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Defining Hyoplasia of the Atlas: A Cadaveric Study

Michael P. Kelly, MD¹, Yasushi Oshima, MD, PhD², Jin S. Yeom, MD, PhD³, Rashmi Agarwal, MD⁴, Navkirat S. Bajwa, MD⁵, and K. Daniel Riew, MD¹

¹Washington University School of Medicine, Department of Orthopedic Surgery, Saint Louis, MO

²University of Tokyo, Department of Orthopaedic Surgery, Tokyo, Japan

³Department of Orthopaedic Surgery, Seoul National University, Seoul, Republic of Korea

⁴Department of Orthopaedic Surgery, University of Pittsburgh, Pittsburgh, PA

⁵University of South Alabama, College of Medicine, Mobile, AL

Abstract

Study Design—Cadaveric Study

Objective—To define congenital hypoplasia of the atlas.

Summary of Background Data—Little has been written about hypoplasia of the atlas and it is usually described in the setting of other skeletal dysplasias or syndromes.

Methods—543 Cervical spine specimens were randomly selected from the Hamann Todd Collection. Sagittal and coronal diameters of the atlas, axis, and C3 (when available) and the dens diameter were measured using digital calipers. Correction for modern size and radiographic magnification was performed. Hypoplasia of the atlas was defined as the lowest 2.5% of measurements. The correlation between inner sagittal diameters at C1 and C3 was calculated.

Results—The mean C1 inner sagittal diameter was 30.8 ± 2.4 mm (Range 23.5 – 38.1mm). We defined C1 hypoplasia as an inner sagittal diameter value representing the smallest 2.5% of subjects. Since the mean was 30.8mm, hypoplasia was defined as a diameter ≤ 26.1 mm. Correcting for size and magnification of radiographs, hypoplasia is defined as an inner sagittal diameter of the atlas of 28.9 mm. Approximately 10% of cases had a dens that occupied more than 40% of the spinal canal at C1, thus not following Steel's Rule of Thirds. There was only a moderate correlation between the spinal canal diameter at C1 and at C3 ($r=0.483$, $N=345$, $P<0.001$).

Conclusions—With an inner sagittal diameter of 26mm or less, one may describe the atlas as hypoplastic. 10% of the specimens had an odontoid process that occupied more than 40% of the spinal canal at C1. There was little correlation between the inner sagittal diameter at C1 and the diameter at C3.

Keywords

cervical stenosis; Congenital Stenosis; Hypoplasia; myelopathy; atlas

Introduction

Subaxial cervical spinal stenosis is a common condition which both puts patients at risk for developing cervical spondylotic myelopathy and increases the risk of spinal cord injury with trauma.^{1–4} Stenosis at the level of the atlas is far less common, due in part to the particular form and function of the atlantoaxial articulation. Steel described the “rule of thirds”, where the odontoid process, spinal cord, and space each occupy one third of the canal diameter at the atlas.⁵ Thus, the spinal canal at the level of the atlas is often tolerant of instability and trauma, as is evidenced by the low rate of spinal cord injury in the setting of odontoid process fractures.

Hypoplasia of the atlas is a rare entity that may cause spinal stenosis and myelopathy. The literature describing this pathology consists mostly of case reports or small case series.^{6–16} Many reported cases are associated with other developmental abnormalities, such as spondyloepiphyseal dysplasia congenita or Down Syndrome. Also, other developmental abnormalities, such as failure of fusion of C1, are commonly coexistent making pure, pathologic hypoplasia less common. There have been several studies examining anatomy at the level of the atlas and axis.^{17,18} These studies, however, have been underpowered, performed on radiographs, or both. No adequately powered study exists to define hypoplasia of the atlas.

We sought to examine and measure an appropriate number of cadaveric cervical spine specimens, to define the normal diameter of the atlas, and to propose a value, defining hypoplasia as the lowest 2.5% of measurements, which may form the basis for future clinical research. We also sought to determine whether subaxial canal diameters correlate to supra-axial canal diameters and to evaluate the frequency with which the “rule of thirds” does not apply.

