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ARTICLE

Incidence of basilar invagination in patients with tonsillar herniation – a case control craniometrical study

Incidência da invaginação basilar com heriação tonsilar – estudo da craniometria do tipo caso-controle

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ABSTRACT

A retrospective case-control study based on craniometrical evaluation was performed to evaluate the incidence of basilar invagination (BI). Patients with symptomatic tonsillar herniation treated surgically had craniometrical parameters evaluated based on CT scan reconstructions before surgery. BI was diagnosed when the tip of the odontoid trespassed the Chamberlain's line in three different thresholds found in the literature: 2, 5 or 6.6 mm. In the surgical group (SU), the mean distance of the tip of the odontoid process above the Chamberlain's line was 12 mm *versus* 1.2 mm in the control (CO) group ($p<0.0001$). The number of patients with BI according to the threshold used (2, 5 or 6.6 mm) in the SU group was respectively 19 (95%), 16 (80%) and 15 (75%) and in the CO group it was 15 (37%), 4 (10%) and 2 (5%).

Keywords: basilar invagination, tonsillar herniation, craniometrical evaluation.

RESUMO

Realizamos estudo retrospectivo tipo caso-controle baseado na avaliação craniométrica para avaliar a incidência da Invaginação Basilar (IB). Pacientes com herniação tonsilar sintomática tratada cirurgicamente foram avaliados quanto a parâmetros craniométricos obtidos em reconstrução de TC antes da cirurgia. IB foi diagnosticada quando a ponta do odontóide passava acima da linha de Chamberlain em 2, 5 ou 6,6 mm. No grupo cirúrgico (CI), a distância média da ponta do odontóide acima da linha de Chamberlain foi de 12 mm *versus* 1,2 mm no grupo controle (CO) ($p<0.0001$). O número de pacientes com IB conforme o critério diagnóstico usado (2, 5 ou 6,6 mm) foi de 19 (95%), 16 (80%) e 15 (75%) no grupo CI, respectivamente, contra 15 (37%), 4 (10%) e 2 (5%) no grupo CO. Pacientes com herniação tonsilar tinham maior incidência de IB comparados ao grupo controle.

Palavras-chave: invaginação basilar, heriação tonsilar, avaliação craniometrical.

Basilar invagination (BI) is a developmental anomaly of the occipital bone and upper cervical spine resulting in an abnormally high vertebral column prolapsed into the skull base[1]. As a radiological finding, its diagnosis is made when the tip of the odontoid process crosses above the Chamberlain's line (a line traced from the posterior margin of the hard palate to the dorsal margin of the foramen magnum). According to different authors, in order to establish the diagnosis of BI, the odontoid tip should be above the Chamberlain's line in 2 mm, 5 mm or even 6.6 mm[2,3,4].

Chiari type I malformation (CM), described in 1891 by Hans Chiari, is probably the most important and prevalent symptomatic congenital craniocervical malformation[2,3,4,5]. It is secondary to a caudal rhombencephalon abnormality with displacement of the cerebellar tonsils into the cervical canal, also commonly associated with syringomyelia[6,7,8]. Diagnosis of CM is mainly based on finding of tonsillar ectopia. However, there is controversy regarding the amount of herniation to be labeled as abnormal. Most radiologists define CM as when the tonsils descend more than 5 mm below the foramen magnum. Other radiological

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findings can also be present, such as compression of the posterior fossa subarachnoid space, excessive crowding in the posterior fossa, a small posterior fossa volume, excessive slope of the tentorium, medullary kinking and basilar impression[9].

Several large surgical series have described the association between CM and BI, although most of them did not report the exact criteria used for diagnosing BI. The incidence varies among the different studies. Milhorat et al. reported that in 364 patients treated for symptomatic CM, 22 (6%) had BI[9]. In a series published by Fenoy et al., with 234 patients treated for hindbrain herniation, 56 (23.9%) had BI[10]. Silva et al. reported that 67 (65.6%) of 102 patients with CM had BI, an even greater incidence than previous studies[6].

The variability of incidence of BI in patients with CM may be due to the lack of strict criteria for diagnosis of this congenital axial skeleton anomaly. We performed a case-control study in order to evaluate the relationship between CM and BI. Our primary hypothesis is that the incidence of BI will be higher in patients with tonsillar herniation and, using more sensitive criteria for diagnosis, almost all these patients would also have the diagnosis of BI.

