Cervical Spine Deformity in Long-Standing, Untreated Congenital Muscular Torticollis

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Background: Congenital muscular torticollis (CMT) is a benign condition. With early diagnosis and appropriate management, it can be cured completely, leaving no residual deformity. However, long-standing, untreated CMT can lead to permanent craniofacial deformities and asymmetry.

Methods: Four adult patients presented to the author with long-standing, untreated CMT. Initial clinical assessment demonstrated tightness of the sternocleidomastoid muscle on the affected side. Investigation of cervical spine using 3-dimensional computed tomography scans with cervical segmentation allowed a 3-dimensional module to be separately created for each vertebra to detect any anatomical changes.

Results: A change in the axis of the vertebral column was noted when compared to that of the skull. Also, there were apparent anatomical changes affecting the vertebrae, which were most noticeable at the level of the atlas and axis vertebrae. These changes decreased gradually till reaching the seventh cervical vertebra, which appeared to be normal in all patients. The changes in the atlas vertebra were mostly due to its intimate relation with the skull base. The changes of the axis were the most significant, affecting mainly the superior articular facet, the lamina, and the body.

Conclusions: There were seemingly permanent changes along the cervical spine region in the adult patients with long-standing, untreated CMT in the form of bending and rotation deformities that might result in residual torticollis postoperatively.

Key Words: Cervical spine deformities, congenital muscular torticollis, vertebral segmentation

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Torticollis is an old Latin term that means "twisted neck." It is a symptom of various underlying diseases. Congenital muscular torticollis (CMT) is one of the most common causes of this condition. It is defined as a neck deformity that is detected at birth or immediately after birth due to a shortening of the sternocleidomastoid muscle (SCM).[1]

Congenital muscular torticollis is the third most common childhood congenital musculoskeletal anomaly, with an incidence ranging from 0.3% to 3.92%.[2,3] The male-to-female ratio for CMT is 3:2.[4] Despite many theories that have been proposed to explain such shortening, the actual reason is still unclear. However, consistent pathological findings are endomysial fibrosis and muscle fiber atrophy at the microscopic level.[5] Muscular torticollis is classified into 3 main groups according to the presence of a mass or tightness.[6]

Congenital muscular torticollis is rather a benign condition, and with early diagnosis and appropriate management, it can be cured completely without any residual deformity. However, long-standing, untreated CMT can lead to permanent cranial and facial deformities and asymmetry.[7] In addition to pain and a limitation of neck movement that might last forever,[8] typical facial deformities include frontal bone depression, zygomatic bone asymmetry, a posteriorly positioned ear on the affected side, and deviation of the chin to the other side. On the cranial level, positional plagiocephaly is frequently detected in patients with CMT.[9,10]

Although craniofacial deformities in CMT have been frequently studied from different perspectives, cervical spine changes in long-standing, untreated CMT have not been well studied. Therefore, our study has focused on exploring the changes that might occur to the cervical vertebrae of adult patients with known long-standing, untreated CMT.

MATERIALS AND METHODS

Four patients were diagnosed as having CMT with a clinically apparent, tight SCM on 1 side. The youngest patient was a 21-year-old female with right-sided CMT, the second youngest was a 21-year-old male with left-sided CMT, the third was a 24-year-old female with right-sided CMT, and the oldest patient was a 38-year-old male with left-sided CMT. None of those patients had received any type of treatment at any time before the time of presentation. The patients' chief complaint was the abnormal head tilting position rather than any functional or neurological complaints. Exclusion of other causes of torticollis was done to confirm the diagnosis of CMT. Facial examination revealed asymptomatic asymmetry with variable degrees of deformities among the patients for which the patients had not sought any medical advice, either before or at the time of presentation. There were no signs of any neurological deficits in any of the patients.

