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IMAGING (L MECHTLER, SECTION EDITOR)

Basilar Invagination, Basilar Impression, and Platybasia: Clinical and Imaging Aspects

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Abstract The congenital and acquired deformities of the craniovertebral junction (CVJ), such as basilar invagination, basilar impression, or platybasia, can present in the form of slowly progressive or acute neurologic deterioration. In many cases, an insidious headache is the only symptom and can be a diagnostic challenge for the neurologist. Proper imaging studies as well as recognizing often associated neurologic or systemic conditions are required for early diagnosis and effective therapy. In the current report, the primary focus will be on clinical aspects of these CVJ abnormalities; the pathologic and radiologic aspects, such as developmental and pathophysiologic background or radiographic analysis, will be discussed briefly, confined to clinically relevant data.

Keywords Basilar impression · Platybasia · MRI · Chiari malformation · Craniovertebral junction · Osteogenesis imperfecta

This article is part of the Topical Collection on *Imaging*

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Introduction

Basilar invagination and basilar impression are congenital and acquired forms of the craniovertebral junction (CVJ) anomalies, where the odontoid process of C2 prolapses into the foramen magnum. Basilar invagination can be a solitary finding resulting from a number of developmental anomalies, or it can be associated with a more complex developmental process such as Chiari malformation. Basilar impression occurs secondary by definition and involves the abnormal softening of the bone. One specific form is cranial settling, a term reserved for deformation of CVJ caused by rheumatoid arthritis. Prolapse of odontoid process in addition to anomalies of the CVJ may lead to the compression of the cervicomedullary junction, adjacent vascular structures, or cerebrospinal fluid spaces and may cause muscle spasm. This can result in a wide spectrum of neurologic symptoms, differing in acuity and severity depending on the clinical presentation. Basilar invagination, basilar impression, cranial settling, and platybasia are often synonymous in the literature, but it is important to distinguish these entities. Platybasia is the flattening of the skull base and may be a solitary finding but is more likely associated with other forms of skull base or CVJ deformity.

Basilar Invagination, Basilar Impression, and Platybasia: Definition and Associated Pathologies

Basilar invagination is a developmental anomaly of the CVJ where the odontoid process is positioned abnormally upward and backward, prolapsing into the foramen magnum [1]. It is often a result of CVJ developmental disorders, where the bone tissue remains normal, such as basioccipital (clivus) hypoplasia, atlas hypoplasia, occipital condyle hypoplasia, achondroplasia, or an incomplete ring of C1 with spreading of the lateral masses and atlanto-occipital assimilation. Chiari malformation is often associated with basilar invagination and

other CVJ abnormalities and may be present in up to 33–38 % of patients [2]. In a recent retrospective study of patients with Chiari malformation who underwent surgery, 13 % (46 out of 350) had associated basilar invagination, and 67 % (31 out of 46) of those patients had a ventral compression in the foramen magnum [3]. Occipital condyle hypoplasia results in decreased skull base height with basilar invagination. Due to flattening of the occipital condyles, the skull base may actually ascend rather than descend. Patients with achondroplasia have a small foramen magnum, shortened clivus, and abnormal odontoid process with resulting invagination and possible medullary compression [4]. Atlanto-occipital assimilation, also known as occipitalization of the atlas, is the most common developmental abnormality of the CVJ. It can be associated with a variety of congenital anomalies, which may be present in approximately 20 % of patients. This results in limited mobility at the atlanto-occipital junction and C1–2 instability in 50 % of patients. In Klippel–Feil syndrome, many CVJ anomalies are frequently present including basilar invagination, odontoid hypoplasia, atlanto-occipital assimilation, platybasia, and the Chiari type I malformation [5••].

In the case of *basilar impression*, the same malpositioning of the odontoid process is secondary to traumatic changes or bone softening in diseases such as Paget disease, osteomalacia, severe osteoporosis, osteogenesis imperfecta, rickets, hyperparathyroidism, renal osteodystrophy, skull base infection, or Hurler syndrome. It is also referred to as "secondary basilar invagination" [6–8]. Rheumatoid arthritis results in basilar impression due to the loss of axial supporting structures in the upper cervical spine, and it is estimated that 10 % of affected patients are consequently at risk for sudden death due to instability [9•]. Basilar impression was reported at a frequency of 25 % in a clinical screening study that included 87 patients with osteogenesis imperfecta [10]. Basilar impression was found at the highest frequency in type IV B, and 50 % of these patients had neurologic signs of compression of posterior fossa structures. In another study, basilar impression was found in 17 % of 47 patients who had type III osteogenesis imperfecta (Table 1).