METHOD

We performed a retrospective craniometrical study of 20 adult patients with tonsillar herniation secondary to congenital craniocervical junction anomalies treated from 2009-12 in our institution, Clinical Hospital of the State University of Campinas, a tertiary neurosurgical center. All these patients underwent posterior fossa decompression for treatment of symptomatic tonsillar herniation, with or without concomitant fixation and fusion. Fusion was performed in patients with evidence of instability on static radiological exams (C1-2 or C2-3 listhesis or occipital-C1-2 subluxation of the facet joints) or in dynamic CT scans (increase mobility between C1-2). A control group of forty adult patients treated for non craniocervical pathologies was randomly chosen as a control group, chosen by random selection from the hospital radiological database. In both groups, only patients older than 17 years were selected. All had at least a complete preoperative CT scan with axial, sagittal and coronal reconstruction of the craniocervical junction.

In addition to clinical data (age and sex), the following radiological measurements were collected in both groups (see Figures 1 and 2 with cases examples) by two researchers (RNM and UCB) and all their measurements were

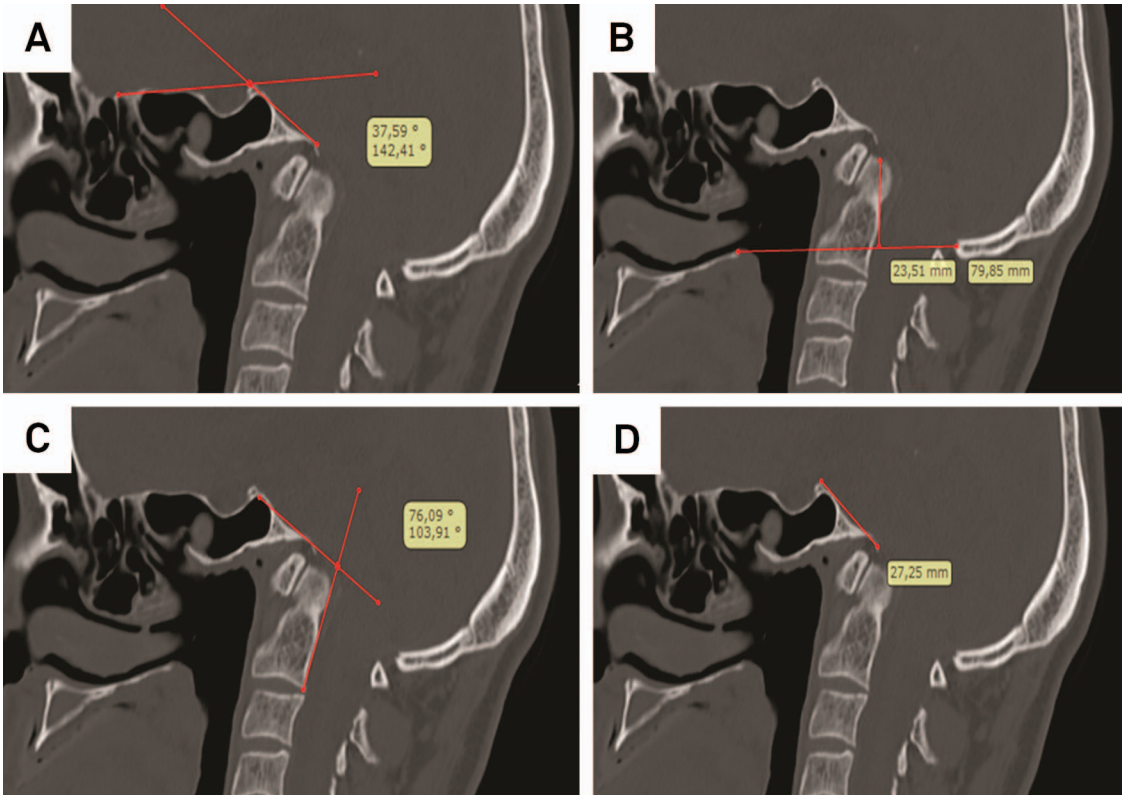

Figure 1. Measurements performed in a case patient. Welcher basal angle: it was measured from a line extending from the planum sphenoidale and another from the clivus (A). Chamberlain's line - extending from the posterior edge of the hard palate to the opistium with the tip from the odontoid process (B). The clivus canal angle: measured from a line along the clivus crossing with a line along the posterior portion of the body of the axis (C).The clivus extension (D).

