**[Malformations of the cranio-cervical junction: basilar impression](https://ajbm.net/malformations-of-the-cranio-cervical-junction-basilar-impression/)**

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**Abstract**

Cranio-cervical junction abnormalities are a rare combination of congenital or acquired malformations, linked to the occipital-cervical region that often leads to severe neurological disorders. There is a great clinical polymorphism and high variability in the natural history of these conditions, which determines therapeutic outcomes difficult to assess in a global sense. The authors of this report give an account of the case of malformative bone complex of cranio-cervical junction and carry out a non-systematic search of the literature.

We analyze the clinical case of a 19-year-old woman who presents hypoesthesia in the four limbs, spastic paraparesis that began one year ago, and evolved to tetraparesis. On physical examination, there is quadriparesis, pronounced in the lower limbs, hyperreflexia, and Babinski sign bilaterally and the Hoffman sign. The hypothesis of myelopathy was made. X-ray suggested the possibility of a malformation of the cranio-cervical junction suggesting basilar impression with an odontoid process anomaly. Computed tomography, suspected myelopathy, demonstrates an anomaly of the cranio-cervical junction, basilar impression. MRI is performed of the cranio-cervical junction concluding a basilar impression with significant additional cord damage.

**Keywords**: Cranio-cervical malformations; Basilar impression; Platybasia; High cord compression

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**Introduction**

Anatomically, the cranio-cervical junction (CCJ) is composed of the lowermost region of the skull, or foramen magnum area, and the most superior segment of the cervical spine. This structure has evolved to address the dichotomy of the neurological processes stemming from the brain above the cranio-cervical junction with the spinal cord moving below. As a result of the impressive array of structures passing through so narrow a space, there are multiple important clinical and surgical repercussions in individuals with malformations of the cranio-cervical junction. Individuals with malformations of the cranio-cervical junction experience problems of impaired vascular physiology, compressed nerves, and disturbance in cerebrospinal fluid flow.

These, in turn, present as a host of neurological symptoms, many of which are potentially life-threatening. The purpose of this essay is to illustrate the malformations of the cranio-cervical junction that require surgical manipulation in order to return a patient to an acceptable state. We will demonstrate how these malformations can be categorized based on the gross anatomical defects, then contrast them with the symptoms typically presented and the physiological rationale for these symptoms. Case studies will then be presented regarding the three most common malformations experienced at the cranio-cervical junction - Chiari malformation, Basilar Invagination, and Atlanto-axial dislocation. These three malformations embody the main groups of defects experienced at the cranio-cervical junction and in whom only surgical intervention can hope to return the patients to their normal functional state.

**Clinical case**

We analyze the clinical case of a 19-year-old woman who presents hypoesthesia in the four limbs, spastic paraparesis that began one year ago, and evolved to tetraparesis. On physical examination, there is quadriparesis, pronounced in the lower limbs, hyperreflexia, and Babinski sign bilaterally and the Hoffman sign. The hypothesis of myelopathy was made. X-ray suggested the possibility of a malformation of the cranio-cervical junction suggesting basilar impression with an odontoid process anomaly. Computed tomography, suspected myelopathy, demonstrates an anomaly of the cranio-cervical junction, basilar impression. MRI is performed of the cranio-cervical junction concluding a basilar impression with significant additional cord damage.

This dorsal cord compression is due not only to the extrusion of the abnormal odontoid process in the canal, but also to malposition following the presence of this osteoarthritis in a frightened position. The bad position of this odontoid process is responsible for the multistructural very high cervical compression. The basilar impression is defined as the decrease of the distance between the F. of the McRae Maxillary base and the level of the posterior arch of the Atloid Process less than 35 mm, in the adult person. The interest of this clinical case lies in the exceptional occurrence of basilar impression. Basilar impression is a malformation of the cranio-cervical junction due to the embryological development of this anatomical region. During the sixth week of embryonic development, a chondrification center occurs (Meckel's cartilage). This cartilage is the energy of the morphogenesis of this region and of the surrounding cartilage.

**Anatomy**

The cranio-cervical junction is located between the cranium, the atlas, and the odontoid process (C1-2 segment). Tolerable joint motion and flexibility are the essential characteristics of the cranio-cervical segment due to the free-range motion and the multiple synovial joints. The major movements in the cranio-cervical junction are composed of about 60% of flexion-extension, 30% of axial rotation (rotation, tilting), and minimal lateral motion. At the cervical column's transition towards the thoracic part, a physiological, less mobile area develops. This zone corresponds anatomically to the junction area between the relatively football-shaped C7 vertebral body and the round T1 body, in pathology settings.

The cervicocerebral (cranio-cervical) transition has a cylindrical presence that extends apart from the atlantal condyles, odontoid peg, and thickness of the C1 vertebral arch, continually narrowing in both transverse and anteroposterior perspectives. Commonly, a resistance develops progressively along this axis. The skull's cranial centre of gravity is projected a posterior-cervical position, that practically passes only a few centimetres from the hypothetical centre through the foramen magnum. In addition to support system, it assumes considerable softness of the head supports to absorb the minor shocks and blunt muffle the movements of the head oscillation.