Table 1 Most common causes of primary and secondary basilar invagination

Basilar invagination	Basilar impression
Chiari malformation	Paget disease
Basioccipital hypoplasia	Osteomalacia
Atlas hypoplasia	Severe osteoporosis
Occipital condyle hypoplasia	Osteogenesis imperfecta
Achondroplasia	Rickets
Incomplete ring of C1	Hyperparathyroidism
Atlanto-occipital assimilation	Renal osteodystrophy
Fused upper vertebrae	Skull base infection
	Hurler syndrome

Platybasia is characterized as flattening of the skull base determined by lines drawn from the nasion to the tuberculum and the basion to the tuberculum. This abnormality is often associated with basilar impression and basilar invagination. Platybasia alone does not usually cause symptoms unless it is associated with basilar invagination [11].

Craniometric Measurements to Define Basilar Invagination and Platybasia

Basilar Invagination

The three reference lines traditionally and widely used in diagnosing basilar invagination are the Chamberlain, the McGregor, and the McRae lines [12]. These methods of measurements have good specificity and sensitivity and are easily reproducible, granting them clinical importance. All three lines were established on lateral radiographs of the skull.

The Chamberlain line runs from the hard palate to the opisthion, which is the midpoint of the posterior margin of the foramen magnum (Fig. 1). The tip of the odontoid process typically lies inferior to this line—1.8 mm in men and 1 mm in women, on average, according to a study by Cronin et al. [9•]. It is generally considered to be normal if the dens extends above this line, not more than 2.5 mm, although the range varies based on the source. Basilar invagination is considered if the odontoid tip extends more than 5 mm above the Chamberlain line [1, 6, 13].

Since the precise identification of the opisthion on lateral radiographs is not always possible, McGregor proposed a

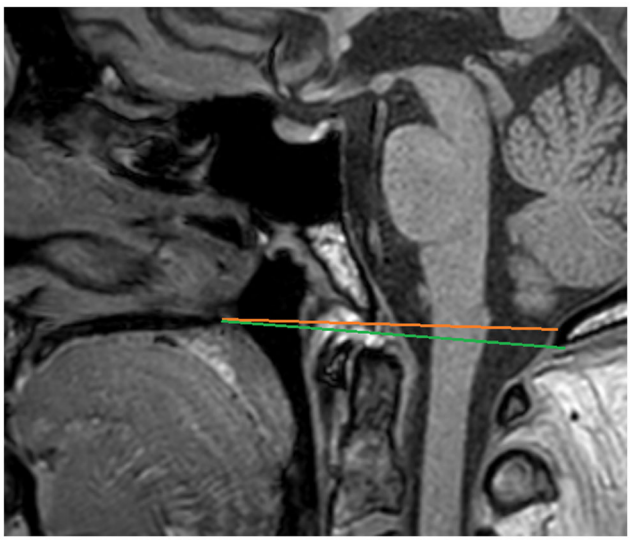

Fig. 1 Normal craniovertebral junction on a sagittal T1-weighted image. The Chamberlain line (*brown*) runs form the hard palate to the opisthion. The McGregor line (*green*) runs from the hard palate to the lowest point of occipital squama. The tip of the odontoid process typically lies below these lines in normal cases

modification of this line [5••]. The McGregor line runs from the hard palate to the lowest point of occipital squama (Fig. 1). This line is 2 mm inferior to the Chamberlain line. Normally, the tip of the dens extends no more than 4.5 mm above this line and is generally considered abnormal if the tip of the odontoid projects greater than 7 mm above the McGregor line (Fig. 2) [13].

The McRae line is defined by the line from the basion (the midpoint of the anterior margin of foramen magnum) to the opisthion and is basically the anteroposterior diameter of foramen magnum measured in the midline [1]. Normally, the dens should be below this line; in healthy adults, the average distance between the tip of dens and the McRae line was found to be 4.6 mm in women and 5.3 mm in men [9•].