On presentation, 3-dimensional (3D) computed tomography (CT) scans of the skull and cervical spine were obtained for further investigation. The CT scans were obtained using the Siemens CT scanner (Siemens Medical Solutions, Erlangen, Germany; Somatom Sensation 64, matrix size 512 × 512, facial 1/1 mm pac). Segmentations of the cervical vertebrae were done using AnalyzeDirect AVW (Mayo Clinic, Rochester, MN), as shown in Figure 1. A separate 3D reconstructed module was done for each vertebra so that they could be studied separately. This 3D reconstruction enabled each vertebra to be translated and rotated freely. A midsagittal plane was established

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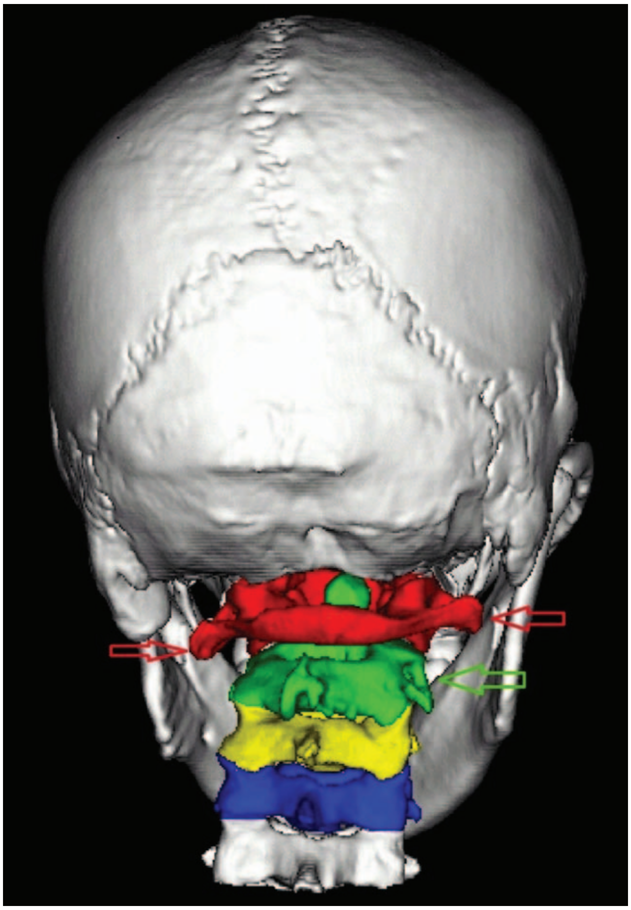

FIGURE 1. Posterior view of the skull and cervical spine showing segmentation of the upper 5 cervical vertebrae with growth changes, most apparent on the level of the atlas and axis: (red) atlas, (green) axis, (yellow) third vertebra, (blue) fourth vertebra, and (gray) fifth vertebra. The red arrow points to the level of the transverse process of the atlas vertebra, and the green arrow points toward the rotational deformity of the axis vertebra.

across each vertebra to assess any rotation deformity. For the atlas vertebra, the midsagittal plane was created across the anterior and posterior tubercle, while for the axis vertebra, it was created across the center of the odontoid process and passed through both the body and the tip of the spinous process (Fig. 2). For the remaining vertebrae, it was extended through the center of the anterior aspect of the body to the tip of the spinous process. Each atlas and axis vertebra was divided according to these midsagittal plans into 2 halves. To avoid any bias or error, adjustment of the position of both vertebrae was done before division so that they would be vertically and horizontally aligned. For each vertebra, the volumes of both sides were measured to detect any significant changes. The volume measurement was done using the volume render measurement tool in the AnalyzeDirect AVW software. Both sides were initially segmented and then subsequently divided and measured at the same threshold to avoid any bias due to different thresholds.

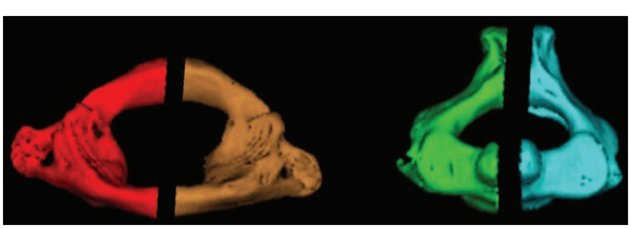

FIGURE 2. Top view of atlas and axis vertebrae with the midsagittal plane; for the atlas vertebra, it passes through the anterior and posterior tubercle, and for the axis vertebra, it passes through the center of odontoid process, the body, and the tip of spinous process.

