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Atlas Assimilation Patterns in Different Types of Adult Craniocervical Junction Malformations

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Study Design. This is a cross-sectional analysis of resonance magnetic images of 111 patients with craniocervical malformations and those of normal subjects.

Objective. To test the hypothesis that atlas assimilation is associated with basilar invagination (BI) and atlas's anterior arch assimilation is associated with craniocervical instability and type I BI.

Summary of Background Data. Atlas assimilation is the most common malformation in the craniocervical junction. This condition has been associated with craniocervical instability and BI in isolated cases.

Methods. We evaluated midline Magnetic Resonance Images (MRIs) (and/or CT scans) from patients with craniocervical junction malformation and normal subjects. The patients were separated into 3 groups: Chiari type I malformation, BI type I, and type II. The atlas assimilations were classified according to their embryological origins as follows: posterior, anterior, and both arches assimilation.

Results. We studied the craniometric values of 111 subjects, 78 with craniocervical junction malformation and 33 without malformations. Of the 78 malformations, 51 patients had Chiari type I and 27 had BI, of whom 10 presented with type I and 17 with type II BI. In the Chiari group, 41 showed no assimilation of the atlas. In the type I BI group, all patients presented with anterior arch assimilation, either in isolation or associated with assimilation of the posterior arch. 63% of the patients with type II BI presented with posterior arch assimilation, either in isolation or associated with anterior arch assimilation. In the control group, no patients had atlas assimilation.

Conclusion. Anterior atlas assimilation leads to type I BI. Posterior atlas assimilation more frequently leads to type II BI. Separation in terms of anterior *versus* posterior atlas assimilation reflects a more accurate understanding of the clinical and embryological differences in craniocervical junction malformations.

Key words: atlas assimilation, basilar invagination, cervical atlas, Chiari malformation.

Level of Evidence: N/A

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Craniovertebral junction malformation (CCJM) is a complicated and controversial topic in the literature. Although there are several smaller malformation possibilities in the craniocervical junction, in the clinical context as regards adults, these malformations have been described as Chiari malformations (CMs) and basilar invagination (BI).[1,2] CM is a herniation of the cerebellar tonsil inside the foramen magnum and BI is a condition in which there is a prolapse of the upper cervical spine in the direction of the cranial base.[2,3]

More recently, BI has been subdivided into type I, with direct brainstem compression due to odontoid process indentation, and type II, with a reduction in posterior cranial fossa volume and CM.[4] The description of new cases of BI associated with instability has allowed the identification of a new pattern of malformation, independent of the presence of CM but associated with the type of atlas assimilation. The atlas's anterior arch assimilation has been found to be invariable in cases of BI with instability. The embryonic sites of formation of the anterior arch of the atlas and atlantoaxial ligament system are the same, such that the assimilation of the anterior arch of the atlas is often associated with atlantoaxial instability.[5]

Regardless of the presence of cerebellar tonsil herniation, BI has been divided into type I and type II: with and without instability, respectively. The type of atlas assimilation seems to be important in determining the type of malformation[3]

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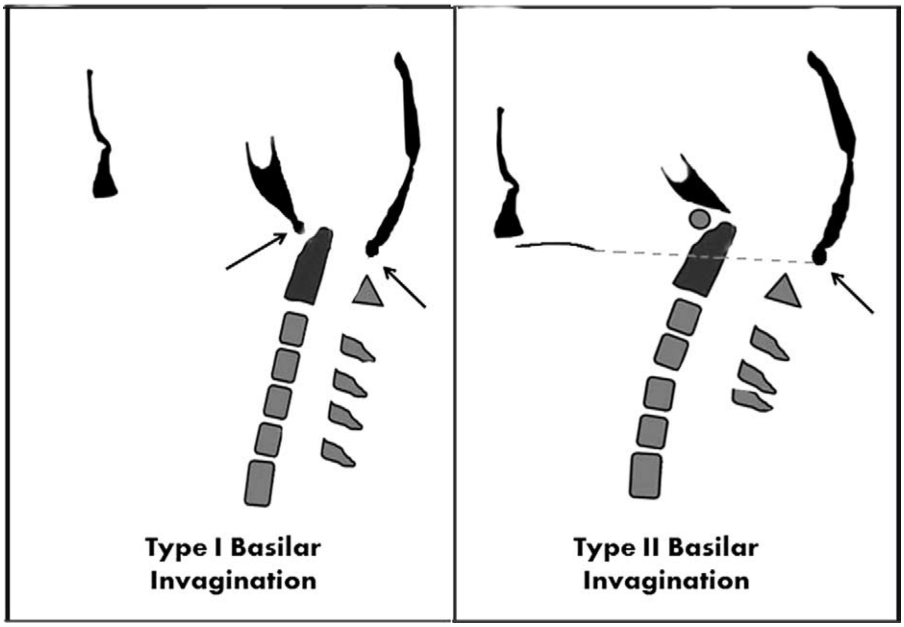

