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Basilar Invagination: cranio-cervical kyphosis rather than prolapse from the upper cervical spine

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ABSTRACT

Background: Adult craniocervical junction malformations have been described as Chiari malformation (CM) and Basilar invagination (BI). Recently, angular craniometric studies have identified differences among subtypes of malformations and reveled new relationships between skull and spine.

Objective: The scope of this study is to summarize the knowledge related to craniometric relationship in pathophysiology of these diseases.

Results: In CM, angular craniometric measures are not different from normal controls. In BI type I and II there is an increased craniocervical Kyphosis associated to cervical spine lordosis. Chamberlain's line violation evaluation reveals that there is a downward angulation of the skull towards the upper cervical spine in BI.

Conclusion: BI should be considered a craniometrical kyphosis rather than a prolapse of the cervical spine to the skull base.

Introduction

Craniocervical junction malformations (CCJM) have been described more frequently as adult Chiari Malformation (CM) and Basilar Invagination (BI)[1]. Chiari malformation is defined as the herniation of the cerebellar tonsils in the foramen magnum with compression of the neural structures and / or of the cerebrospinal fluid at the foramen magnum level[2]. Basilar invagination (BI) is classically described as a developmental anomaly of the craniovertebral junction in which the odontoid abnormally prolapses into the foramen magnum[3]. BI was recently divided into those associated with craniocervical instability in which odontoid process invaginates inside foramen magnum (Type I Basilar Invagination) and those not associated with instability but with flattened basecranium (Type II Basilar invagination) (**Figure 1**)[4,5,6]. Linear and angular craniometric studies helped to clarify pathophysiology of these malformations[5]. Craniometric analysis of skull and cervical spine has shown interrelation between craniocervical kyphosis and cervical spine lordosis suggesting some new correlation with potential therapeutic importance[5]. The objective of this study is to demonstrate the role of craniocervical junction kyphosis in pathophysiology of basilar invagination, confronting with the classic concept of upper cervical spine prolapsed into the skull base.

Methods

To demonstrate the objective of this study, three types of published data will be presented: First, data comparing craniocervical angular measurements in normal subjects, Chiari malformation and Basilar

Invagination patients will be displayed. Specific measurements for BI will be pointed[5]. After that, relationship between craniocervical junction kyphosis and cervical lordosis angle will be analyzed[5,7]. At last, data revealing craniocervical kyphosis reduction, odontoid process prolapse normalization, cervical angle lordosis correction after skull realignment and basilar invagination treatment will be presented[8,9,10,11,12].

Angular Craniometric Point Definitions

Only four points have been suggested to measure sagittal human Cranio-cervical junction angulations in magnetic resonance image **(**Figure 1**)**: Nasion, Top of dorsum sellae, Basion and Opistion[7,13,14].

Craniometric points permit to identify and measure primary cranial angles (Basal and Boogard´s angle) and secondary craniocervical angles (Clivus canal and Cervical lordosis angle)[5]. (Figure 2).

Results

Angular craniometric differences among normal subjects, Basilar Invagination and Chiari Malformation[5]

In a previous paper[5], 73 patients with CCJM and 33 normal subjects were studied. Sixty-seven percent had CM and 32 % BI. One third of BI patients were classified as Type I and two-thirds as BI type II. The Basal angle (BA), Clivus canal angle, Boogard's and cervical lordosis angle were compared in normal controls (CTRL), Chiari malformation and Basilar invagination patients (Figure 2). The Basal angle is fundamental to the diagnosis of Platibasia and Boogard's angle is indicative of clival horizontalization. These angles