Materials and Methods

Five hundred forty-three adult cervical spines from Caucasians and African Americans were randomly selected from the Hamann Todd Collection—a collection of over 3000 skeleton remains from the late nineteenth and early twentieth centuries from Cleveland, Ohio. This sample size is appropriate for a population larger than 264 million, giving the 95% confidence interval with a 5% margin of error.¹⁹ Digital calipers (Mitutoyo, Kawasaki, Japan) were used to perform all measurements. The inner sagittal diameter of C1 was measured from the posteriormost portion of the fovea dentis to the posterior most portion of the ventral lamina. Inner sagittal diameters were measured at C2 and C3 by calculating the distance from the posteriormost portion of the vertebral body to the posteriormost portion of the ventral lamina. Inner coronal diameters were measured at C1, C2, and C3 by calculating the distance between lateralmost portions of the lateral mass-lamina junctions at C1 and the distance between the lateralmost portions of the pedicle-lamina junctions at C2 and C3. The

diameter of the dens was measured at the widest point. Measured values were recorded, via the calipers, directly into a computer database, without transcription.

The mean values of the measurements were calculated, along with standard deviations. Histograms were plotted to ensure a normal distribution. Congenital hypoplasia was defined as values falling below two standard deviations from the mean. Between group comparisons were made using unpaired two-sample t tests, organized both by gender and by race. A Pearson correlation coefficient was calculated between the anterior to posterior diameter at C1 and C3 (supra-axial and subaxial). Statistical significance was defined as $p < 0.05$.

The Hamann Todd collection consists of skeletons from the turn of the 20th century. At this time, the average height was smaller than current times. Males have grown from an average height of 172.1cm to 175.3cm (+1.8%). Females have grown from an average height of 161.3cm to 162.5cm (+0.8%) To account for the change in the increase in height of males and females since the Hamann Todd Collection was created, the measured diameters for males were increased 1.8% and females increased 0.8%.⁴ Lee et al have previously shown that radiographic measurements 9.9% larger than the physically measured values of canal diameter with specimens from this collection.⁴ This value was obtained by comparing 45 radiographic measurements with the corresponding direct measurement. To account for radiographic magnification, measurements were increased 9.9% when reporting values as would be obtained from lateral radiographs. All statistical calculations were performed using SPSS v20.0 (IBM, Armonk, New York).

Results

Demographics

Five hundred forty-three adult cervical spine specimens were randomly selected from the Hamann Todd Collection. There were 283 males and 260 females. Two hundred ninety four specimens came from Caucasians and 249 from African-Americans. The mean age of the specimens was 47.0 ± 16.9 years (Male: 48.7 ± 14.8 , Female: 45.2 ± 18.7 , $p = 0.034$). From these specimens, 541 had complete C1 and C2 rings and were included in the statistical analysis (Table 1). Three hundred and forty three specimens had C1 and C3 measurements available for correlations between the diameter at the atlas and the first subaxial level.

C1 Measurements

The mean C1 sagittal diameter was 30.8 ± 2.4 mm (Range 23.5 – 38.1mm). Males had larger C1 sagittal diameter than females (Male 31.8 ± 2.1 mm vs Female 29.8 ± 2.2 mm, $p < 0.001$). Caucasian specimens were slightly larger versus African American specimens (Caucasians: 31.3 ± 2.3 mm vs African Americans 30.3 ± 2.2 mm, $p < 0.001$).

Definition of C1 Hypoplasia

We defined C1 hypoplasia as an inner sagittal diameter values falling in the lowest 2.5% of measurements (1.96 standard deviations below the mean). Since the mean was 30.8mm, hypoplasia was defined as a diameter ≤ 26.1 mm (Figure 1).

C2 Measurements

The mean C2 sagittal diameter was $18.8 \pm 2.2\text{mm}$ (Range 12.8 – 25.7mm). As expected, males had larger C2 sagittal diameter than females (Male $19.5 \pm 2.3\text{mm}$ vs Female $18.1 \pm 1.8\text{mm}$, $p < 0.001$). As for the C1 parameters, Caucasian specimens were slightly larger than African American specimens (Caucasians: $19.4 \pm 2.2\text{mm}$ vs African Americans $18.2 \pm 2.0\text{mm}$ $p < 0.001$).

