Quantitative Measurements of the Skull Base and Craniovertebral Junction in Congenital Occipitalization of the Atlas: A Computed Tomography–Based Anatomic Study

Rui Zong, Yiheng Yin, Guangyu Qiao, Yazhou Jin, Xinguang Yu

■ OBJECTIVE: **To study the craniovertebral junction and determine the anatomic characteristics of occipitalization of the atlas (OC) by computed tomography (CT) imaging.**

■ METHODS: **We retrospectively reviewed 80 cases of patients with OC who underwent cervical CT scanning between March 2012 and March 2014. Forty healthy subjects were recruited as a control cohort. Fusion pattern and associated osseous anomalies were recorded. Fifteen linear quantitative parameters were measured to study the outlet of the foramen magnum, angular dimension of the skull base, posterior cranial fossa, and height of the odontoid process.**

■ RESULTS: **The most common fusion pattern was the complete osseous fusion (83.75%). Fifty-four patients (67.5%) presented with other osseous anomalies. Measurements of the OC group, such as the length of the clivus, cranial canal angle, and height of the odontoid process, were significantly different than those of the control group. The correlation analysis showed that the C1 lateral facet inclination was significantly correlated with the age of onset.**

■ CONCLUSIONS: **The outlet of the foramen magnum is severely impaired in patients with OC, and the presence of other osseous anomalies is common. Deformity is not confined to the region of the assimilated atlas; the clivus and odontoid process are also shorter than normal. The lateral facet inclination likely influences disease progression.**

INTRODUCTION

Congenital bony fusion of the atlas vertebra to the base of the occipital bone is referred to as assimilation of the atlas, also known as occipitalization of the atlas (OC), occipitocervical synostosis, atlanto-occipital fusion, and atlanto-occipital nonsegmentation/assimilation. OC is caused by a failure of segmentation between the fourth occipital sclerotome and the first cervical sclerotome.[1,2] It is the most common congenital malformation involving the craniovertebral junction (CVJ), with a global incidence of 0.25%–3.63%.[2-6]

Most patients with OC likely remain asymptomatic. For patients with atlantoaxial instability or basilar invagination, symptoms may occur because of compression of the spinal cord, tonsillar herniation, or syringomyelia. Occipitocervical fixation and fusion is usually recommended to treat patients with progressive symptomatic segmental instability or neurologic deficits. Therefore, a clear understanding of the anatomic features of the CVJ of OC is essential for clinicians to manage consequent complications of OC. However, the available information on the morphologic features of OC is insufficient and is mostly based on gross observations or case reports. This study was designed to quantitatively

Key words
- Anatomy
- Atlanto-occipital fusion
- Atlas assimilation
- Craniovertebral junction
- Foramen magnum
- Occipitalization

Abbreviations and Acronyms
AADI: Anterior atlantodental interval
BA: Basal angle
CCA: Clivus-canal angle
CIA: Coronal inferior C1 facet angle
CT: Computed tomography
CVJ: Craniovertebral junction
DCL: Distance from the tip of the odontoid to the Chamberlain line
FM: Foramen magnum
HB: Height of the C2 body
HO: Height of the odontoid process
OC: Occipitalization of the atlas
PADI: Posterior atlantodental interval
PCF: Posterior cranial fossa
SIA: Sagittal inferior C1 facet angle

Department of Neurosurgery, PLA General Hospital, Beijing, China

To whom correspondence should be addressed: Xinguang Yu, M.D., Ph.D. [E-mail: xinguang_yu@263.net]

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evaluate the CVJ and skull base of OC, and thereby give insights into the pathogenesis of congenital OC.

MATERIALS AND METHODS

Subjects

After institutional review board approval, a retrospective review of patients operated for congenital OC at PLA General Hospital between March 2012 and March 2014 was performed. The study group enrolled 80 patients (28 men, 52 women; mean age, 35.8 ± 11.7 years). All patients were symptomatic and were diagnosed with congenital atlas assimilation. Patients with any pathologic findings, such as tumor, infection, traumatic condition, inflammatory condition, or rheumatoid arthritis, were excluded. The clinical characteristics of patients are summarized in Table 1. Additionally, subjects of the control cohort were randomly selected from our institute's clinical records database. A total of 40 subjects (26 men, 14 women; mean age, 49.0 ± 14.1 years) were recruited. All control subjects underwent cervical computed tomography (CT) scan for the reason of minor cervical or head injury and showed normal radiologic findings. Those who presented with any craniovertebral anomaly, assimilation of the atlas, bone fracture, cervical spondylolisthesis, or other cervical disease were excluded.

