to mild to moderate deformities that can be successfully treated with less invasive methods (eg, positioning, nasopharyngeal airways, tongue-to-lip adhesion, etc). Second, the lack of systematically collected clinical data (eg, serial polysomnography was not feasible in these clinical cohorts) precludes the authors' ability to correlate the morphologic changes to clinical airway findings *between* subgroups. Although there is no universally valid tool for assessment, the present analyses represent the authors' institutional experience with standardized CT scans. Multidisciplinary evaluation with assessments of feeding, growth, breathing, and sleep are needed not only to stratify patients into treatment groups but also to assess the effects of treatment.<sup>34</sup> Third, this is a limited time assessment, because most patients in all groups were younger than 1 year. The available data do not allow the authors to comment on the stability of morphologic changes, further changes with growth, alterations in inferior alveolar nerve function, temporomandibular joint dysfunction, or dental development compared with



**FIGURE 3 (cont'd).** E, Patient with RS without mandibular distraction osteogenesis at 14 months of age. F, Patient with RS without mandibular distraction osteogenesis at 69 months of age. RS, Robin sequence.

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the control groups. In this subgroup of patients with micrognathia related to RS, the authors can state that those treated as infants with MDO have an end-consolidation body-symphysis morphology that is markedly similar to that of a normal infant mandible.

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