Management of Basilar Invagination

Tratamento de Invaginação Basilar

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RESUMO

A invaginação basilar (IB) constitui-se de uma anomalia do desenvolvimento da região crânio-cervical que resulta no prolapso da coluna cervical superior na base do crânio, comumente associada com outras anormalidades do neuro-eixo, tais como malformação de Chiari do tipo I e siringomielia. Neste artigo, revisamos os conceitos necessários para entender e tratar os pacientes com IB. O tratamento é discutido com base na classificação proposta por Goel, que divide a IB em dois grupos: grupo A - pacientes com elementos de instabilidade na junção crânio-cervical e grupo B - pacientes com IB secundária à hipoplasia do clivus. O tratamento no grupo A consiste no realinhamento e na estabilização da junção crânio-cervical, muitas vezes através de uma abordagem por via posterior isolada, evitando a morbidade inerente às descompressões por via anterior. No grupo B, a descompressão do forame magno é o tratamento de escolha. As técnicas cirúrgicas a serem utilizadas dependem da anatomia do paciente e da experiência do cirurgião. Resultados cirúrgicos adequados podem ser obtidos com o entendimento dos conceitos e formas de tratamento das diferentes apresentações da IB.

Palavras Chave: invaginação basilar, classificação, tratamento.

ABSTRACT

Basilar invagination (BI) is a development anomaly of the craniocervical junction that results in a prolapsed of the upper cervical spine into the skull base, commonly associated to other bone and neural axis abnormalities, like Chiari I malformation and syringomyelia. In this paper, we review the concepts necessary to understand and treat BI. The most comprehensive and accepted classification system is the proposed by Goel, which divides patients with BI into two groups, as it follows: group A) patients with clear elements of instability; and group B) BI secondary to clivus hypoplasia. Treatment in group A includes craniocervical realignment and stabilization, most of the times using an isolated posterior approach, obviating an unnecessary and morbidity of the anterior decompression. In group B, foramen magnum decompression is the treatment of choice. Surgical techniques should be adequate according to patient’s anatomy and surgeon’s experience. Good surgical results can be obtained with the understanding of the main concepts and treatment options of BI.

Keywords: basilar invagination, classification, diagnosis, surgical treatment.

INTRODUCTION

Basilar invagination (BI) is a radiographic finding that consists in a development anomaly of the craniocervical junction in which the odontoid process is at least 2 mm above Chamberlain’s line (although up to 6.6 mm has been advocated as the normal limit), resulting in a prolapse of the upper cervical spine into the skull base6,10,24. Bone anomalies can cause or be associated with BI. The most commonly findings are clivus hypoplasia, condyle hypoplasia, anomalies of the atlas formation and atlanto-occipital assimilation24,25,26. In addition, about 30% of the patients with BI have also neural axis abnormalities, like Chiari malformation, syringomyelia and hydrocephalus10.

In this paper, we review the basic concepts necessary to understand and treat BI based on its classification and the different surgical strategies adopted to treat this challenge and complex anomaly.

MATERIAL AND METHODS

A systematic review of the literature using the MedLine Database (National Library of Medicine), covering the period from 1980 to 2012, was performed. The search strategy involved the keywords “basilar invagination treatment”. Inclusion criteria included English language (or translated text), diagnosis of basilar invagination, discussion about management and surgical treatment of this entity. No age

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restrictions were included. All case series, clinical studies and review articles were also included. In addition, a manual search of all referenced articles not found in the search was performed. 315 articles were initially found. Cross references of classic historical articles were also included. We excluded some articles based on repeated information for the purpose of our review. After all abstracts were screened, 31 articles were evaluated and included in our review.

**Basic craniocervical craniometry (figure 1)**

Some basic craniocervical craniometrical concepts are necessary to diagnosis, understand and treat BI. A brief review of the main craniometrical points is exposed, as it follows:

Basion – it refers to the anterior margin of the foramen magnum.

Opisthion – it refers to the posterior margin of the foramen magnum.