CT Acquisition and Data Analysis

CT scans were performed, and the CT DICOM data (0.625-mm slice thickness) were imported into MIMICS 15.0 software (Materialise, Leuven, Belgium). To ensure that all measurements were standardized, images were resliced and reconstructed based on the Frankfort horizontal plane. The CT-derived anatomic parameters were divided into the following classes: gross morphology, outlet of the foramen magnum (FM), angular measurements of the skull base, dimensions of posterior cranial fossa (PCF), and height of odontoid (Figure 1). The quantitative measurements were performed by 2 raters (R. Zong and Y. Jin) individually, and then averaged. The gross observations were recorded on agreement between these 2 researchers.

Table 1. Principal Presentation of 80 Cases with Occipitalization

Symptoms and Signs	Number of Cases	%
Appearance changes	65	81.25
Occiput/neck pain	57	71.25
Quadriparesis	55	68.75
Sensory loss	51	63.75
Ataxia	48	60.00
Lower cranial nerve dysfunction	32	40.00
Dyspnea or sleep apnea	15	18.75
Hemiparesis	14	17.50
Sphincter disturbance	13	16.25

Gross Observation

OC was classified into zones based on the failure of segmentation: the anterior arch (zone 1), the lateral masses (zone 2), and the posterior arch (zone 3) of the atlas.[7] Other associated osseous anomalies were also recorded.

Outlet of the FM

The anterior atlantodental interval (AADI) was measured as the distance between the anterior assimilated C1 arch and the odontoid process on the midsagittal image. The posterior atlantodental interval (PADI) was defined as the distance between the odontoid process and the posterior assimilated C1 arch.

The distance from the tip of the odontoid to the Chamberlain line (DCL) was measured. When the odontoid tip was above the Chamberlain line, the value was defined as positive; otherwise, the value was negative. DCL >5 mm was considered basilar invagination.

The anterior-posterior length and the horizontal width of the FM were also calculated.

Angular Measurements of Skull Base

The sagittal inferior C1 facet angle (SIA) was measured on a parasagittal section passing through the C1 lateral facet. The coronal inferior C1 facet angle (CIA) was measured on a coronal section crossing the C1 lateral mass. Both the angles were formed as a line extending from the facet and a horizontal line.

The basal angle (BA) was measured as the angle formed by an intersection of a line from the nasion to the dorsum sellae and a line from the dorsum sellae to the basion. The diagnosis of platybasia (flattening of the skull base) was made when the BA was >133°.[8,9]

The clivus-canal angle (CCA) was measured as the angle formed by a line from the dorsum sellae to the basion and a line extending from the dorsal portion of the C2 body.

Linear Dimension of the PCF

The clivus length (from the dorsum sellae to the basion), supraocciput length (from the opisthion to the internal occipital protuberance), Twining line (from the dorsum sellae to the internal occipital protuberance), and height of the PCF (calculated as a perpendicular line from the opisthion to the Twining line) were measured in a midsagittal image.[10]

Height of the Odontoid

The height of the odontoid process (HO) was defined as the distance between the apices and the superior articular facet of the C2 vertebra. The height of the C2 body (HB) was defined as the distance between the transverse process and the inferior border of the C2 body. The HB in patients with C2-3 fusion was calculated from the transverse process of C2 and the inferior border of the C2-3 fused vertebral body. Both the HO and HB were measured along the axial line of the odontoid process.

Statistical Analysis

Statistical analysis was performed using SPSS 17.0 software (IBM Inc., Chicago, Illinois, USA). Data are presented as mean ± SE. For simplicity, the bilateral SIA and CIA values were averaged prior to analysis. An independent-samples t test was applied, as appropriate, to determine the significance of measurements (except the