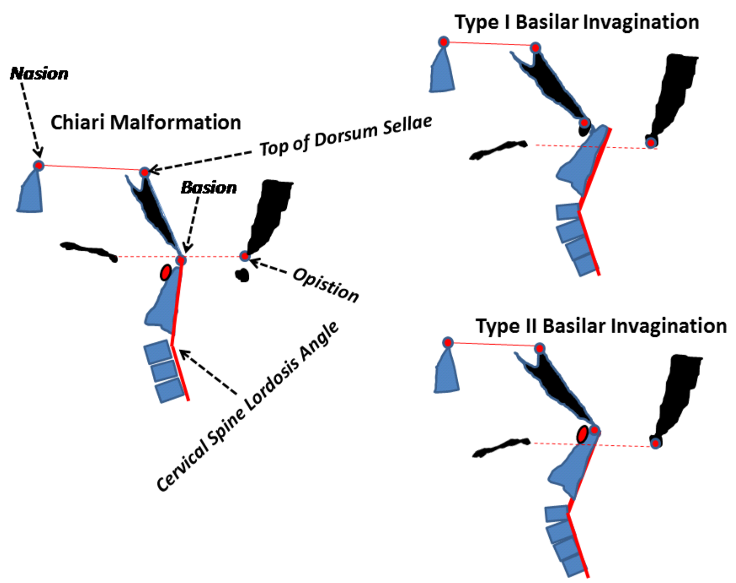

Figure 1: Circles illustrate the 4 craniometric points (Nasium, Top of dorsum sellae, Basion, Opistion) in Chiari Malformation (Upper left),Type I Basilar Invagination (Upper right) and Type II Basilar Invagination (Lower right).

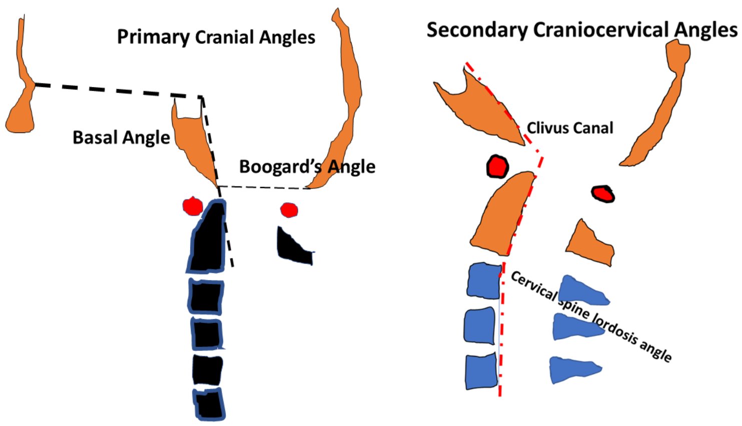

Figure 2: Craniometric ilustrations of the Basal angle (BA), Clivus canal angle, Boogard's and cervical lordosis angle[5].

were described with a greater detail in another paper[5]. Both, Clivus Canal Angle and Cervical Spine Lordosis Angle are sufficient to summarize the relation of craniocervical kyphosis and cervical spine lordosis and will be used in this manuscript to its main objective.

Clivus Canal Angle (CCA)

An important difference among Basilar invagination , CM and also with control subjects is related to CCA: in BI this angle was smaller (sharper) than those of CM and CTRL groups (p <0.001) (Figures 1 and 2).

Cervical Spine Lordosis Angle

The BI group showed higher lordosis (lower angle) than the CM (p<0.001) and the CTRL groups (p<0.001). There were no differences between the CM and the CTRL groups (p=0.347) (Figures 1 and 2).

Correlation between cervical lordosis angle and Clivus canal angle[5,7]

Cervical lordosis angle assumed a direct relation with Clivus canal angle (R= 0.439;p<0,01).

An acute clivus canal angle was associated with an acute Lordosis angle. This suggests that cervical lordosis compensates Cranio-cervical kyphosis in the maintenance of the horizontal gaze (Figures 2 and 3).

The Chamberlain's line violation (CLV)[7]

The position of hard palate reveals angular and linear relationship between skull and upper cervical spine. For centuries, the Chamberlain's line (CL) was used as a recognizable relationship between hard palate and Odontoid tip. Violation of CL by Odontoid was considered diagnosis of Basilar Invagination but the amount of violation varied abroad in the literature[7]. Recently the CLV in BI has been objectively measured in the MRI era[7].

In a previous paper[7], a total of 97 subjects were studied: 32 normal subjects, 41 CM patients, 9 basilar invagination type 1 (BI1) patients, and 15 basilar invagination type 2 (BI2) patients.

The mean CLV violation in the groups were: The control group, 0.16 ± 0.45 cm; The CM group, 0.32 ± 0.48 cm; The BI1 group, 1.35 ± 0.5 cm; and the BI2 group, 1.98 ± 0.18 cm. CLV was not different between CM and normal subjects.

There was a strong correlation between CLV and the clivus Canal angle (R = 0.7, P = 0.000) and Cervical Lordosis angle (R = -0.39, *P* = 0.000). As the craniocervical angle becomes kyphotic, cervical spine becomes lordotic in a compensatory way.