The Wackenheim line (or Wackenheim's clivus baseline) is drawn along the posterior surface of the clivus and extends to the cervical spinal canal. The dens should lie immediately anterior to it. Various sources define pathologic conditions differently. According to Smoker, an intersection of the Wackenheim line with 1/3 of the odontoid process is still normal, while Ross considers any intersection with the tip of the dens abnormal [6, 13]. The intersection of the Wackenheim line and the body or base of C2 suggests anterior craniocervical dislocation, which most likely results from trauma.

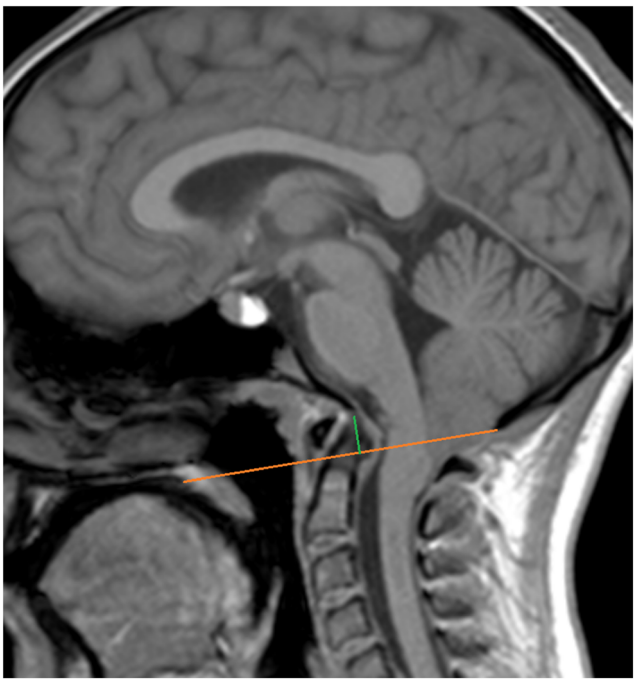

Fig. 2 Basilar invagination on a sagittal T1-weighted image. The tip of the odontoid process extends 8 mm above the McGregor line (*brown*). The tip of the odontoid prolapses backward and causes the compression of the cervicomedullary junction. Magnetic resonance imaging (MRI) allows superior visualization of the brainstem and spinal cord compared to computer tomography and should be performed to rule out secondary parenchymal changes

Basilar invagination can be caused by condylar hypoplasia, which can be measured with the atlanto-occipital joint angle. This angle is constructed at the intersection of tangents drawn parallel to the atlantooccipital joints on the coronal view of the skull. The average angle measures 124° to 127°; it becomes more obtuse with occipital condylar hypoplasia and may even approach 180° or more [5••].

There are other craniometric measurements and reference lines used for the diagnosis of basilar invagination, such as the Klaus height index, Bull angle, Redlund-Johnell method, Ranawat method, Fischgold bimastoid line, Fischgold digastric line, Clarks stations, and the Francesconi invagination index [13]. Many of these measurements suffer from poor sensitivity and specificity and therefore, are of less clinical importance.

Platybasia

Platybasia by definition is the flattening of the skull base as measured by the Welcher-basal angle. This angle is formed at the intersection of lines drawn from nasion to tuberculum sella and basion to tuberculum. It is 132° on average and considered normal when less than 140° (Fig. 3). Platybasia is defined by a Welcher basal angle greater than 140°. Most patients with basilar invagination have a normal basal angle and do not have associated platybasia [11, 13]. In the case of platybasia, the position of the Wackenheim clivus baseline is usually normal.

The clivus-canal angle (also craniovertebral angle or Wackenheim-canal angle) is measured at the intersection of