Figure 1. Basilar Invagination classification. Left: Basilar invagination type I associated with craniocervical instability (black arrows: assimilation of archs anterior and posterior). Right: Basilar invagination type II (dotted line: Chamberlain line; black arrow: arch posterior assimilation).

(Fig. 1). The objective of this work is to test the hypothesis that, in the adult CCJM, atlas assimilation is associated with BI and that the atlas's anterior arch assimilation is associated with craniocervical instability and type I BI.

MATERIALS AND METHODS

The study was approved by the ethics committee and developed in accordance with international ethical regulations.

We studied images database of adult patients with primary craniocervical junction malformations. Patients with basilar impression secondary to rheumatoid arthritis, as well as patients with trauma, tumor or infection in the craniocervical junction, were excluded from the study. The patients with CCJM were separated into 3 groups:

1. Symptomatic patients with cerebellar tonsil herniation, posterior fossa structures and cisterna magna compression were diagnosed with Chiari type I malformation or symptomatic adult CM;
2. Patients with odontoid invagination into the foramen magnum were classified as having type I BI; and
3. Patients with odontoid invagination toward the skull base but without invagination into the foramen magnum were classified as having type II BI[6] (Fig. 1).

The control group was composed of a sample of normal MRI scans, classified as such by the radiology service, which were matched by age and sex to scans from the CCJM group.

For this study, we evaluated the midline MRI T1 and T2-weighted scans (and/or CT scans) of patients with CCJM and normal subjects. The images came from a database of MRIs of patients treated between 1996 and 2012 in 2 public hospitals in São Paulo, Brazil, and the data were analyzed as a cross-sectional (transversal) study.

Atlas Arch Assimilation Classification

The atlas arch assimilations were classified according to their embryological origin: (Fig. 2)

- Posterior arch assimilation,
- Anterior arch assimilation, or
- Assimilation of both the anterior and posterior arches.

The prevalence of the atlas assimilation patterns was described for each type of abnormality.

Statistic

Student *t* test was used to test the mean difference between the case and control groups' ages. The χ^2 test was used to compare the sex distribution among the 3 CCJM subgroups and the control group.

RESULTS

We studied the craniometric values of 111 subjects, of whom 78 had CCJM and 33 were normal subjects.

Of the 78 malformations, 51 patients had a CM and 27 had BI, of whom 10 had type I BI and 17 presented with type II BI. 1 patient in the CM group and another in the type II BI group had MRI or CT scans that were inadequate for analysis, resulting a final total of 109 subjects for analysis. There were no statistical differences between the age or sex distribution of CCJM and control groups. Demographic data of the final sample are detailed in Table 1.

Atlas Assimilation Patterns

Chiari Malformation Group

In the CM group, 41 showed no assimilation of the atlas (82%) (Fig. 3).

TABLE 1. Demographic Data of Age (Mean ± Standard Deviation) and Sex Distribution for Each Group

Group	Age	% Sex (Male/Female)
BI I	46.0 ± 10	50/50
BI II	50.3 ± 10	55/45
Chiari	46.1 ± 14	43/57
Control	44.9 ± 12	57/43

18% of patients had atlas assimilation: none had isolated anterior arch assimilation, 6 had isolated posterior atlas assimilation (12%) and 3 (6%) had assimilation of both arches (Fig. 4).

