Morphometry of the outlet of the foramen magnum in crania with atlantooccipital fusion

Laboratory investigation

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Object. Assimilation of the atlas to the occiput may result in symptoms that are often compressive in nature around the outlet of the foramen magnum. The aim of the present study was to elucidate the morphological features of the bone through this foramen.

Methods. Thirteen adult skulls with atlantooccipital fusion underwent morphometrical analysis of the outlet of the foramen magnum.

Results. All specimens but one were found to have a decreased area of the outlet of the foramen magnum. In those 12 specimens, a decrease of 15%–35% was seen. Fusions of the atlas that were based primarily along the anterior rim of the foramen magnum resulted in more obstruction of its outlet. In general, the horizontal diameters of the outlet of these foramina were more decreased from the normal range.

Conclusions. These findings demonstrate that in the majority of cases, assimilation of the atlas to the occiput results in a compromised outlet of the foramen magnum. (DOI: 10.3171/2011.3.SPINE10828)

Key Words • anatomy • skull base • neurosurgery • foramen magnum

First described in 1844 by Rokitansky, whose findings were discussed by Hadley,9 occipitalization of the atlas occurs in approximately 0.08%–3% of the population, most commonly between the anterior arch of the atlas and the anterior foramen magnum.12,15 Also known as atlantooccipital assimilation, the varying degrees of this anomaly, which is the most commonly recognized bony variation of the craniovertebral junction, produce a wide range of effects, which are usually asymptomatic, but may result in neurological compromise.9,15,21,25,31 A determination of the clinical outcome of this state depends on the particular anatomical findings. In some cases, the foramen magnum can be constricted, with symptoms exacerbated by head movement,26,29 resulting in the so-called foramen magnum syndrome.15 Often these patients have compensatory hypermobility of the atlantoaxial joint, and may have compression of the vertebral artery with resultant ischemic symp-toms.1,19,22,23,26,31,34 Atlantooccipital fusion is also commonly associated with the presence of a posteriorly constricting band of dura mater that may groove the spinal cord.26

Although it is usually asymptomatic, occipitalization of the atlas can have mild to detrimental signs and symp-toms.14,21,27,32,38 Medullary compression2 and death have been attributed to this bony anomaly.35 Various studies report a wide variety of neurological symptoms for different patients. These include limited range of motion in the neck due to pain, motor deficit in the upper limbs, difficulty walking, spasticity, and numbness. Rotation of the neck or extension of the jaw may exaggerate symptoms.18 For example, Iwata et al.12 reported on a patient with occipitalization of the atlas who developed hypalgesia and hypesthesia of both hands and the right foot with maximal mouth opening and neck extension.

This article contains some figures that are displayed in color online but in black and white in the print edition.
The condition can result from one of two different origins.2,3,14,15 The atlas may be melded to the occipital bone due to a local inflammation caused by infection or tumor, or it may be congenital.15 Bézi2 proposed that “the term ‘assimilation’ be reserved for congenital cases and those of an inflammatory origin should be called ‘synostosis’.” With congenital forms, this fusion is the result of a failure of segmentation between the skull and the atlas.2,5–7,9,15,26,29,31 However, regardless of the origin of the fused atlantooccipital joint, the outcome can produce the same compression of nervous and vascular tissues.

Methods

Thirteen dry skulls exhibiting atlantooccipital fusion from 6 female and 7 male adult skeletons were examined. These were derived from commercial sources of skeletons, and were supplied to our laboratories over several decades and by various sources. Other than the single fusion anomaly, none of the dry skulls revealed any evidence of gross pathological entities, previous surgical procedures, or traumatic lesions. Based on overall bone fusion and degradation, the age of the specimens was approximated (using standard methods) to be between 40 and 75 years at death.

The foramen magnum in each specimen was viewed internally and externally (internal and external to the level of the hypoglossal canal, respectively), recorded with a digital camera (Nikon Coolpix S5), and studied using a computer-assisted image analysis system, with Lucia software version 5.0 (2000, edition for Windows XP); the imaging analysis system was made by Nikon (Laboratory Imaging, Ltd.). The digital camera was connected to an image processor (Nvidia GeForce 6800 GT) linked to a mainframe computer. Digitized images of the foramina were stored in the Lucia program (2048 × 1536 pixels). After applying a standard 1-mm scale to all images, the program was able to calculate pixel differences between 2 selected points, such as the width of a foramen, as previously described.33 This allowed easy and accurate translation of pixel differences into metric measurements. These measurements included the AP and horizontal diameters and surface area of the inner aspect of the foramen magnum, and the outlet of this foramen where the assimilation was found. Statistical analysis between male and female skulls was performed using Statistica for Windows, with significance set at p < 0.05.