The importance of Atlas assimilation in Basilar invagination[15]

In one study, in the type I BI group, all patients presented with anterior arch assimilation (AAA), and 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. It was suggested that anterior atlas assimilation leads to type I BI. Posterior atlas assimilation more frequently leads to type II BI. At least in Type I BI, AAA is strongly associated with craniocervical instability and craniocervical junction kyphosis **(Figure 3.)**

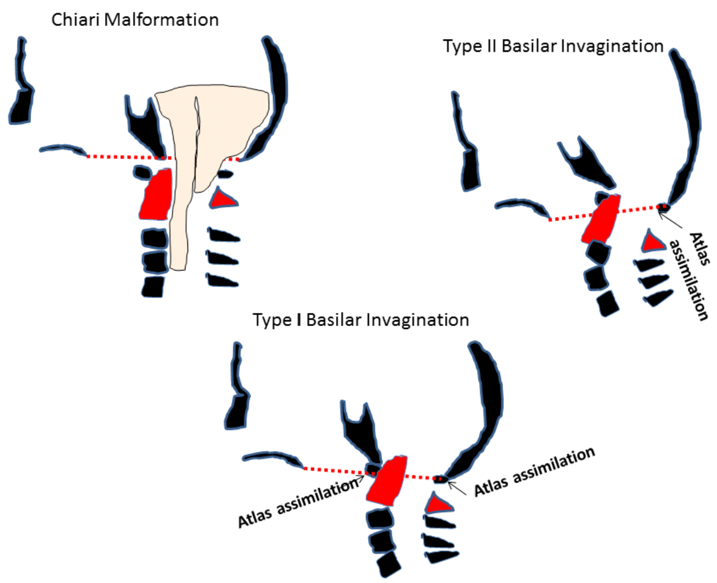

Figure 3: The red dotted line represents Chamberlain's line violation in the three types of CCJM. Note Atlas assimilation in Basilar invagination figures.

The effect of craniocervical kyphosis realignment in BI[8-12] (Figures 4 and 5)

Several papers have demonstrated the role of reduction by traction or distractive surgeries in restoring craniocervical kyphosis.

Publications of skull realignment have demonstrated that some altered craniometric angulations previous considered as part of the BI syndrome can be normalized: the clivus canal angle becomes less acute, the craniocervical kyphosis was reduced, the tentorial slope became more vertical. The odontoid tip becames projected under the McRae line. In this way, skull realignment treats Basilar Invagination.

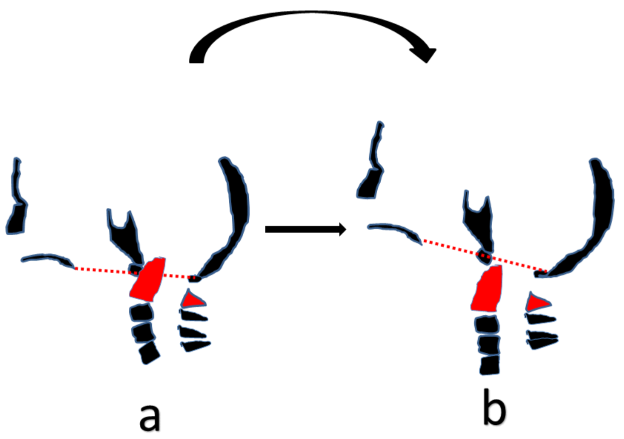

Figure 4: Illustration of Type I Basilar Invagination reduction. Reduction (frequently done by head extension) normalizes CLV (red dotted line), Cranio-cervical kyphosis and cervical spine lordosis.