Type I BI Group

In type I BI group, 2 patients (20%) had isolated assimilation of the anterior arch and 80% had assimilation of both arches. All of the patients were presented with atlas assimilation. Alone or associated with the assimilation of the posterior arch, all patients (100%) had anterior arch assimilation (Fig. 5 and 6).

Type II BI Group

In type II BI group, 2 patients had isolated assimilation of the anterior arch (12%), 6 had isolated assimilation of the posterior arch (38%), 4 patients had assimilation of both arches (25%), and 4 had no assimilation (25%). 63% of the type II BI patients had posterior arch assimilation, isolated or associated with anterior arch assimilation (Fig. 7).

Control Group

In the control group, no patients had atlas assimilation (Table 2).

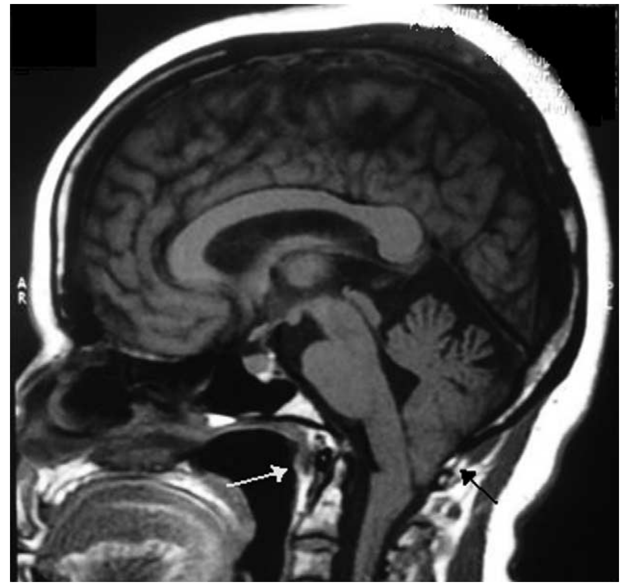

Figure 3. Adult Chiari Malformation. There is no atlas assimilation. Note the distinct anterior (yellow arrow) and posterior (black arrow) atlas arcs.

DISCUSSION

MRI elucidated new knowledge about malformations of the craniocervical junction, revealing the real anatomy of the anatomic structures involved.

Although a multitude of isolated malformations in the craniocervical junction are possible,[1] from a clinical point of view, the adult CCJM has been described in terms of CM and BI.[2] The most commonly associated bone anomaly in CCJM is atlas assimilation. However, isolated atlas assimilation rarely produces clinical pathology.[1]

The developmental abnormalities of the craniocervical junction have been embryologically divided into malformations of the central pillar and malformations of the surrounding rings. Malformations of the central pillar would give rise to BI, whereas malformations of the surrounding rings, which participate in occipital bone development, would be responsible for CM[7] (Fig. 2).