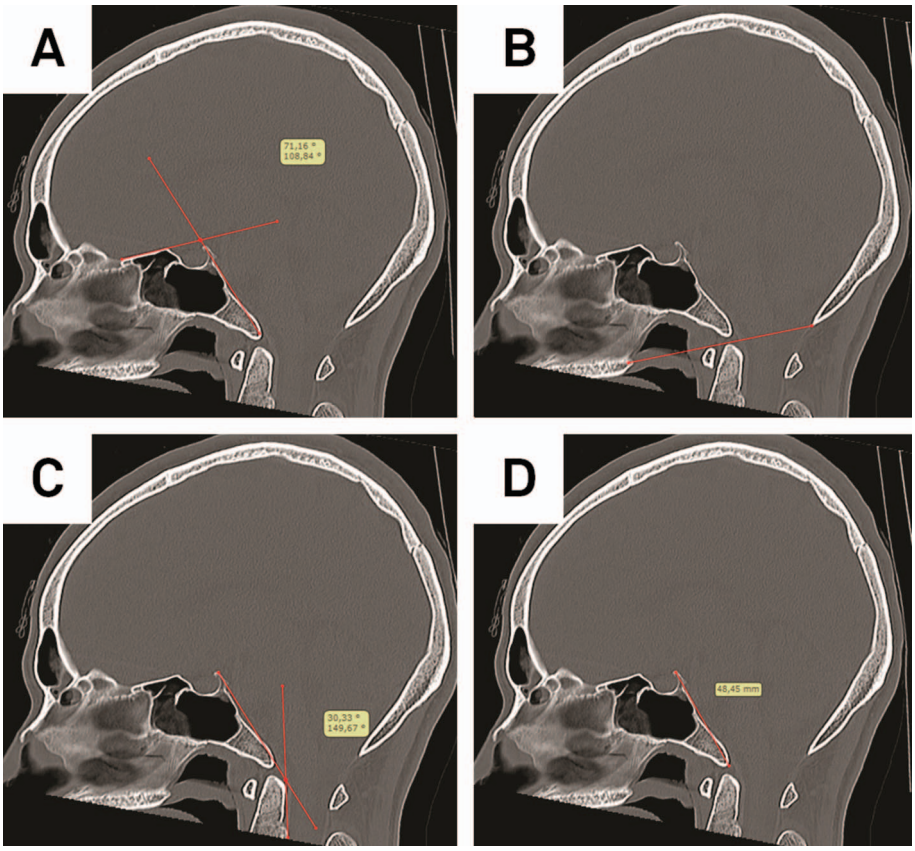

Figure 2. Measurements performed in a control patient. Welcher basal angle: it was measured from a line extending from the planum sphenoidale and another from the clivus (A). Chamberlain's line - extending from the posterior edge of the hard palate to the opistium (B). The clivus canal angle: measured from a line along the clivus crossing with a line along the posterior portion of the body of the axis (C). The clivus extension (D).

checked for accuracy by the main researcher (AFJ) to attest precision of the measurements:

1) Clivus-canal angle: It was measured at the meeting point of a line drawn along the clivus and of a line drawn along the posterior portion of the body of the axis;

2) Welcher basal angle: It was measured from a line extending from the planum sphenoidale crossing with another from the clivus;

3) Presence of tonsilar herniation greater than >5 mm;

4) Clivus length;

5) Distance of the tip of the odontoid process by applying the Chamberlain's line;

6) Incidence of basilar invagination: It was calculated in both groups using three different criteria proposed in the literature: tip of the odontoid above 2 mm[8], 5 mm[13] or at least 6.6 mm above the Chamberlain's line[14].

Analysis of the obtained data of the craniometrical parameters between groups was performed using an unpaired t-test; comparisons of gender and number of patients were analyzed using a two-tailed Fisher's exact test. Statistical significance was determined at $p<0.05$. There were no external funding sources for this study. Ethical approval was obtained (P 16122313.9.00005404).

RESULTS

Surgical group

There were 23 patients with craniocervical junction anomalies treated in the study period, but three of them were excluded from the study because they had acquired anomalies (one craniocervical compression secondary to tuberculosis and two patients with rheumatoid arthritis and atlanto-axial instability). A total of 20 patients were included. All were treated surgically with foramen magnum decompression for symptomatic tonsillar herniation. Three patients (3/20 - 15%) had clear C1-2 instability with anterior lysthesis of C1 over C2 and underwent craniocervical fusion with instrumentation (Table 1).