McRae line – defines the plane of the foramen magnum. It is formed by a line from the basion to the opisthion. The tip of the odontoid process should fall below this line.

Platybasia – this term refers to a flattening of the skull base. It can be associated to BI but also may present in isolation. The diagnosis of platybasia is made based on the measurement of the Welcher Basal Angle. This angle is defined by a line extended from the nasium to the anterior tubercullum sellae and other from this last to the basion. In normal subjects, it should be less than 140°. Patients with increasing in this angle had the diagnosis of platybasia.

Wackenheim Clivus Baseline or Basilar line – this is a line along the clivus extending inferiorly into the upper cervical spine canal. It should fall tangent to the posterior aspect of the tip of the odontoid process. The angle formed by the intersection of the line constructed along the posterior surface of the axis body and the Wackenheim clivus baseline is called as craniovertebral or clivus-canal angle. It normally ranges from 150° to 180°, changing about 30° with flexion and extension positions. Craniovertebral kyphosis can be attributed when the clivus-canal angle is less than 150°.

**Craniocervical junction – anatomic anomalies of clinical importance in the treatment of bi**

Some anatomic development anomalies are associated with BI. The most important are presented below:

Atlantooccipital Assimilation - assimilation of the atlas is caused by a failure of segmentation between the skull and the first cervical vertebra, completely or partially. Although the Wackenheim clivus baseline may be normal, the clivus-canal angle may be decreased. It’s commonly associated with fusion of the axis and third cervical vertebra. This association (atlantooccipital assimilation and axis-C3 fusion) is associated with atlantoaxial subluxation in about 50% of the cases.

Basilar impression – it refers to the acquired form of BI secondary to softening of the bone and joints at the base of the skull. The main etiologies include rheumatoid arthritis, Paget disease, infection, tumors, among others. This term was proposed by Virchow, in 1876, 19 years later of the introduction of the term platybasia.

Basiocciput (clivus) Hypoplasia – represents the hypoplasia of the basiocciput that results in shortening of the clivus and BI (violation of the Chamberlain line by the tip of the odontoid), decreasing the clivus-canal angle and producing cranio-cervical kyphosis, resulting in deformity at the cervicomedullary junction.

Basion – it refers to the anterior margin of the foramen magnum.
magnum\\textsuperscript{24}.

Condylar Hypoplasia – The underdevelopment of the occipital condyles, with a flattened appearance, leads to basilar invagination, secondary to violation of the Chamberlain line and widening of the atlantooccipital joint axis angle. The tip of the odontoid process and the lateral masses of the atlas lie below the McRae line. The lateral mass of the atlas may be fused to the hypoplastic condyles, accentuating the BI and limiting the movements of the atlantoccipital joint\\textsuperscript{24}.

Chiari Type I Malformation – a caudal rhombencephalon abnormality that it is represented by a downward displacement of the cerebellar tonsils and the medial portions of the inferior cerebellar lobes into the cervical spinal canal\\textsuperscript{7,8}. Symptoms are secondary to valve effect by the protruding tonsils at the foramen magnum or direct compression of nervous tissue.

CLASSIFICATION OF BI

Goel proposed a classification of BI based on the presence or absence of clinical and radiological instability\\textsuperscript{12}. This classification is important once it can help surgeons in to decide the best form of surgical treatment:

Group A – This group main characteristic is the presence of a clear instability of the region manifested by the tip of the odontoid process distancing itself from the anterior arch of the atlas or the lower end of the clivus. A case example of BI of this group is presented in Figure 2 and 4.

Diagnostic criteria:
- atlantoaxial dislocation (generally with C1-2 subluxation)
- tip of the odontoid process protruded into the foramen magnum and above the Chamberlain line, McRae line and Wackenheim’s clival line

Patients may have also Chiari malformation with tonsilar herniation and syringomyelia.