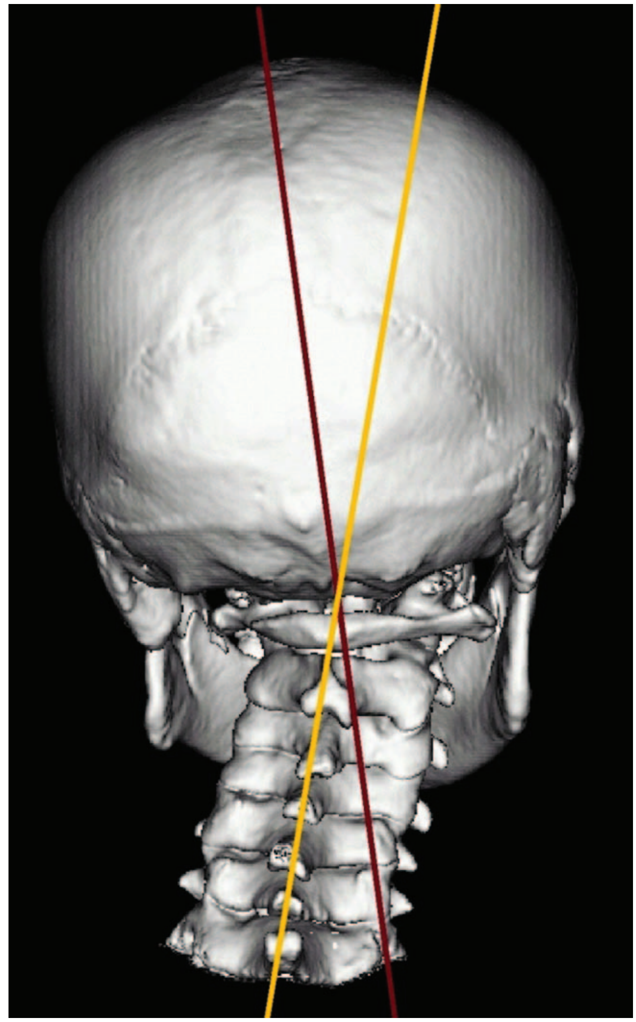

FIGURE 3. Posterior view of the skull and cervical spine; the red line represents the axis of the skull, while the yellow line represents the axis of the cervical vertebrae.

RESULTS

The overall view of the cervical spine showed changes that resembled those of congenital scoliosis due to failure of separation or unsegmented bar. There was a change in the axis of the cervical spine in comparison to that of the skull in the neutral position, as shown in Figure 3. Alteration of the spinous processes' shape and direction from C2 to C6 was also apparent. There was change in the vertical height of the vertebral bodies. The greatest difference was noticed at the level of the atlas and decreased gradually toward the seventh vertebra, which had equal vertical height. There was an apparent difference regarding volume measurements across both sides of the atlas and axis (Table 1).

TABLE 1. Demographic Distribution of the Patients With Volume Measurement Across Both Halves of Atlas and Axis

Age (y)	Sex	Atlas Volume (mm^3)		Axis Volume (mm^3)	
		Affected Side	Nonaffected Side	Affected Side	Nonaffected Side
21	F	5022	4943	6059	6112
21	M	7947	7706	9207	8689
24	F	7609	7481	8756	7858
38	M	6357	5279	7312	7846>

F, female; M, male; y, years.

The atlas vertebra showed significant bending deformity that matched the skull base shape (Fig. 4). There were significant changes in the direction and level of transverse processes, which were more severe in the older patient. There was also mild rotational deformity in comparison to the axis.

The axis vertebra showed the most significant deformity. Almost all of its components exhibited change. The odontoid process showed apparent tilting, and there were significant changes in the shape, slope, and size of the superior articular facet. The facet on the affected side showed a larger surface area and more sloping. Also, the lamina showed changes in its direction and curvature, since it appeared convex on the unaffected side and concave on other side (Figs. 5–7).

As we proceed downwards, the vertebrae started to show less significant changes until the seventh vertebra, which showed normal morphology without any apparent deformities (Fig. 8). These cervical spine deformities resulted in residual postoperative torticollis despite adequate surgical myomectomy and improvement of head tilting.

Regarding the skull shape, there was apparent mandibular asymmetry with chin deviation toward the affected side, as well as mild dental occlusal tilting. The maxilla and zygomatic bone on the affected side looked retruded and shorter than the other side. There was alteration in the shape and vertical height of the orbit when compared to the unaffected side (Fig. 9). There was posterior plagiocephaly that always affected the ipsilateral side, and mild frontal depression was also noticed in all patients. These facial findings matched the skull base changes that occurred secondary to the atlanto-occipital joint changes.