Ten (50%) were males and ten (50%) were females. Age ranged from 19 to 73 years-old (mean of 44.3, median of 44.5 years-old, standard deviation of 15.1). The basal angle in this group ranged from 95.5 to 143.6º (mean of 120.2º, median of 118.6º and standard deviation of 12.8). The clivus canal angle ranged from 92 to 171º (mean of 133.3º, median of 133.4º and standard deviation of 20.3). All of the twenty patients had tonsillar herniation greater than 5 mm. The clivus length ranged from 11.6 to 42.3 mm (mean of 28.3, median of 28.6 mm and standard

Table 1. Clinical and radiological data of the patients included in the present study.

		20 cases	40 control	
Sex	Men	10 (50%)	26 (65%)	p>0.05
	Female	10 (50%)	14 (35%)	
Age (years-old)	Range	19 to 73	18 to 88	p>0.05
	Mean	44.3	45.6	
	Median	44.5	41.5	
	Standard deviation	15.1	20.3	
Basal angle	Range	95.5-143.6°	94-130.8°	
	Mean	120.2°	112.5°	p=0.0104
	Median	118.6°	112.9°	
	Standard deviation	12.8	9.4°	
Clivus-canal angle	Range	92-171°	139-169°	
	Mean	133.3°	154.9°	p<0.0001
	Median	133.4°	155°	
	Standard deviation	20.3°	5°	
Clivus length	Range	11.6 to 42.3 mm	29.3 to 49.4 mm	
	Mean	28.3 mm	38.2 mm	p<0.0001
	Median	28.6 mm	37.6 mm	
	Standard deviation	7.8 mm	4.1 mm	
Relationship between the tip of the odontoid / Chamberlain's line	Range	0 to 26.3 mm	-4.9 to 9 mm	
	Mean	12	1.2	p<0.0001
	Median	8.7	0.25	
	Standard Deviation	8.1	3.23	

deviation of 7.8). All patients had the tip of the odontoid process crossing above the Chamberlain's line, ranging from 0 to 26.3 mm (mean of 12 mm, median of 8.7 and standard deviation of 8.1 mm). According to the criteria used for diagnosis, the number of patients with BI ranged as follows (Table 2):

1) Tip of the odontoid at least 2 mm above the Chamberlain's line (criteria proposed by Goel et al.)[8] – 19 of 20 patients (95%) had the diagnosis of BI;

2) Tip of the odontoid at least 5 mm above the Chamberlain's line (criteria proposed by Smith et al.)[13] – 16 of 20 patients (80%) had the diagnosis of BI;

3) Tip of the odontoid at least 6.6 mm above the Chamberlain's line (upper limit proposed by Smoker et al.)[14] – 15 of 20 patients (75%) had the diagnosis of BI.

Control Group

Forty patients entered the control group. There were 14 (35%) females and 26 (65%) males in the control group. Age ranged from 18 to 88 year-old (mean of 45.6, median of 41.5 years-old, standard deviation of 20.3). The basal angle in this group ranged from 94 to 130.8° (mean of 112.5°, median of 112.9° and standard deviation of 9.4). The clivus canal angle ranged from 139 to 169° (mean of 154.9°, median of 155° and standard deviation of 5). None of the forty patients had tonsillar herniation. The clivus length ranged from 29.3 to 49.4 mm (mean of 38.2, median of 37.6 mm and standard deviation of 4.1). The tip of the odontoid process above the Chamberlain's line ranged from -4.9 to 9 mm (mean of 1.2 mm, median of 0.25 and standard deviation of 3.23 mm).

Number of patients with BI according to the criteria of diagnosis – the relationship between the Chamberlain's line and the tip of the odontoid (Table 2):

1) Tip of the odontoid 2 mm above the Chamberlain's line[8] – 15 of 40 patients (37.5%) had the diagnosis of BI;

2) Tip of the odontoid 5 mm above the Chamberlain's line[13] – 4 of 40 patients (10%) had the diagnosis of BI;

3) Tip of the odontoid 6 mm above the Chamberlain's line[14] – 2 of 40 patients (5%) had the diagnosis of BI.

Comparison between groups

There were no statistical differences in sex or age (p>0.05) between groups. The basal angle (p=0.0105), the clivus-canal angle (p<0.0001), the clivus length (p<0.0001) and relationship between the tip of the odontoid and Chamberlain's line were statistically different comparing both groups. Comparing the diagnosis of BI between groups, there were statistical differences in all the proposed diagnostic parameters. Those differences increased progressively along with the increase in the distance of the tip of the odontoid from the Chamberlain's line: 2 mm: p=0.0494; 5 mm: p=0.0005; 6.6 mm: p=0.0001.