CLINICAL AND RADIOLOGICAL EVALUATION

Patients can present with symptoms of BI at all ages, but most commonly at the second and third decade of life. Some clinical signs can be clearly seen at the first inspection, like torticollis, limited neck movements, low hairline implantation and short neck. Most of the patients present with neurological deficits, like muscular weakness, gait abnormalities, posterior column and bladder dysfunction, or even low cranial nerves deficits\\textsuperscript{14,23}. Sometimes, only neck pain is reported. Symptoms onset can be acute, following a minor trauma, but they are generally chronic and progressive\\textsuperscript{23,30}.

Radiological evaluation includes Computer Tomography (CT) scan with sagittal and coronal reconstructions to evaluate the bone anatomy and cranio cervical Magnetic Resonance (MR) imaging, to evaluate the neural axis abnormalities, like Chiari malformation and the presence of syringomyelia. In some cases, when clear instability cannot be inferred in static radiological exams, at least one dynamic study should be performed in patient evaluation, such as a sagittal flexion and extension CT scan or MR at the T2 weighted sequence.

Surgical treatment is indicated in symptomatic patients once BI is a progressive disease that can lead to severe disability or even death. In asymptomatic patients, close clinical and radiological follow-up is necessary, surgery may be an option in presence of mild symptoms, like cervical pain, or pyramidal signs or presence of syrinx.
Group B – There is no instability. The alignment of the anterior arch of the atlas, the odontoid process and the inferior aspect of the clivus remains normal. Probably, this condition is secondary to a complete clivus hypoplasia. Platybasia can be found in both groups, but it occurs most commonly in group B. Figure 3 shows a case example of a BI in this group.

Diagnostic criteria:
- atlantoaxial relation is normal,
- tip of the odontoid process is above the Chamberlain’s line,
- tip of the odontoid is below the Wackenheim’s clival line and the McRae’s line.

The distance between the ponto-medullary junction and the odontoid process is markedly reduced. Posterior cranial fossa volume can be also markedly reduced in this group. Chiari I malformation is commonly found.

**SURGICAL TREATMENT**

Surgical treatment is proposed based on the classification of BI.

**Group A**

Realignment of the craniocervical junction results in indirect neural decompression. Goel, in 2004, proposed an innovative posterior approach with C1-2 joint distraction and direct lateral mass fixation involving placement of spacers in the joints to reduce odontoid migration and reestablish craniocervical alignment. Although technically challenging, this is the only surgical option that treats C1-2 instability directly, being considered the gold standard of treatment in patients of group A. This technique requires a long learning curve, with expertise in C1 - C2 fixation using screws, once most of these patients have vertebral artery anomalies and small lateral masses or atlas assimilation. As an alternative, other surgical options can be used. One of these options is preoperative traction and, if reduction is obtained, an occipitocervical fusion can be performed. Although occipital fixation with screws is preferred, sometimes the occipital bone is thin, precluding screw insertion. In these cases, wiring techniques are then preferred, even though a less rigid construction is obtained.

Jian et al., 2010, published a series of 27 out of 28 patients...
successfully treated with direct intraoperative posterior occipito-C2 pedicle screws reduction with distraction and fixation of BI with atlantoaxial dislocation. After tightening the rod in the occipital bone, distraction is performed, moving the odontoid inferiorly, and then inserting C2 rod holder17. It can be an option in patients whose anatomy precludes C1 screw insertion or in cases without reduction of C1-2 subluxation with preoperative traction. The potential disadvantages of this technique, when compared with C1-2 distraction and fixation, is that it can leads to craniocervical fixation in flexion and an unnecessary occipital fusion in patients without atlas assimilation.

Nowadays, anterior approaches are rarely used, being indicate only for patients with irreducible ventral compression of the brainstem. Patients treated in other institutions with previous occipito-cervical fusion without reduction of C1-2 subluxation may have some benefits from an anterior decompression. A direct transoral-transpharyngeal approach is the most common form of anterior decompression18,20. Endoscopic transnasal, transoral and even transcervical approaches have been also proposed, followed by a posterior occipito-cervical fusion in selected cases, with or without an additional posterior decompression2,31.