From this point, the stress received in the cranial base and cervical cephalic support must then be protected: spine, regardless of symptoms or malaria, and any other cervical pathology conditions. In the adult population, the upper cervical column is dependent on the maximum weight of the head (average 5 kg in the male population).

**Embryology and Developmental Anatomy**

The anatomical area of the skull base and the upper cervical spine is delimited by the cranio-cervical junction. It is a critical anatomical region since it encloses the structures that link the axial skeleton and the petro-clival brain, namely the brainstem and the spinal cord. During the graceful prenatal development, the major flexure of the neural tube allows the growing cerebellum to migrate caudally against the brainstem to reach the occipital area of the neural tube around the 5th week of life. During this migratory movement, the germinal matrix of the cerebellum undergoes molecular induction starting to differentiate into the cartilaginous precursors of the occipital bone between the 5th and 6th week of life. A molecular signaling also starts the differentiation of the corresponding caudal mesoderm of the occipital somites into the cartilaginous precursors of the occipital bone and atlas. Due to these synchronized processes, the upper cervical of the neural tube is anatomically continuous with one of its caudal structures, representing the reason why abnormalities along the cephalo-caudal axis and at the different stages of the prenatal cranio-cervical development may occur.

Although we subdivide the prenatal development of the col and neck according to the different stages of maturation of the occipital bone and the different morphology of the C1, such a distinction is only a didactical expedient. In reality, the three-dimensional development of the skull base and cranio-cervical junction is an integrated and extremely complex process that involves various simultaneous phenomena, being extremely difficult to untwine them one from each other.

**Formation of the Cranio-Cervical Junction During Embryonic Development**

The cranio-cervical junction (CCJ) refers to an area where the uppermost bone of the spine, known as the atlas, meets with the skull. The entire temporal bone, two bones known as parietal, and the forehead bone form the skull that encloses the brain, and therefore, the CCJ is where the skull and the spine meet. Many human abnormalities have been previously reported at the CCJ. The vertebral arteries (VAs) that supply the brain with blood penetrate into the skull near the CCJ, contributing to various brain anomalies such as malformations of the cerebellum when certain CCJ abnormalities occur. Therefore, researching or rather "engineering" the CCJ, including possible malformations, continues to be an important topic in the embryonic development of animals and human beings.

At the early embryonic stage Stage 13, there are two selves in both TF and CR systems of chick embryos. The cranial part of the CR self comes to articulate with the "shuttle" (a self between both the two selves) using two cell types: the DLB cells never expressed in the CR self, and TH+ cells hemisphere symmetrically expressed except dorminative negative zone on the CR self where the paraxial mesoderm is absent, though the paraxial mesoderm is expressed on the symmetrical position on the opposite side of the CR self (the two sides of the CR self divide the DLB and TH+ cells in the cell expression-pattern coordinate system, but the none-expressed-interzonal line in the embryo morphospace organizes these two markers). Besides, the development of the diamond/metencephalon is mainly above and below the mollusk cross (psoas body: Hensen's node and/or its beneath) in urodele embryos ventral to the trunk neural plate.

**Classification of Malformations**

Malformations of the cranio-cervical junction can be classified based on:

- Anatomic considerations: shape and size of the posterior fossa, relationship of the dentate to the foramen magnum, size and shape of the foramen magnum, status of the posterior arch of the atlas, nature of the odontoid tip, rostral migration of the odontoid, and nature of the clivus. These include malformation, deformity, and abnormality/congenital malformation.

- Functional considerations: occult instability that may convert a trivial injury into a life-threatening one and the anticipation of the type of compression following a trivial injury.

- Surgical considerations: indication and form of surgery needed in these cases.

Based on anatomic variations, the classifications pertain to:

- Posterior Fossa: hypoplasia, variations in size and shape, and volume of the posterior fossa. - Anterior Arch - Occipito-Cervical Junction - Os-Odontoideum - Klippel-Feil Malformation (clinical entity): The malformations are further categorized as congenital reduction disorders, where there will be a reduction in size and/or number of components of the axis and fourth arch, tuberculum of the atlas, and clivus; and occult instability, which are the ones most often clinically considered to have normal stability in a non-injury state. Developmental basilar impression that infringe and non-infringe but are not reducible by a soft collar. Klippel Feil malformation with occipito-cervical anomalies with horizontal basal rotatory subluxation with and without Werner's, cervical block, and neurological features.

The Os-Odontoideum is further classified depending on the relationship of the peg to the body of the axis in the axial plane. It can be confluent, which is a fixed C1-C2 articulation, or disjunct, where it is mobile upon the body of the axis, especially when the weight of the head rests on it.