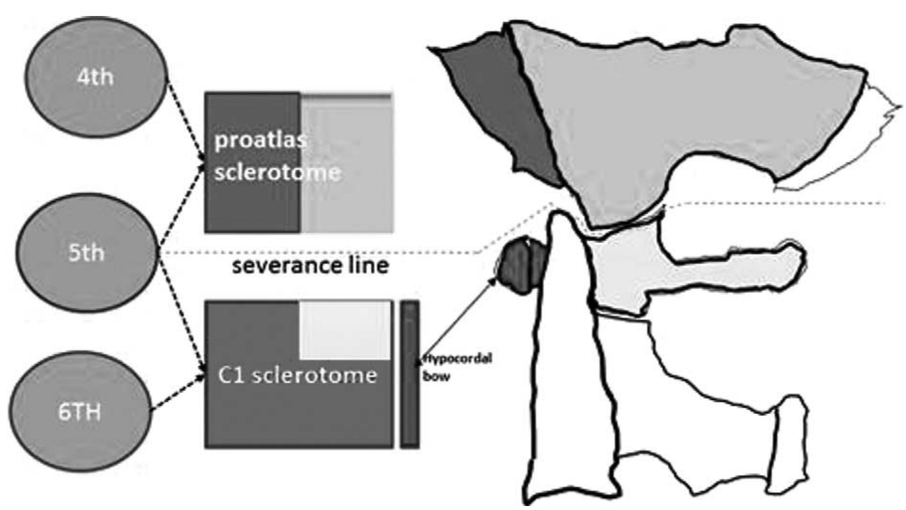

Figure 2. Embryological origin of craniocervical bones. Origin of craniocervical bones according to embryology. The 4th and 5th somites are the origin of the proatlas sclerotome and then the occipital bone. The 5th and 6th somites are the origin of the C1 sclerotome. In the 4th developmental week, the occipital and cervical bones are separated by a severance line.

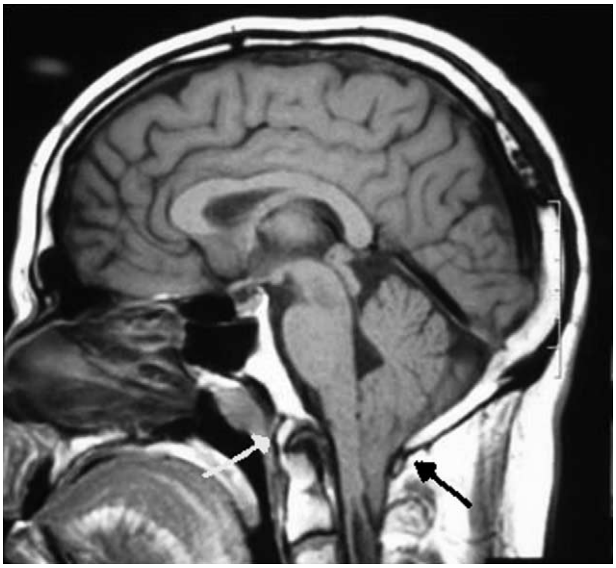

Figure 4. Adult Chiari Malformation. Note the anterior atlas assimilation (yellow arrow) and posterior atlas assimilation (black arrow).

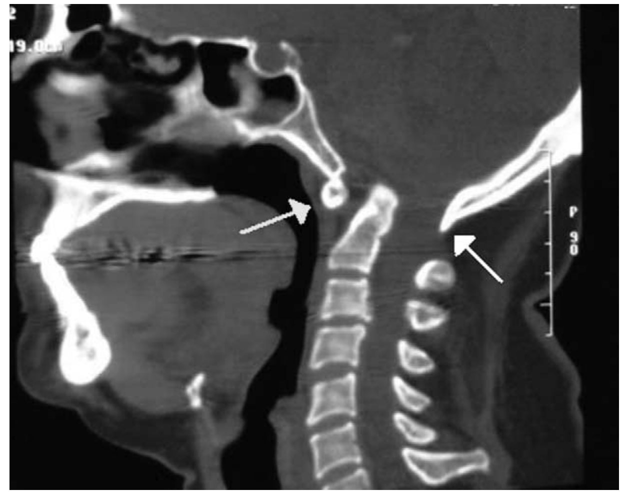

Figure 6. Type I basilar invagination. Note the odontoid process inside the foramen magnum. Yellow arrow: anterior atlas assimilation. White arrow: posterior atlas assimilation.

Morphometric studies have revealed that these malformations belong to a continuum of malformations, with CM representing the least-affected pole and BI the most affected pole.[2,3]

Classification of types of BI was performed long ago by Menezes *et al*.[8] They described BI in terms of therapeutic prognosis: namely, as reducible *versus* irreducible cases. In some so-called reducible cases, the authors showed the presence of anterior atlas assimilation.[8]

Goel classified invagination in terms of types I and II. Type I is associated with instability and ventral neural compression caused by the axis's dens, and type II is associated with CM. Experience has revealed that the presence of CM (tonsillar herniation) or lack thereof has been erratic and inconsistent.[4]