DISCUSSION

The cervical spine supports the cranium and enables its 3D movement in space. Although head movement is generated through the action of several muscles, the type of movements primarily depends on the structural integrity of the cervical vertebrae.[11] From an anatomical and physiological point of view, the cervical spine can be divided into 4 main subunits: the atlas, the axis, the C2–C3 junction, and typical cervical vertebrae C3–C7.[11] Growth of the vertebral body may be affected by genetic, racial, postural, or occupational factors. Generally, the rate of growth of the vertebral bodies appears to be more in the vertical direction than in the anteroposterior direction.[12]

FIGURE 4. Back view of the atlas vertebra, with the midsagittal plane showing apparent changes in the level and the direction of the transverse processes; it is directed downwards on the affected side.

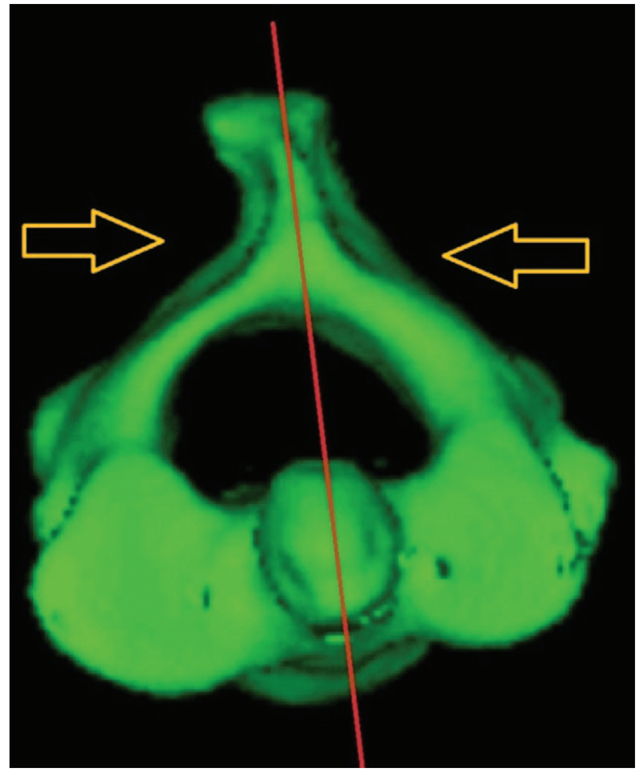

FIGURE 5. Top view of the axis vertebra, with the midsagittal plane showing changes in the morphology of the lamina on both sides (arrows), changes in the direction of the spinous processes, and changes in the shape and surface area of the superior articular facet.

The atlas vertebra is the first unique structure in the cervical spine. It is formed of 3 parts: the anterior, the posterior arches, and the lateral masses. Unlike other vertebrae, the atlas does not have either a body or a spinous process. It receives occipital condyles into its superior articular sockets to form the atlanto-occipital joint, which allows only for the nodding movement. This kind of limited movement causes the head and atlas vertebra to move as a single unit.[13,14] The strong atlanto-occipital joint and its limited mobility might explain the bending deformity at the level of the atlas vertebra in the patients with CMT in this study, as it matches the shape of their tilted skull base. Since the movements of the atlas vertebra are mainly passive and mediated through the muscles that act on the head (eg, SCM and splenius capitis), the movements are transmitted through the atlanto-occipital joints to the atlas.[15] Therefore, in the patients with CMT in this study, a rotational deformity was noted that matched the head rotation toward the affected side. This finding is in line with the results of Ozer et al, who observed a significant rotational deformity of the atlas vertebra along the axis vertebra in their study on major craniofacial deformities along the lower facial portion.[16]

The axis is the second unique structure, due to the presence of the odontoid process and having a very specialized superior articular facet that permits the head to rotate around a strong odontoid process (the median atlantoaxial joint). In this study, the changes that occurred in the superior articular facet might have been due to