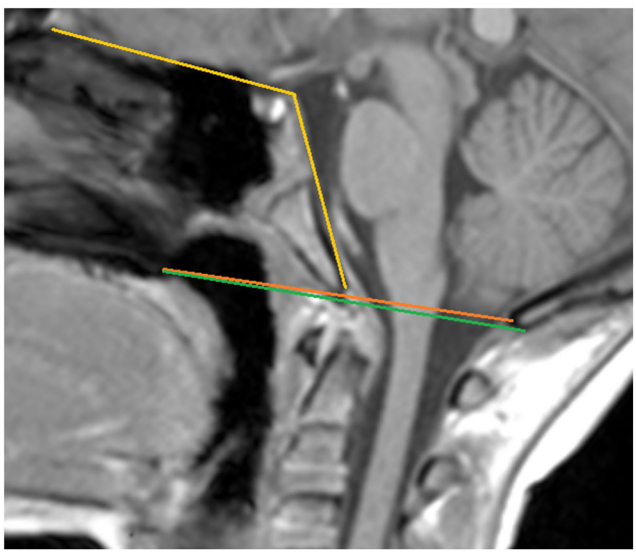

Fig. 3 Demonstration of the three main measurements of the craniovertebral junction on a T1-weighted image of a pediatric patient. The distance between the Chamberlain (*brown*) and the McGregor (*green*) lines is small. The Welcher-basal angle (*yellow*) is formed by a line running from the nasion to the tuberculum sella and a second line running from the tuberculum along the posterior margin of the clivus. A Welcher-basal angle larger then 140° indicates platybasia

Wackenheim line (drawn along the clivus) and a line along the posterior margin of C2–3 bodies. It ranges normally between 150° in flexion and 180° in extension. Ventral spinal cord compression may occur with angles less than 150°. In the case of platybasia, the Chamberlain line and Wackenheim clivus baseline may not be violated; however, the clivus-canal angle may become abnormal and cause the compression of the cervicomedullary junction [6].

Imaging of Basilar Invagination and Platybasia

Conventional Imaging

The Chamberlain and McGregor lines were first established on plain lateral radiography of the cervical spine over 50 years ago [9•]. However, for optimal assessment of the CVJ, the study should include all bony structures between the sphenoid base and the C2 base. The tuberculum sella, in the case of a cervical spine study, or the C2 body in the case of a skull radiograph, may be excluded from the field of view; the adjacent osseous structures may obscure the region of interest. This, in general, makes the X-ray less favorable against computed tomography (CT) or magnetic resonance imaging (MRI) in the study of the CVJ [5••]. With the advent of high-resolution 3D imaging, it became possible to create orthogonal reconstructions of one 3D dataset, which allowed adapting radiographic skull measurements to CT. CT is the best tool for imaging bone and should always be the first choice for posttraumatic changes, evaluation of the osseous structures, visualization of the relation of different bony elements, or imaging of soft tissue calcifications. Imaging from CT may assist in deciding whether the CVJ deformity is congenital or acquired, by differentiating between normal and abnormal bone (Fig. 4).

MRI is a multiplane acquisition technique, and thus, the mentioned measurements can be demonstrated on 2D sagittal images, although 3D techniques are also available and widely used in routine imaging of the skull.

The advantage of the MRI in the region of CVJ is to visualize subtle changes in the soft tissue and cervicomedullary junction. It is recommended that the connective tissues of the CVJ as well as their pathologic changes be studied exclusively by MRI, unless the degree of soft tissue calcification is in question. Visualization of the ligaments, membranes, and joints of the CVJ is still not ideal with MRI, but far superior as compared to CT. The normal ligaments appear as dark bands on standard T2 spin-echo sequences, and these uniformly dark structures are often hard to differentiate, but once they are damaged, they show increased signal intensity or irregularity, which may be appreciated only on MRI [14]. Similarly, when there are clinical or imaging signs of brainstem or upper spinal cord compression, MRI is the

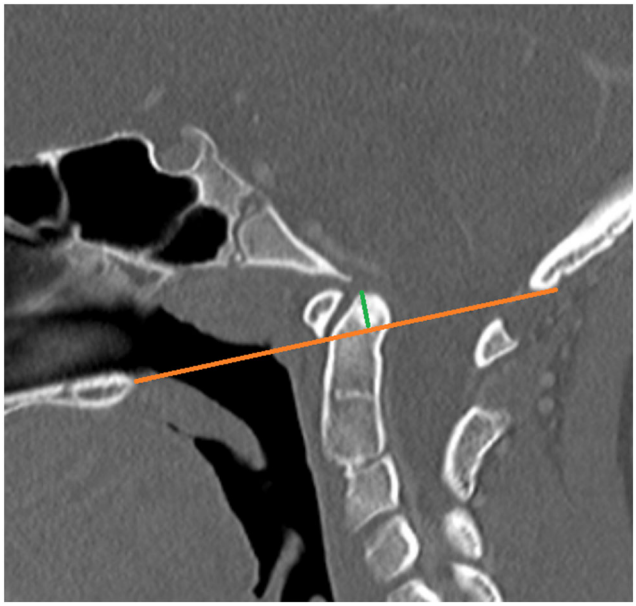

Fig. 4 Basilar invagination as demonstrated by a sagittal reconstruction of a CT scan. The thin slice, isotropic acquisition, and the applied bone filter allow the excellent visualization of the bony structures and more precise measurements. The green line indicates that the tip of the odontoid process extends 6.6 mm above the McGregor line (brown)

