INTRODUCTION

Chiari malformations (CMs) and basilar invagination (BI – BI1 and BI2) are the most common cranio cervical junction malformations (CCJM) seen in adults; both may also be associated with syringomyelia.\[1,3,4,6,7\]

BI1 is associated with cranio cervical instability, odontoid insinuation toward the foramen magnum, and often anterior arch of the atlas assimilation.\[2-4\] BI Type 2 (BI2) is not associated with instability, but there is an exacerbation of the clivus canal angle and cranio cervical kyphosis resulting in ventral compression of the brainstem and cranial nerves.\[2-4\] Syringomyelia is frequently encountered in all of these three types of CCJM and can be correlated with a variety of increased signs and symptoms (SS).\[1\] Here, we analyzed the different frequencies of SS, between these three CCJM subtypes with/without syringomyelia.
METHODS

Study design

This study represented a cross-sectional evaluation of prospectively collected data conducted between September 2002 and April 2014 using Bindal's scale. All patients filled out informed consent to enter in this study. The research protocol was approved by the research ethics committee (CAAE 32361614.6.0000.5551).

There were 89 consecutive symptomatic patients with CCJM; 46 (52%) had CM, 43 (48%) had BI; among those with BI: 13 (30%) had BI1, and 30 (70%) had BI2. Females represented 57% of the patients with CCJM (51 patients), and they averaged 47.7 years of age (range: 11–77 years). All exhibited varying degrees of SS attributed to neural structure/posterior fossa compression and/or CSF blockage. Syringomyelia was present in 44 (49%) of all CCJM (CM and BI) patients.

Definitions and diagnostic criteria of CM, B11, and B12 malformations [Figure 1]

**CM**

Herniation (variable) of the cerebellar tonsils below the foramen magnum results in compression of the tonsils in the cisterna magna without odontoid insinuation toward the skull base.\(^3,4\)

**BI**

BI1: Odontoid insinuation toward the foramen magnum, violating the McRae line, associated with the anterior arch of the atlas assimilation, and craniocervical instability.\(^2-4,7\)

BI2: Odontoid axis process above the Chamberlain line, without violation of the foramen magnum.\(^2-4,7\)

**Syringomyelia**

Syringomyelia constitutes a fluid accumulation around the spinal cord central canal, identified as intramedullary hypersignals on T2-weighted magnetic resonance imaging (MRI).\(^1,4\)

Assessment of SS of CCJM

Bindal's standardized scale longitudinally evaluated CCJM patients' clinical manifestations and provided the following score for follow-up comparisons; each sign or symptom received a score of 10, and the score (symptomatic burden) was defined as the sum of the SS presented by the patients [Table 1].\(^1\)

### Statistical evaluation

Bindal's scores for the three types of malformations were compared using ANOVA (the Kruskal–Wallis test). SS were described by percentages and were compared between types using the Chi-squared test; this was similarly applied to patients with/without syringomyelia [Graphs 1-5 and Table 2].

| Table 1: Bindal’s scale. |
|--------------------------|
| Braintstem compression   | Syringomyelia          |
| Vertigo                 |                          |
| Diplopia                |                          |
| Hoarseness              |                          |
| Swallowing difficulty   |                          |
| Tinnitus                |                          |
| Persistent cough        |                          |
| Hearing loss            |                          |
| Nystagmus               |                          |
| Sleep apnea             |                          |
| Ataxia                  |                          |
| Shoulder pain           |                          |
| Weakness (sign)         |                          |
| Weakness (symptoms)     |                          |
| Spasticity              |                          |
| Extremity numbness      |                          |
| Neck pain               |                          |
| Headache                |                          |
| Atrophy                 |                          |
| Dissociated sensory loss|                          |
| Scoliosis               |                          |
| Extremity pain          |                          |

Based on: Bindal AK, Dunsker SB; Tew JM. (1995) Chiari I Malformation: Classification and Management. Neurosurgery

Figure 1: Craniocervical junction malformations.
RESULTS

Prevalent SS among three CCJM malformations

Predominant SS for patients with these three malformations (CCJM, B11, and B12) included neck pain, ataxia, numbness, and weakness of the extremities (e.g., with added headaches for CM patients and sleep apnea for BI patients). For CM patients, SS in descending order included neck pain, headache, and numbness of extremities followed by weakness [Graph 1]. For patients with B11, SS in descending order included weakness and neck pain, ataxia, sleep apnea, and extremity numbness [Graph 2]. For B12 patients, SS included (descending order) weakness, ataxia, nystagmus, sleep apnea, extremity numbness, neck pain, and headache [Graph 3].

The mean Bindal's score for each type of CCJM was 74.6 for CM, 78.5 for B11, and 78 for B12 (Kruskal–Wallis test, \( P = 0.9 \)) [Graph 4]. There were significant differences in SS, swallowing difficulty (\( P = 0.03 \)) and nystagmus (\( P = 0.007 \)) for the three groups [Table 2].