Summary of treatment in BI - Group A – surgical options

Treatment of Choice:
1) C1-2 distraction and manipulation of the joints using spacers.

Treatment Options:
1) Preoperative traction followed by occipito-Cervical fusion if reduction was obtained or;
2) Direct intraoperative distraction using occipito-C2 pedicle instrumentation.

Anterior approaches: irreducible C1-2 subluxation with ventral compression or previously fixed patients without reduction.

Group B

Treatment in this group is mainly based on posterior foramen magnum decompression12. Some authors advocate a small suboccipital craniectomy, to avoid postoperative cerebellar herniation5,6. However, a large craniectomy has been proposed by others reshaping the small volume of the posterior fossa22.

Opening of the dura mater and coagulation of the tonsils even in presence of a syrinx, is controversially. Many authors advocate that these maneuvers lead to an unnecessary risk of complications such as CSF leak, meningitis and infection14. Others advocate radical tonsil coagulation and duraplasty to improve the neural decompression and increase posterior fossa volume1,4,15.

Preoperative traction is not helpful in changing craniocervical parameters of patients in this group12,14. This can be explained by the fact that there is no instability in these patients with clivus hypoplasia.

Failure in obtain clinical improvement after posterior fossa decompression have been reported5,11. Worsening of craniocervical kyphosis can exacerbate cervicomедullary compression over the odontoid16. In patients with an important kyphotic clivus-canal angle (less than 130°), the brainstem can suffers severe deformatives stresses, resulting in clinical deterioration16. Once traction is not useful in to restore craniometrical parameters, these patients may have clinical benefits with craniocervical fusion in mild extension, improving clivus-canal angle in up to 30° to decrease brainstem deformity and stress16,24.

Summary of treatment in BI - Group B – surgical options

Treatment of Choice:
1) Posterior foramen magnum decompression.

Options: small versus large craniectomy/ duraplasty/ tonsil coagulation

(Preoperative traction is not useful in this group)

Treatment Option:
1) Craniocervical fusion in mild extension in patients with clinical deterioration after posterior fossa decompression or severe kyphosis at the craniocervical junction (small clivus-canal angle).
Treatment of BI is guided according to the presence (Group A) or absence (Group B) of C1-2 instability. Surgery can improve patient symptoms and restore quality of life. Understand the different types of BI, as well as the associated neural axis abnormalities are necessary to optimize surgical results. Moreover, a correct radiological evaluation of bone and neural anatomy allows surgeons to choose the best surgical technique, improving patient outcome.

Table 1: Craniometrical Parameters of the Craniovertebral Junction

<table>
<thead>
<tr>
<th>Chamberlain Line</th>
<th>Extends from the posterior portion of the hard palate to the opisthion. The odontoid tip should be located below this line in normal subjects (or until 2 mm above it)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Welcher Basal Angle</td>
<td>Defined by a line extended from the nasium to the anterior tuberculum sellae and other from this last to the basion. Should be less than 140o. Increasing in this angle leads to the diagnosis of platybasia</td>
</tr>
<tr>
<td>McRae line</td>
<td>A line from the basion to the opisthion. Defines the plan of the foramen magnum</td>
</tr>
<tr>
<td>Wackenheim Clivus Baseline or Basilar line</td>
<td>A line along the clivus that extending it inferiorly into the upper cervical spine canal. It should be fall tangent to the posterior aspect of the tip of the odontoid process</td>
</tr>
<tr>
<td>Craniovertebral or clivus-canal angle</td>
<td>Angle formed by the intersection of the line constructed along the posterior surface of the body and the Wackenheim clivus baseline. It normally ranges from 150o-180o</td>
</tr>
</tbody>
</table>

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craniocele junction.” Childs Nerv Syst 2008;24(10):1187-93.


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Consents were obtained from the patient presented in this paper.
Financial Disclosure: The authors have no financial interest in this article.
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