method of choice. MRI is also superior in the imaging of bone edema; however, in visualizing the bone structure, it generally performs worse than CT. Also, although both CT and MRI images can suffer from artifacts caused by metallic objects in postoperative studies, MRI may be more susceptible to these due to field inhomogeneities and frequency offsets, and CT can be more reliable. For this reason, the two modalities have complementary roles in the imaging of CVJ, and the application of both may be justified in certain clinical settings.

The standard reference lines and measurements can be performed on CT and MR similar to the way they were originally established on plain film more than 50 years ago. However, there are differences and modality-specific variations. In case of MR, it can be difficult to define the edge of the hard palate on sagittal T1 images, and one should look for the cortical bone dark signal on T1-weighted images. To define basilar invagination, it is recommended that one measure the shortest perpendicular distance from the given line to the tip of the odontoid [9•]. The thickness (or apparent thickness) of the hard palate can pose a technical problem as well. In most papers, the posterior end of the hard palate is treated as a point, and in many published images, it is indeed a point; however, in the daily routine on MR images, in many cases, it seems to have a thickness comparable to that of the occipital squama [5••]. In some patients, this may raise the possibility of significantly affecting the measurement results. Most reports do not mention this problem, and it may not have a clinical impact; however, it is important to utilize a method that is clinically reliable and reproducible. Cronin et al. used the postero-

superior margin of the hard palate in defining McGregor s line. Some measurements also can differ in their normal range in CT and MR, compared to X-ray [9•].

Dynamic Imaging of the Craniovertebral Junction—Imaging of Instability

The dynamic X-ray, CT, or MR imaging can demonstrate craniocervical instability and may be the only method to visualize functional stenosis of the spinal canal or cord compression. Recently, dynamic CT and MR have been used to confirm the instability of C1-C2 in patients with os odontoideum [15] and to detect atlantoaxial instability with cord compression where it was not seen in neutral position [16] in various abnormalities of CVJ and to compare the CVJ region of healthy subjects and patients with rheumatoid arthritis. In a study by Asri et al., sequences of CT in flexion and extension have been used to depict the degree of abnormal motion between C1 and C2 in patients with previously confirmed os odontoideum. A dynamic MR of the region in cine mode may be more efficient, since it provides real-time images and holds more information about soft tissues and the spinal cord. The main goal with these sequences should be to demonstrate the functional abnormalities, such as instability or position-dependent compression of the brainstem; therefore, other quality issues can be secondary. A lower resolution scan with motion artifacts and with a field-of-view that covers only the CVJ area, but not the rest of the skull, is acceptable.

In a study by Gupta et al., sagittal and axial T1 and T2 sequences were applied in neutral position as well as in flexion and extension. Although this was not cine-type real-time imaging of the moving spine, it facilitated the diagnosis of atlantoaxial instability, functional spinal canal narrowing, and variability in cord compression with changing position of the neck. Dynamic MRI was able to detect cases of cord compression that were not seen in neutral position. CT of the neck was also performed in 80 % of the patients, in supine position after flexion and extension with 2-mm thin slices and was also definitive. The functional changes in the spinal canal and atlantoaxial displacement were studied by Reijnierse et al. and Karhu et al. [17, 18]. It was reported that on flexion, there is statistically significant narrowing of the subarachnoid space at the atlantoaxial level and below C2. Karhu et al. studied 31 patients with rheumatoid arthritis and 20 healthy subjects in an open MRI scanner during flexion and extension. Additionally, the MRI study was compared with X-ray in patients with rheumatoid arthritis. They found that in the neutral position, C1 was oriented normally, but in a flexed position in relation to both the occiput and C2. The image was abnormal compared to healthy subjects. The atlantoaxial subluxation was found to be greater on lateral radiographs than in MRI in flexion. In the patients compared to healthy subjects, there was considerably more cord impingement in flexion than in other positions.