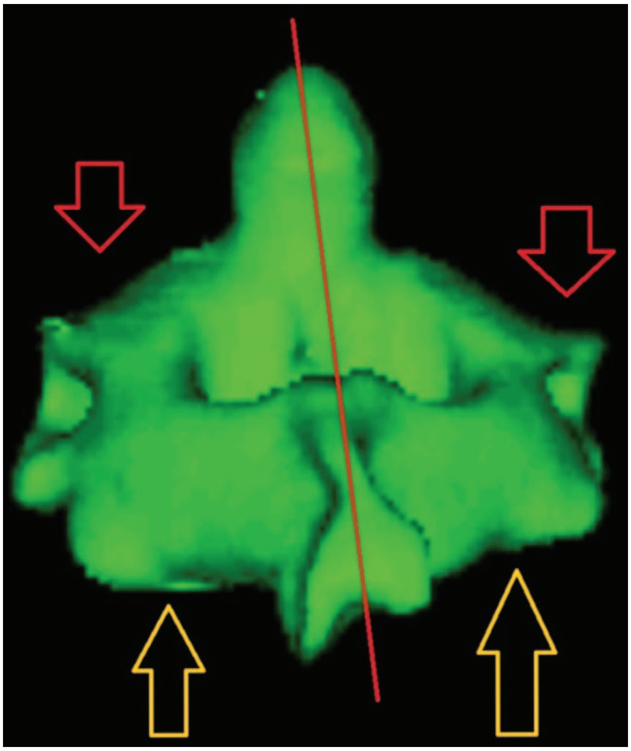

FIGURE 6. Posterior view of the axis vertebra, with the midsagittal plane showing changes in the sloping of the superior articular facet (top arrows) and changes in the vertical height of the lamina (bottom arrows).

FIGURE 8. Anterior view of the axis vertebra (top), with the third and fourth vertebrae showing sliding of the axis vertebra to the opposite side of the tilting along the C2–C3 junction, as well as decreasing deformities along the third and fourth vertebrae.

the unequal weight force transduction across the lateral atlantoaxial joint, since the weight is mainly transmitted through the lateral atlantoaxial joints.[17] This was reflected anatomically in the changes in the shape and sloping of the facets. Also, restraint to the movement of the atlas along the medial and lateral atlantoaxial joints is not typical movement, due to the absence of ligaments that are able to limit the flexion and extension movements. During the axial rotation, the alar ligament and the capsule of lateral atlantoaxial joint can limit such movement.[18] All these factors together might make the axis liable for rotational deformity and account for the

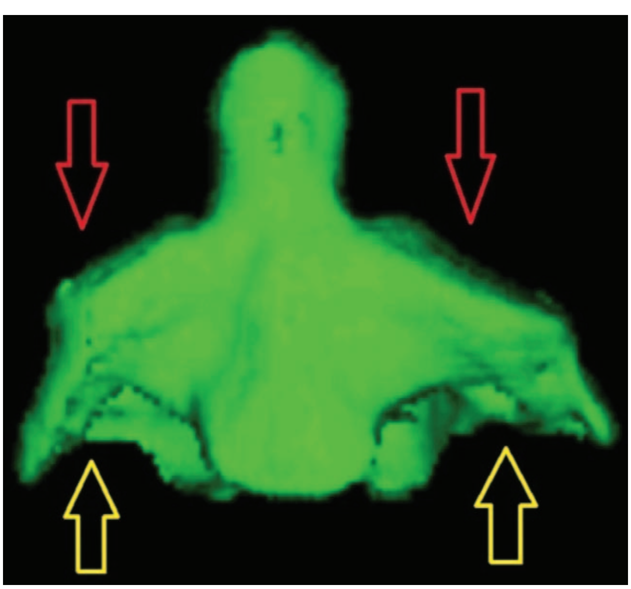

FIGURE 7. Anterior view of the axis vertebra, showing changes in the sloping of the superior articular facet (top arrows) and changes of the vertical height on either side of the body of the vertebra (bottom arrows).