Table 2. Relationship between the diagnosis of BI according to the criteria used for diagnosis in both case and control group.

Distance of the tip of the odontoid from the Chamberlain's line	Diagnosis of basilar Invagination				p
	Cases – 20 patients		Control – 40 patients		
	N	%	N	%	
>2 mm	19	95	15	37.5	0.0494
>5 mm	16	80	4	10	0.0005
>6 mm	15	75	2	5	0.0001

DISCUSSION

The first description of BI has been attributed to Ackermann in 1790, who described a craniocervical congenital malformation in Cretins from the Alps which consisted in a small posterior fossa with projection of the foramen magnum into the posterior fossa[11]. Since then, the exact definition of BI is still debated. This study suggests that a more stringent, universally applied criterion for BI diagnosis is needed to improve research and clinical outcomes.

The main etiologies of BI are clivus (basiocciput) hypoplasia, occipital condyle hypoplasia, atlas hypoplasia, atlanto-occipital assimilation and congenital atlanto-axial instability[12,13,14]. Several neural axis abnormalities are commonly described in association with BI, especially CM and syringomyelia, although hydrocephalus and syringobulbia can also be found[3]. CM was described in 1891 by Hans Chiari, as an associated reduction of the posterior fossa volume with tonsillar herniation, specifically classified according to the severity of the malformation (Chiari types 1, 2 and 3)[15,16]. Although CM and BI have much in common, they are not described as the same pathological process, but as different clinical entities.

A rigid and standardized criterion for diagnosis according to relation of the tip of the odontoid process with the Chamberlain's line is not defined in the literature, where distances range from 2 to 6.6 mm[2,3,4]. This variation on the diagnosis criteria can lead to under or over diagnosis of patients with BI and its relationship with tonsillar herniation, as shown in our results. Besides, according to the criteria utilized, the real incidence of BI, as well as its association with CM in the general population can be completely different. In our study, we observed that when the criteria used for diagnosis was modified, the incidence of BI ranged from 75 to 95% in the group with tonsillar herniation (cases) and from 5 to 37.5% in the control group. Moreover, we noticed that many patients considered to be normal had the tip of the odontoid above the Chamberlain's line. Using more sensitive criteria for diagnosing BI, as the one proposed by Goel et al. of 2 mm, we can have an extremely high prevalence (95%) of BI in the CM group. Regarding our other craniometrical measurements, platybasia is represented by the flattening of the skull base and can be attributed to an increase in the basal angle[4]. There was statistical difference between groups (p=0.0105), suggesting that a flat cranial base can be associated with tonsillar herniation and neurological symptoms of CM. The same observation is that smaller clivus length was associated with a greater prevalence of neurological symptoms and of surgical treatment. The case group had a smaller clivus (28.3 mm mean and 28.3 median) compared to the control group (38.2 mm mean and 37.6 median), suggesting that an underdevelopment of the basiocciput was associated with tonsillar herniation. Some patients can present with clinical symptoms of CM even without tonsillar herniation. Millhorat et al. proposed that CM should not be defined just for tonsillar herniation[9]. They suggested that CM is secondary to hindbrain malformation and some patients can have CM due to the obliteration of the retrocerebellar cerebrospinal fluid space without herniation – that was present in all the 364 patients treated. The presence of platybasia and a smaller clivus length in the tonsillar herniation can favoring an upper displacement of the cervical spine into the cranial base, explaining the higher incidence of BI in the case compared to control group.

Different from our findings, Botelho and Ferreira did not find differences in craniometrical measurements performed in Chiari malformation patients and control groups, but BI patients had wider basal angles and sharper clivus canal angles[17]. Of note, for these authors, BI was diagnosed when the tip of the odontoid was at least 5 mm up the Chamberlain's line, possible excluding patients with less severe forms of BI that also would have the diagnosis of CM.

Although limited by a small sample of patients and a non-blinded radiological evaluation, our study is the first of to our knowledge to address the issue of the lack of a standard radiological criterion leading to a wide range incidence of BI among CM series published in literature.

In conclusion, our study suggests that changing the thresholds for diagnosis BI can also change the prevalence of BI in patients with CM. Our findings suggest a strict relationship between CM and BI and the need for a rigid diagnosis criterion for further studies evaluating these diseases.

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