Symptoms Associated with Basilar Invagination/Impression and Platybasia

Due to the multitude of causes resulting in basilar invagination or impression, the symptomatology is very diverse. The symptoms caused solely by the disturbed mechanics of the CVJ region can mix with the symptoms resulting from the compression of the medulla, the cervicomedullary junction, or the upper cervical spinal cord. Obstruction of the CSF circulation may be present as well, leading to syringomyelia. The direct CNS symptoms may include signs of medullary dysfunction, such as nystagmus, dysphagia, ataxia, dysmetria, or cranial nerve palsy. There may be signs of myelopathy, such as motor and sensory abnormalities or vegetative dysfunctions. The clinical picture may be altered according to the underlying or accompanying pathologic process.

When CNS compression is not present, the mechanical effects of abnormal dens position may be responsible for symptoms. Exertional and symptomatic cough headaches seem to be the most frequently associated to basilar invagination or impression. It is almost exclusively described as an occipital or suboccipital headache [19, 20, 21••]. Pascual and colleagues analyzed 72 cases of benign and symptomatic cough-, exercise-induced (formerly exertional), and sexual headaches [20]. These headache forms are different clinical entities with different age of onset, clinical course, or treatment response, although similar precipitant factors. All of the patients with symptomatic cough headache had Chiari I malformation, based on CT and MR images. These patients had occipital and suboccipital pain, in five cases with frontotemporal radiation. Basilar invagination was not reported, although was not specifically mentioned either. No CVJ abnormalities were found related to benign cough headache, benign exercise-induced headache, or benign sexual headache; however, CT or MR was performed only in 11 out of 13, 6 out of 15, and 8 out of 13 patients in these groups, respectively; therefore, it cannot be excluded that CVJ abnormality was present in some of these cases. Symptomatic cough headache can be caused by basilar invagination or impression among other hindbrain abnormalities, such as meningioma, Schwannoma, or posterior fossa cyst. It can be precipitated by laughing, weight lifting, or acute body or head postural changes in addition of coughing [19]. In another study by Pascual and colleagues, 97 patients were evaluated for cough, exertional, or sexual headache, and 45 % were found to have an underlying intracranial abnormality, and 80 % of secondary cough headaches were related to Chiari I [21••]. Cutrer mentions basilar impression and platybasia as possible causes of benign cough headache.

The clinical course and symptoms may be different when basilar invagination is associated with Chiari malformation. Goel et al. reported on 190 surgically treated patients with basilar invagination stratified based on presence or absence of associated Chiari malformation. Among those patients who had invagination without Chiari, symptom onset was relatively acute, and the most common signs and symptoms included weakness (100 %), neck pain (59 %), posterior column dysfunction (39 %), bowel and bladder disturbance (28 %), and paresthesia (25 %). Localized findings included torticollis (69 %), restricted neck movements (59 %), low hairline (48 %), webbed neck (47 %), and short neck (41 %).

In the 102 patients with basilar invagination with associated Chiari malformation, the duration of symptoms was relatively long-standing and slowly progressive, and the most common signs and symptoms included weakness (94 %), paresthesia (79 %), posterior column and spinothalamic tract disturbance (56 %), and ataxia (47 %). Common localized findings included short neck (50 %), webbed neck (38 %), and low hairline (37 %); however, headache or neck pain was not among the most common symptoms [1, 22].

Congenital skull base deformities can present as neck pain, which may be related to change of posture and resulting muscle spasm. In a recently reported case, basilar invagination was identified as a cause for recurrent torticollis and exertional headache. In this patient, no other CVJ abnormalities were diagnosed. This calls the attention to the possibility that exercise-induced headache with torticollis might be the only neurologic symptom of basilar invagination [23].

Other types of headache, such as migraine, are less likely associated with basilar invagination. In the study by Eidlitz-Markus, out of 600 total children with migraine who suffered from craniocervical and occipital pain, none of the subjects had basilar invagination [24].