Results

Eight specimens (62%) were found to have primarily anterior fusion, 1 (7.7%) primarily posterior fusion, 3 (23%) posterolateral fusion, and 1 (7.7%) complete fusion. Of these, 12 fusions (92.3%) resulted in less surface area at the outlet of the foramen magnum compared with a normal range.33 The inner aspect of the foramen had an area of 405–702 mm² (mean 603 mm²), and the outlet of the foramen had an average area of 410 mm² (range 280–503 mm²) (Figs. 1 and 2). Fusions that were primarily anterior were more likely to result in constriction of the outlet of the foramen magnum (Fig. 3). The total decrease in surface area of all specimens was between 15% and 35%. The inner AP diameter of the foramen magnum for all skulls ranged from 2.8 to 4 cm (mean 3.3 cm). The mean inner horizontal diameter for all foramina ranged from 2.6 to 3.6 cm (mean 3.1 cm). The mean outer AP and horizontal diameters were 2.5 and 2.3 cm, respectively. In all specimens, the narrowed outlet of the foramen magnum was due to the occipitalization and not to malformation of the skull base. The left styloid process...
Foramen magnum in atlantooccipital fusion

![Fig. 3. Photograph showing an example of the internal aspect of the foramen magnum outlet in a skull with atlantooccipital fusion that is primarily anterior in nature.](image)

...of one specimen was elongated, and in another specimen the posterior arch of C-1 was bifid. Statistical analysis revealed no difference in the type of fusion between sexes. Skulls were more likely to have a decrease in their horizontal versus AP outlet diameters (p < 0.05).

**Discussion**

In an earlier study, we performed a morphometric analysis of the foramen magnum in normal adult crania. For these, the mean surface area of the foramen magnum was 558 mm², with a range of 385–779 mm². From this group, 20.8% had a surface area < 500 mm², 66.6% had a surface area between 500 and 600 mm², and 12.5% had a surface area > 600 mm². The mean AP diameter of the foramen magnum was 3.1 cm, with a range of 2.5–3.7 cm. The mean horizontal diameter was 2.7 cm, with a range of 2.4–3.5 cm. In the present study, all but one specimen with atlantooccipital fusion was found to have a compromised outlet of the foramen magnum. This compromise was found to be most common in the horizontal diameter of the outlet of the foramen, and resulted in a 15%–35% decrease in the area of this region. Interestingly, the diameters of the inlets of all foramina were found to be within normal range.

The foramen magnum varies in shape in children and adults. Lang classified the shapes in 5 groups, as follows: 1) composed of 2 semicircles (adults 41.2% and children 18.4%); 2) an elongated circle (adults 22.4% and children 20.4%); 3) egg-shaped (adults 17.6% and children 25.5%); 4) rhomboidal (adults 11.8% and children 31.6%); and 5) rounded (adults 7% and children 4%). This author found that the average AP diameter of the foramen was 35 mm and the average horizontal diameter was 30 mm. It is generally accepted that when the normal diameter of the foramen magnum (35 mm) is reduced to approximately 19 mm, symptoms may manifest. For reference, the normal AP diameter of the atlas is 27–35 mm, and the average width of this space anterior to the facet joints is 31 mm.

Embryologically, the sclerotomes of the first 4 somites eventually fuse to form the occipital bone and the posterior part of the foramen magnum, and the first 2 sclerotomes form the basiocciput. The third sclerotome, as it forms the jugular tubercles, is responsible for development of the exoccipital centers. A key component in this process is the fourth sclerotome, also known as the proatlas. The proatlas contains 3 parts: the hypocentrum, forming the anterior tubercle of the clivus; the centrum, a precursor of the apical ligament and the apex of the dens; and the neural arch, which is further subdivided into ventral-rostral and dorsal-caudal components. The occipital condyles and the U-shaped anterior margin of the foramen magnum as well as the alar and cruciate ligaments arise from the ventral-rostral portion of this embryological part. The dorsal-caudal division forms the superior part of the posterior arch and the lateral masses of C-1. Consequently, anomalies and malformations of the occipital sclerotomes result in irregular geometry of the foramen magnum and related structures.