In another, separate analysis, BI was described as type I, associated with instability and vertical dens insinuation into the foramen magnum, and type II, in which there is projection of the odontoid in the direction of the cranial base, without invagination into the foramen magnum and without instability (Fig. 1).[3]

Analyses of reports of unstable BI[3,9–11] and the analysis of our sample have shown that, for type I BI, anterior arch

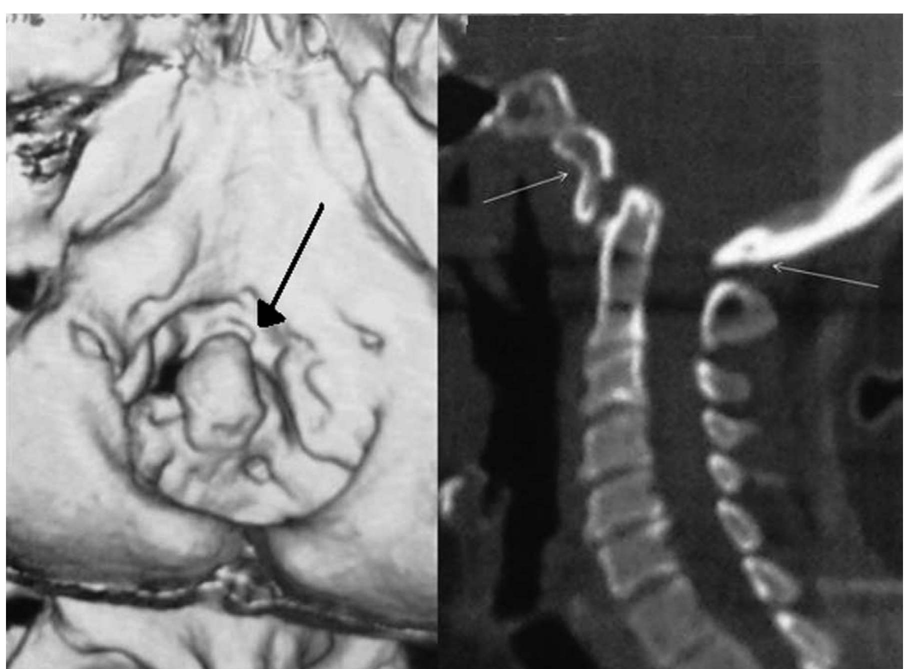

Figure 5. Left: axial view of anterior and posterior atlas assimilation. Note that the atlas has become part of the skull (black arrow). Right: Sagittal view. Anterior and posterior arch assimilation (white arrows).

TABLE 2. Frequency of Assimilations Types for Each Group

CJMM	Anterior Arc	Posterior Arc	Both Arcs	None Assimilation	% Assimilations
BI I	2 (20%)	0	8 (80%)	0	100%
BI II	2 (12%)	6 (38%)	4 (25%)	4 (25%)	75%
Chiari	0	6 (12%)	3 (6%)	41 (82%)	18%
Control	0	0	0	33 (100%)	0%

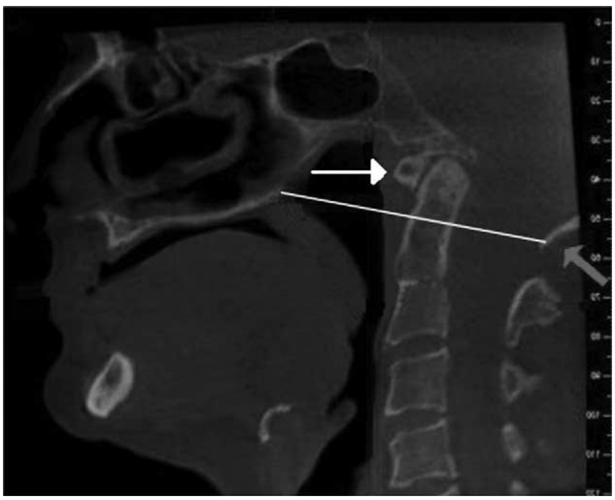

Figure 7. Type II basilar invagination. Yellow line: Chamberlain's line. White arrow: anterior arch. Red arrow: posterior arch assimilation. There is a block at the clivus preventing the dens from migrating upward to the foramen magnum.

assimilation is a basic condition for pathology. Anterior atlas assimilation leads to type I BI unless the odontoid process' upward migration is blocked by a bone bulkhead—commonly, the clivus (Fig. 7).