Dens Measurements

The mean Dens Diameter was $11.1 \pm 1.0\text{mm}$ (Range 8.0 – 14.2mm). Males had larger Dens Diameters than females (Male $11.7 \pm 0.8\text{mm}$ vs Female $10.4 \pm 0.9\text{mm}$, $p < 0.001$). Caucasian specimens were slightly larger than African American specimens (Caucasians: $11.4 \pm 1.0\text{mm}$ vs African Americans $10.7 \pm 0.9\text{mm}$, $p < 0.001$).

Relative Measures

The dens anterior to posterior diameter occupied 36% of the C1 canal diameter, on average. We found that approximately 10% of cases had a dens that occupied more than 40% of the spinal canal at C1, thus not following Steel's Rule of Thirds. There was only a moderate correlation between the spinal canal diameter at C1 and at C3 (Figure 2; $r = 0.497$, $N = 340$, $P < 0.001$).

Correction for Modern Body Size

The mean C1 sagittal diameter, correcting for modern body size, was $34.3 \pm 2.7\text{mm}$ (Range 26.0–42.6mm). The mean sagittal diameter at C2 was $21.0 \pm 2.5\text{mm}$ (Range 14.2–28.8mm) and the mean dens diameter was $12.3 \pm 1.2\text{mm}$ (Range 8.8–15.9mm). The mean C1 sagittal diameter in males was $35.5 \pm 2.3\text{mm}$ and in females 33.0 ± 2.4 . We have defined C1 hypoplasia as measurements equal to 26.1mm or less. As clinicians will be evaluating patients based upon lateral radiographs, this value, corrected for modern size and radiographic magnification is 28.9mm (Figure 3).

Discussion

While there are a number of case reports describing hypoplasia of the atlas causing myelopathy, to our knowledge, there are no adequately powered studies that have examined the range of normal values for canal measurements at the level of the atlas.^{4,20} As cervical spondylotic myelopathy is a common complaint in the spine surgeon's clinic, they must be aware that hypoplasia of the atlas may cause spinal cord compression, with symptoms similar to subaxial conditions. This is particularly important as many routine magnetic resonance imaging (MRI) scans begin with axial images at the level of the axis, with no imaging of the canal at the level of the atlas and the odontoid process. Thus, without a threshold for concern, there may be misdiagnosis and a delay to prompt management. The purpose of this study was to define a value for C1 hypoplasia using population norms from an appropriate sized cohort of specimens.

The mean C1 anterior-posterior diameter in this cohort was 30.8mm. We defined C1 hypoplasia as values falling nearly two standard deviations below the mean, which

represents the lowest 2.5% of the population. A direct measurement of 26.1mm may be defined as hypoplastic. When corrected for the larger size of contemporary subjects and for the magnification of radiographs, this was approximately 29mm.⁴ We found that 10% of the samples did not follow the “Rule of Thirds.” That is, 10% of cases had a dens diameter that occupied more than 40% of the spinal canal at C1, thus limiting the space available for the cord, particularly when soft tissues are accounted for. Also important to note is that we found only a weak to moderate correlation between supra-axial canal diameters and subaxial canal diameters. This should alert physicians to evaluate C1 when managing a patient with evidence of cervical myelopathy, as C1 stenosis may exist in the absence of subaxial stenosis.

The vast majority of previous publications on this subject are case reports and reviews of the literature.^{6–16,18,20–23} Musha and Mizutani measured the sagittal inside diameter (equivalent to our C1 anterior-posterior diameter) of 300 adult Japanese men and women.¹⁸ They also defined hypoplasia as the lowest 2.5% of values. In women, 29.4mm was considered hypoplastic, while 30.5mm was hypoplastic in men. These estimates correlate well with our value of 29mm. Their subjects were all Asians, though, and thus not generalizable to a Caucasian and African American population. Doherty and Heggeness measured the anterior-posterior diameter of 88 C1 vertebral specimens.²⁰ The mean value was 32mm and the lower 2.5% 28mm, higher than our calculated value. Their sample size is too small for an estimate of the population as a whole and this value may overestimate what should be considered hypoplasia. None of these studies reported the canal diameter at subaxial levels. We have shown that the inner sagittal diameter of C1 was only moderately correlated to the inner sagittal diameter of C3. The implication of this finding is that one may fail to diagnosis stenosis at C1, if one assumes that the lack of subaxial congenital stenosis implies a normal canal above. Also, none of the aforementioned studies examined the C1 space available for the cord (SAC) in relation to the dens and the C1 inner sagittal diameter. In 10% of our patients Steel’s “Rule of Thirds” was violated, leaving a smaller SAC than usually considered.