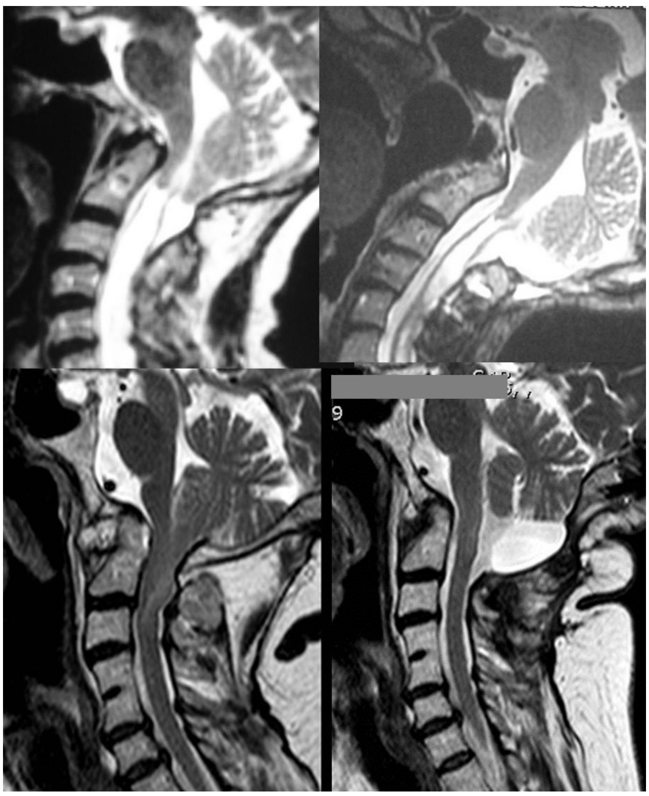

Figure 5: Upper part: type II BI before skull realignment (Left) and after (Right).Lower part: type I BI before skull realignment (Left) and after

Figure 4 illustrates a type I BI with extension of the head.

Discussion

The (BI) on its part has been described as a developmental anomaly of the craniovertebral junction in which the odontoid abnormally prolapses into the foramen magnum[3].

BI has been classified into those associated with craniocervical instability in which odontoid process invaginates inside foramen magnum (Type I Basilar Invagination) and those not associated with instability but with somehow flattened basecranium (Type II Basilar invagination) (Figure 1)[4-6].

Morphological studies in the CCJM demonstrated that there was, in most cases, an underdevelopment of the posterior fossa[16]. Chiari malformation may or may not be associated with basilar invagination.

Chiari malformation has been defined as the herniation of the cerebellar tonsils in the foramen magnum with compression of the neural structures and / or of the cerebrospinal fluid at the foramen magnum level. The 5 mm limit of cerebellar tonsil herniation has been popularized after the work of Aboulezz et al[2]. However, after the identification Chiari malformation cases without evident tonsillar herniation, but associated with syringomyelia and overcrowding of posterior fossa structures (Chiari Zero)[17], the amount of tonsil herniation became less important relative to the overcrowding of neural structures at the foramen magnum that became the Paramount for the malformation identification[17].

Chiari malformations have been considered for centuries as a neuro-dysgenesis. After the experimental work of Marin Padilla[16] treating hamsters with high doses of vitamin A, the pathophysiological approach turned to mesodermal defects.

Several disorders of metabolism and bone formation may lead to the under-development of the occipital bone, a shallow posterior fossa and secondary cerebellar tonsils herniation. However, in most cases of Chiari malformations, no primary cause is identified. Chromosomes 9 and 5 have been associated with Chiari malformation[18].

Angular correlations between skull and spine have been studied in anthropology and evolutionary biology and actually in Cranio-cervical junction malformations[19].

For centuries, basilar invagination has been described as the prolapse of superior cervical spine towards skull.

Craniometric studies revealed that in basilar invagination, different from normal subjects and CM, there is an decrement of Clivus canal angle, indicating craniocervical kyphosis and enlargement of cervical spine lordosis. There is an exaggerated angulation in kyphosis from the skull towards the spine.

The analysis using Chamberlain's line violation bring data of the Skull position related to the upper cervical spine. The hard palate is part of the "viscerocranium". As the skull bends anterior and inferiorly toward the spine, more violated became the Chamberlain's line by odontoid process, more acute the clivus-canal angle, more intense the Cranio-cervical kyphosis and ventral brainstem compression and more lordotic becomes the cervical spine, probably equilibrating the horizontal gaze.

Surgical series have shown that normalization of the clivus canal angle, fusion-stabilization is associated with clinical improvement and craniocervical junction kyphosis can be improved by skull realignment, contributing to the notion of Skull involvement in BI pathogenesis.

Unfortunately, not all BI are reduced. Meneses et all[20]. called attention that majority of cases should be reducible if treated earlier.

Conclusion

Data presented suggests that the classic definition as a prolapse of the upper cervical spine towards the skull base must be changed to concept of Cranio-cervical kyphosis leading to basilar invagination.

Conflict of interest declaration

There is no conflict of interest to be declared.

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