The foramen magnum is a fundamental component in the complex interaction of bony, ligamentous, and muscular structures composing the craniovertebral junction. The shape and size of this foramen are critical parameters for the manifestation of clinical signs and symptoms in craniovertebral pathological entities. Such signs and symptoms include motor myelopathy, sensory abnormalities, brainstem and lower cranial nerve dysfunctions, and signs and symptoms referable to vascular compromise. Diseases associated with anomalies of the foramen magnum include occipital vertebrae, basilar invagination, condylar hypoplasia, and atlas assimilation. Interestingly, one report found that the persistence of the sphenoccipital synchondrosis, aggravated by the coexistence of basilar invagination, resulted in stenosis at the foramen magnum.

Among developmental and acquired craniovertebral junction disorders, achondroplasia is the most commonly reported, with a frequently associated stenotic foramen magnum. The AP and horizontal diameters of the foramen magnum have been found to be independent risk factors in these patients, who often need craniovertebral decompression. Among developmental and acquired craniovertebral junction disorders, achondroplasia is the most commonly reported, with a frequently associated stenotic foramen magnum. A wide foramen magnum has also been appreciated in patients with diastrophic dysplasia. Other diseases associated with stenosis of the craniovertebral junction include craniofacial dysplasia, Jeune asphyxiating thoracic dystrophy, and spherophakia-brachymorphism (Marchesani syndrome). As a case example, a patient of ours with Chiari I malformation presented with a month-long history of left abducen nerve palsy and periods of dyspnea that lasted for several seconds. Imaging revealed occipitalization of the atlas, with significant ventral compression of the spinomedullary junction due to this osteological malformation (Fig. 4). Following posterior fossa decompression, the left abducen nerve palsy improved, and the patient no longer complained of difficulty breathing.
were relieved following posterior fossa decompression. Note the atlan-

tooccipital fusion and ventral compression.

Conclusions

Although atlantooccipital assimilation does not al-

ways produce detrimental effects, it is a condition

that must be well understood and consequently addressed

by clinicians. The most important task is determining

whether the abnormality might lead to the compression of

nerve tissue, which can cause a vast array of neurological

symptoms ranging from mild numbness to sudden death.

Our findings demonstrate that in the majority of cases of

atlantooccipital assimilation, the outlet of the foramen

magnum is compromised by 15%–35%.

Disclosure

The authors report no conflict of interest concerning the mate-
ials or methods used in this study or the findings specified in this paper.

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cript and approved it for submission: all authors. Study supervision: Tubbs.

References


4. Fenoy AJ, Menezes AH, Fenoy KA: Craniovertebral junction fusions in patients with hindbrain herniation and syringohy-


7. Gladstone RJ, Wakeley CPG: Variations of occipito-atlantal joint in relation to metemeric structure of craniovertebral re-


11. Holliday PO III, Davis C Jr, Angelo J: Multiple meningiomas of the cervical spinal cord associated with Klippel-Feil malfor-


14. List CF: Neurologic syndromes accompanying developmental anomalies of occipital bone, atlas and axis. Arch Neurol Psy-


chiatry 45:577–616, 1941

15. Martellacci S, Ben Salem D, Méjean N, Sautreaux JL, Krausé D: A case of foramen magnum syndrome caused by atlanto-


171–181, 1948


19. Menezes AH: Craniovertebral junction database analysis: in-


25. Peyton WT, Peterson HO: Congenital deformities in the re-


gion of the foramen magnum: basilar impression. Radiology 38:131–144, 1942


27. Sakai K, Tsutsui T: Bow hunter’s stroke associated with at-


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Schüller A: A diagnosis of “basilar impression.” Radiology 34:214–216, 1940
32. Symonds CP, Meadows SP: Compression of spinal cord in neighbourhood of the foramen magnum with a note on the surgical approach. Brain 60:52–84, 1937

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