Atlas assimilation arises when the first cervical somite fails to split into its cranial and caudal components. Consequently, the atlas becomes assimilated into the occipital region.[11,12] Embryologically, it originates from the hypochordal bow and the posterior arch in the C1 lateral sclerotome zone. The assimilation of each arch occurs at different embryonic regions and at different stages of body formation[7] (Fig. 2). The differences in embryological origin between the 2 axial arches support the atlas assimilation classification used in this paper.

Atlas assimilation incidence has been estimated at 0.25% among the general population and 0.5% to 1.0% in Caucasians.[2] Because the mesodermic malformation is the foundation for all CCJMs, from CM to BI, it was presumed to be widely found in all CCJMs. This assumption provided the motivation to include CM cases in this analysis. The normal subjects (control group) were included to reveal differences between the CCJM group and normal subjects, should such differences exist.

The assimilation of the anterior arch of the atlas is present in all type I BI cases, and this assimilation is the necessary condition for this malformation (Fig. 5 and 6).

The assimilation of the anterior atlas arch occurs conjointly with ligament deficiency and prevents the axial ligament stabilizer system from developing properly. Upward movement of the vertical axis may occur.

Several authors have considered atlas assimilation as a predisposing factor for atlantoaxial instability.[5,6,10,13]

Assimilation of the posterior arch of the atlas is most commonly found in type II BI malformation (Fig. 7). In CM, atlas assimilation also occurs, but more rarely.

The most frequent embryological factor seems to be related to the underdevelopment of the occipital bone and not to craniovertebral separation.

Analysis of anterior arch assimilation images suggests that foramen magnum dens invagination may not occur depending on the dens migration axis and the presence of any osseous impediment to the odontoid's rise (Fig. 4 and 7). Incomplete separation between the atlas and the occipital bone is associated with ligament incompetence between the atlas and the odontoid, which allows vertical instability with odontoid insinuation into the foramen magnum and type I BI.[3,9,10,13]

Atlas assimilation may involve the anterior arch of the atlas, the lateral masses, or the entire atlas. Clinical and embryological evidence suggests the separation of atlas assimilation into anterior and posterior types. Lateral and posterior atlas assimilations form 2 parts of the same embryologic problem. Posterior arch assimilation extends the opisthion of the skull downward and moves the Chamberlain line to a lower point in relation to the C2 vertebra (Fig. 7).

The limitations of the study are related to the use of an isolated sample, what is usually observed on most papers on this area. These limitations will be corrected with the conduction of future multicenter studies. This manuscript provides information to evaluate procedures related to the possibilities of reduction and stabilization of instable cases.

CONCLUSION

Normal subjects and subjects presenting with CM and BI types I and II have different distributions of atlas assimilation, suggesting different embryological site malformations. Anterior atlas assimilation leads to type I BI and atlantoaxial instability. Posterior atlas assimilation more frequently leads to type II BI.

The separation of cases in terms of anterior and posterior atlas assimilation reflects a more accurate understanding of the clinical and embryological differences in craniocervical junction malformations.

➢ Key Points

- ❑ Regardless of the presence of cerebellar tonsil herniation, BI has been divided into type I and type II: with and without instability, respectively. The type of atlas assimilation seems important in determining the type of malformation.
- ❑ The assimilation of the anterior atlas arch occurs conjointly with ligament deficiency and prevents the axial ligament stabilizer system from developing properly. Upward movement of the vertical axis may occur.
- ❑ Anterior atlas assimilation was found to be significantly more prevalent in patients with BI type I, and this condition seems to have a relationship with craniocervical instability.
- ❑ BI was described as type I, associated with instability and vertical dens insinuation into the foramen magnum, and type II, in which there is projection of the odontoid in the direction of the cranial base, without invagination into the foramen magnum and without instability.

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