A weakness in this study is that we could not account for soft tissues or atlanto-axial instability. Subtle instability, pannus formation, and an ossified transverse ligament have all been described in the setting of atlas stenosis causing myelopathy. Thus, patients with a C1AP greater than 26mm may develop spinal cord compression, as the effective SAC will be smaller, due to instability or ligament hypertrophy. The size of the spinal cord has little correlation with body size or age, so cord volume is likely similar regardless of bony measurements.¹⁷ Also, we do not have clinical histories or examinations, so many patients may have had asymptomatic stenosis, a finding common in the subaxial cervical spine. Despite these shortcomings, we were able to find a potentially critical value for the anterior-posterior diameter of the atlas. This value needs to be validated in a cohort of patients, with histories, exams, and MRI available.

In the largest series of Caucasian and African American skeletons, we have defined hypoplasia of the atlas as an inner sagittal diameter of 26mm or less. This value is smaller than previously reported estimates. A subsequent clinical study, validating or refuting our definition of C1 hypoplasia is necessary as in many cases without C1–C2 instability, a

smaller C1 sagittal diameter may be tolerated. However, in cases of even subtle instability with hypoplasia, the SAC may narrow even further causing myelopathy. Furthermore, 10% of the specimens had an odontoid process that occupied more than 40% of the spinal canal at C1 indicating that these canals may be less tolerant of subtle instability, narrowing the SAC at the level of the atlas. Clinicians should be aware that hypoplasia of the atlas may be a cause of spinal stenosis in the absence of subaxial stenosis and MRI should include the foramen magnum and atlas, particularly if the C1 ring measures smaller than 29mm on a lateral radiograph.

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1. Congenital hypoplasia of the atlas may be defined as an inner sagittal diameter of C1 of 26mm or less.
2. Ten percent of patients did not follow Steel's "Rule of Thirds," with an odontoid process occupying more of the spinal canal at the atlas than expected.
3. Hypoplasia of the atlas is a potential cause of cervical myelopathy.

Figure 1.
Histogram plot of C1 sagittal diameter measurements with normal distribution curve.
Hypoplasia was defined as the lowest 2.5% (C1 Sagittal diameter $\leq$ 26.2mm).

Figure 2.
Correlation plot between C1 Sagittal Diameter and C3 Sagittal Diameter

Figure 3.
Upright lateral radiograph of a 72 year old gentleman with hypoplasia of C1 and diffuse idiopathic skeletal hyperostosis (Sagittal diameter 28.6mm) causing cervical myelopathy.

Table 1

Sagittal and Coronal Measurements at Atlas and Axis

	Canal Diameter* (mm)				
	C1 Sagittal	C1 Coronal	C2 Sagittal	C2 Coronal	Dens
All specimens (n=543)	30.8 ± 2.4	28.2 ± 2.5	18.8 ± 2.2	23.1 ± 1.7	11.1 ± 1.0
Male (n=283)	31.8 ± 2.1	29.0 ± 2.2	19.5 ± 2.3	23.5 ± 1.7	11.7 ± 0.8
Female (n=260)	29.8 ± 2.2	27.4 ± 2.5	18.1 ± 1.8	22.7 ± 1.7	10.4 ± 0.9
Caucasian (n=294)	31.3 ± 2.3	28.9 ± 2.3	19.4 ± 2.2	23.3 ± 1.7	11.4 ± 1.0
African American (n=249)	30.3 ± 2.2	27.4 ± 2.5	18.2 ± 2.0	22.9 ± 1.7	10.7 ± 0.9
Male compared with female	<0.001	<0.001	<0.001	<0.001	<0.001
Caucasian compared with African American	<0.001	<0.001	<0.001	<0.001	<0.001

* Values Reported as Mean and Standard Deviation