Basilar impression is often associated with osteogenesis imperfecta. The clinical course is slowly progressive and has potentially serious complications, although patients may remain asymptomatic and imaging signs may be present for years before neurologic progression [10]. Sillence and colleagues found that basilar impression was radiologically present long before puberty [25]. Neurologic signs may be present before symptoms, which include nystagmus, facial spasms, cranial nerve paresis, pyramidal tract signs, proprioceptive deficits, and papilledema in cases of hydrocephalus. Neurologic symptoms that typically may develop later include occipital headache, which is worse with movement, coughing, sneezing, or straining; trigeminal neuralgia; imbalance; weakness in arms and legs; and bladder disorders [26].

Platybasia is thought to cause clinical symptoms when it is associated with other CVJ deformities, often basilar invagination or impression. Aside from the neurologic deficit related to compression, it will likely cause occipital headache. In the case of rheumatoid arthritis, chronic neck and occipital pain accompanied by the symptoms of brainstem compression may draw attention to CVJ abnormality [27].

Treatment Options

The definitive treatment for basilar invagination is decompression and stabilization. Surgery should be initiated in the cases where neurologic deterioration is present or foreseen, and the patient is expected to benefit from the procedure. In most cases, the indication is the compression of the cervicomedullary junction or upper cervical cord and the resulting symptoms. This may be caused by the prolapsing dens or CVJ instability; the latter may be functional as well. The preoperative examinations should therefore search for signs of compression on clinical exam and on imaging studies. It has been suggested that both CT and MRI should be performed. CT would reveal any pathology of bony skull base and CVJ region, such as posttraumatic deformations or changes that may result from systemic diseases. MRI, on the other hand, would disclose compression of the cervicomedullary junction and secondary changes, such as myelopathy, syringomyelia, or compressed and displaced cranial nerves. CT or MR angiography would visualize the status of neck and posterior fossa circulation, which may be affected as well. Dynamic studies should be considered in cases where conventional studies do not show direct compression, but secondary imaging signs or clinical signs of compression are present. In patients without neurologic symptoms where basilar invagination is an incidental finding, any therapeutic step should be considered carefully, for it may not be necessary. If some degree of cervicomedullary compression or functional instability is already present without secondary changes and clinical symptoms, or with mild neurologic deficit, a close follow-up and later therapeutic re-evaluation should be considered. The clinical preoperative assessment is also important and should include a thorough neurologic examination as well as the review of general systems, with an emphasis on oral hygiene, pulmonary status, and nutritional status.

Preoperative cervical traction can be performed. If it proves the basilar invagination to be reducible, it may be possible to treat with a posterior-only surgical approach, including decompression and fusion [1]. Traction may be the most beneficial when basilar invagination is not associated with Chiari I. Where basilar invagination is not reducible with traction, ventral decompression and posterior stabilization are performed. The ventral approach generally means a transoropharyngeal approach through which the anterior arch of C1 and the odontoid tip is removed. The anterior decompression is often followed by subsequent posterior decompression, but without an associated Chiari I, the ventral approach alone might be sufficient. When basilar invagination is associated with Chiari I malformation, both anterior and posterior approach may be necessary according to Goel et al. and

Mezenes et al. [22, 28]. In a recent review, Chaudry et al. proposed that an anterior approach should be performed in patients with high cervicomedullary compression and lack of C1-2 mobility.

Conclusion

Basilar invagination and impression will most likely cause exertional or symptomatic cough headaches in the occipital or suboccipital location. Besides the abnormal prolapse of odontoid process, symptomatic cough headache can result from a number of pathologies of the posterior fossa or CVJ, such as Chiari I malformation, brain tumor, meningioma, or Schwannoma. Therefore, MR or CT imaging of the head and CVJ in these cases is suggested, especially when it is a new onset headache. When basilar impression is associated with Chiari malformation, the clinical course is slowly progressive. Basilar impression without Chiari comes with a more acute clinical appearance. In patients who suffer from rheumatoid arthritis or diseases that can cause softening of the bone, symptoms of basilar impression may emerge during the course of the disease, which requires the clinician to pay close attention to the subtle changes of the clinical status. It is important to remember that occipital-suboccipital headache related to physical exertion may be the first or the only early sign of secondary craniovertebral instability.

Compliance with Ethical Standards

Conflict of Interest Nandor Pinter declares grant funding from the Dent Family Foundation.

Jennifer McVige declares no conflict of interest.

Laszlo Mechtler declares personal fees as speaker honoraria from Teva, Allergan, Depomed, and Pernix.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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- • Of importance
- •• Of major importance

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