SS for patients with/without syringomyelia

The mean Bindal's score was 69.3 for the patients without syringomyelia and 83.4 for the syringomyelic patients (Mann–Whitney U-test; \( P = 0.054 \)) [Graph 5]. Weakness (SS), extremity numbness, neck pain, dissociated sensory loss, and atrophy were significantly more frequent (\( P < 0.05 \)) in the patients with syringomyelia.

DISCUSSION

Our understanding of CCJM (e.g., CM, B11, and B12) has evolved with the introduction of computed tomography, MRI, linear and angular craniometric studies, and genetic...
Table 2: Prevalence of signs and symptoms of craniocervical junction malformations as measured using the Chi-squared test.

| Condition                        | Chiari malformation (%) | IB1 (%) | IB2 (%) | X²(p)* |
|----------------------------------|-------------------------|---------|---------|--------|
| Vertigo                          | 41                      | 38      | 47      | 0.85   |
| Swallowing difficulty            | 13                      | 38      | 37      | 0.03   |
| Nystagmus                        | 20                      | 38      | 50      | 0.007  |
| Sleep apnea                      | 35                      | 54      | 50      | 0.29   |
| Ataxia                           | 43                      | 62      | 53      | 0.45   |
| Weakness (sign)                  | 39                      | 54      | 43      | 0.64   |
| Spasticity                       | 41                      | 46      | 37      | 0.92   |
| Extremity numbness               | 59                      | 54      | 50      | 0.75   |
| Neck pain                        | 67                      | 69      | 50      | 0.26   |
| Headache                         | 61                      | 46      | 50      | 0.51   |
| Dissociated sensory loss         | 46                      | 38      | 23      | 0.14   |
| Extremity pain                   | 39                      | 23      | 30      | 0.48   |

*Chi-square test

Graph 3: Prevalence of basilar invagination Type 2 signs and symptoms.

Graph 4: The mean Bindal’s score for each type of craniocervical junction malformations.

Graph 5: The mean Bindal’s score for each craniocervical junction malformation type with and without syringomyelia.
and embryological studies.

Notably, SS for each of these three entities differ. Although the current study found no statistically significant differences in the incidence of sleep apnea between the CCJM types, others indicated that sleep apnea was more frequent and intense in BI patients. In 43 patients with atlantoaxial dislocation and CCJM, Shuhui et al. found the following SS to be prevalent (in descending order): weakness (93%), cervical motor limitations (88%), limb paresthesia (98%), neck and shoulder pain (58%), ataxia (42%), vertigo (23%), and respiratory difficulties (7%).

Goel showed that the following symptoms were more predominant in BI1 patients: paresthesias (55%), spinothermal dysfunction (36%), cervicalgia (77%), and torticollis (41%). The study also reported that BI2 physiopathology was the same as for CM, largely attributable to the diminished cranial fossa volume contributing to SS. Our results showed that the only SS differences in the three subtypes of CCJM were swallowing difficulties (P = 0.03) and nystagmus (P = 0.007) in the BI patients.

**Bindal's scale**

Bindal's scale was created to quantify SS, and in this study, no differences were found in the SS burden between the three types of CCJM, but SS burdens were uniformly increased in all patients with accompanying syringomyelia.

**CONCLUSION**

For the three types of CCJM, swallowing difficulties and nystagmus were more prevalent in BI versus CM, while all CCJM patients with syringomyelia had higher frequencies of weakness, extremity numbness, neck pain, dissociated sensory loss, and atrophy.

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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Bindal AK, Dunsker SB, Tew JM Jr. Chiari I malformation: Classification and management. Neurosurgery 1995;37:1069-74.
2. Botelho RV, Diniz JM. Basilar Invagination: Cranio-cervical kyphosis rather than prolapse from the upper cervical spine. J Neurol Neuromed 2017;2:15-9.
3. Ferreira JA, Botelho RV. The odontoid process invagination in normal subjects, chiari malformation and basilar invagination patients: Pathophysiology correlations with angular craniometry. Surg Neurol Int 2015;6:118.
4. Goel A. Basilar invagination, chiari malformation, syringomyelia: A review. Neurol India 2009;57:235-46.
5. Guerreiro RB, Bittencourt L, Reis RC, Rotta JM, Tufik S, Botelho RV, et al. Upper airway dimensions in patients with cranio-cervical junction malformations with and without sleep apnea. A pilot case-control study. Arq Neuropsiquiatr 2015;73:336-41.
6. Klekamp J. Chiari I malformation with and without basilar impression: A comparative study. Neurosurg Focus 2015;38:E12.
7. Pinter NK, McVig J, Mechler L. Basilar invagination, basilar impression, and platybasia: Clinical and imaging aspects. Curr Pain Headache Rep 2016;20:49.
8. Shuhui G, Jiagang L, Haifeng C, Hao ZB, Qing HS. Surgical management of adult reducible atlantoaxial dislocation, basilar invagination and chiari malformation with syringomyelia. Turk Neurosurg 2016;26:615-21.

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