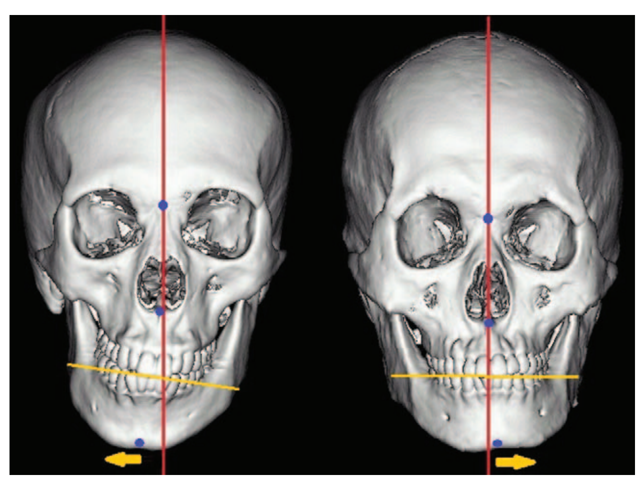

FIGURE 9. Anterior view of the skull of the 21-year-old female patient with right-sided CMT (left-hand picture) and the skull of the 38-year-old male patient with left-sided CMT (right-hand picture); in both patients the red line represent the midline and shows apparent asymmetry across both sides of the face; the yellow arrow shows the direction of the torticollis and also the chin deviation, while the blue dots represent the nation, anterior nasal spine, and the mentum, respectively. CMT, congenital muscular torticollis.

changes across the lamina and the shape of its body compared to other vertebrae noted in this study. These results are in line with the findings of Chen and Ko in their study of 18 children older than 6 years. They reported a change of the superior articular facet and tilting of the odontoid process.[19] Also, our results agree with Oh and Nowacek, who reported radiographic abnormalities in the axis vertebra and lateral atlantoaxis joint during their study of the surgical release of congenital torticollis in adults.[20]

In addition, volume changes were noted across each side of the atlas and axis. These changes showed a regular pattern at the level of the atlas; that is, the affected side was always larger than the other side. However, they showed a different pattern on the level of the axis vertebra; that is, the volume of the affected side appeared larger in 2 patients and smaller in the other 2 patients. These differences across the patients' axis vertebrae might be due to a compensatory rotational deformity of the body of the vertebra itself rather than the laminae and processes, which may be related to the severity of the original pathology. However, a study on a larger scale is recommended to clarify the relationship between the vertebral deformities and volume changes on the affected side.

The C2–C3 junction is another structure that is anatomically and functionally different cervical vertebra. Unlike the typical zygapophysial joints, the superior articular facets of the third vertebra and the inferior articular facets of the axis vertebra have different architectures regarding shape and position compared with other typical vertebrae. It is also operated using a different mechanism; during axial rotation of the neck, the atlas moves away from the direction of the movement,[21] which results in changing the direction of weight transduction across the C2–C3 junction.

Regarding the typical vertebrae (the third to seventh cervical vertebrae), the cervical intervertebral joints are saddle shaped since the opposing surfaces of bodies are slightly curved in the anteroposterior plane and also slightly from side to side.[11] The planes of both intervertebral joints and zygapophysial joints are different to allow the vertebrae to move freely in the sagittal plane across the transverse processes. However, during bending movement, they move across an axis perpendicular to the superior articular facets. These free rocking movements might also play a role in weight force transduction across the vertebrae. This might also be responsible for decreasing the severity of deformities down toward C7 in this study. Since the intervertebral disc does not share or resist pure loading forces during normal physiological conditions, it plays an important role only in severe compressing forces.[22]

According to Chen and Ko, the scoliosis detected in CMT is supposed to be secondary scoliosis since muscle release leads to its correction.[19] However, in this study the authors hypothesized that residual torticollis might be still present despite adequate treatment of the muscle due to the deformity of the spine itself. Since these deformities can occur in long-standing cases, attention must be directed toward the age at which these deformities start to develop to enable appropriate timing of surgical interventions and/or physical therapy.

Despite the fact that craniofacial asymmetry was not our primary point of investigation in this study, our results matched the findings of Jeong et al and Yu et al, who investigated craniofacial asymmetry in adult patients with neglected CMT.[7,10]

CONCLUSIONS

There are definitive vertebral deformities along the cervical spine in long-standing, untreated CMT. The bending and rotation deformities were more clinically significant at the level of the atlas and axis vertebrae and decreased gradually toward the seventh vertebra. The results of our current study highlight the importance of early surgical intervention and management of CMT to avoid the development of these apparently permanent spine deformities. Our findings also point to the need for future exploration of such changes in younger population groups to determine the age at which these changes generally begin.

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