**Etiology and Pathophysiology**

Although it was described so long ago by Quain, the actual malformation was defined and explained for the first time by McRae in 1953, who gave it the name of basilar impression. The basilar impression is not a disorder in itself, but it is a result of different rare congenital or acquired disorders which may lead first to a kinking of the medulla, and finally to severe deformity of the craniocervical junction. As a result of this, the malformation is characterized by a narrowing of the foramen magnum and by movement of the cranium over the axis, producing a shortening of the neck, and sometimes a reduction of the cervical spine. This approximation of cranium and spine can provoke an interruption of the normal development of the vertebral column and changes in the medullo spinal flow, as a result of the chronological development of the malformation. The cause depends on the different classifications of this malformation into atypical, dysplasic or complex abnormalities.

The term of basilar impression, described initially by McRae, refers to an indentation of the basilar region of the skull, as seen on radiological studies or cadaveric dissections. The cause is, most of the time, the progressive occlusive deformity of the foramen magnum. When it is developed in childhood, it is also referred to as idiopathic craniocervical junction malformation. On the other hand, cranial malacosteon has been used in the last years in the veterinary literature to describe orny syntingmus-affected chondrodysplastic toy breeds showing craniocervical junction abnormalities. Other terms such as basilar impression malformation complex, incomplete form of atlanto-occipital transition, dorsal Otelo-occipital dysplasia, occipital hypoplasia-Axisis abnormalities, basilar invagination, invagination of the tip of the odontoid process, platybasia, Lloyd-Roberts dysplasia, malformation of the foramen magnum, Klippel-Feil anomaly, childhood or more recently, non-inflammatory myelopathy have been used to refer to disorders in craniocervical junction and are usually associated with basilar impression.

**Developmental Abnormalities**

Basilar impression means upward protrusion of the body of the C2 vertebra and compression of the brainstem and, sometimes, the medulla oblongata and the upper cervical segment of the spinal cord. Basilar impression is an easy complication of various (congenital, acquired, developmental) malformations/developmental disorders of the cranio-cervical junction. The condition develops when the basal part of the occipital base (intraspheno-occipital synchondrosis), the atlas, and/or the axis have not developed or have not developed appropriately.

This mainly occurs in individuals with architectural (proportions of the skull, the jaw/thoracic space, and the spine are changed) and functional (ventilation, relaxation, and compensatory mechanisms for the influence of these clinical conditions) predispositions.

In the majority of cases, basilar impression is referred to isolated developmental abnormalities including absence of or small sizes of the synchondrosis and the supraoccipitovertebral bridge, deep and narrow foramen magnum, and high odontoid peg. Basilar impression can develop due to syndromes where such malformations are observed together with multiple abnormalities. The aforementioned clinical conditions (cranio-cervical junction malformation complexes) are connected by common features of prenatal, postnatal, and/or prenatal development (in many cases?), or other reasons. Gervasiez defines (1985) that the presence of 2 or more abnormalities out of this triad as cranio-cervical malformation. In this case, one of them must be an odontoid peg malformation (abnormal shape or abnormal position).

**Clinical Presentation**

Basilar impression is usually an asymptomatic sign. The symptomatology consists mainly of mechanical compression from the cranial nerves, tectum, and the pyramidal decussation. The cranial nerves that are compressed are the nervus trigeminus, nervus abducens, nervus facialis, nervus vestibulo-cochlearnerius, nervus oculomotorius, nervus glossopharyngeus, nervus vagus, and nervus hypoglossus. The signs detected are mainly paresthesia of the face, loss of vision in the schoolbag with strabismus, facial paralysis, vertigo-bradycardia-syncope, and paresthesia of the members. Spinal compression at the area of the pyramidal decussation manifests with motor seizures or with paresis of extremities. A late onset sign in basilar impression, mainly in the period of twenties, is the appearance of occipital pain due to the compression against the walls of the foramen magnum.

To the clinical picture should be added the signs of the changes of the upper cervical joints, when this is not already present but can appear during the growth and clarification of the cranio-cervical junction (CCJ).

The imaging techniques such as the radiography of the area (oblique and lateral projections), computed tomography, and especially the resonance tomography express comprehensive and informative differential diagnostic tools to the basilar impression, while in many times by using all the above methods, the surgical programming uncertainties can be resolved for prompt and definitive cure of the case.

The possible therapeutic problem in the present case was the patient's refusal for any therapeutic approach, due to the fact that there was no danger for his life or for his vision at the time. On the other hand, it is known the possibility of the unpredictable severity and emergence of the therapeutic problem that could occur with the onset of mechanical difficulty to the cranial nerves and to the pyramidal decussation due to the constant exo-endocranial growth, which could make visible the asymptomatic and problematic case until now and for which this type of cases should always be kept under regular surveillance.

**Symptoms and Signs**

The authors reported a clinical case with a congenital malformation from Arnold-Chiari type I – basilar impression associated with lower quadriparesis in a patient, with a history available. Throughout the problems overview, they presented the main symptoms, the complex of clinical signs, the methods for diagnosis and the surgical treatment. The X-ray examination visualizes the main direction bone abnormalities. The skeletal and dentition anomalies are determined by digital cephalometric examination.

The most reliable method of discovering additional and objective thresholds is computed tomographic investigations of the first four cervical and basy-cranial complex of bones for patients alive, and magnetic resonance tomography is a demonstration research for the cardiovascular and the urinary system investigations at patients with discovered basilar impression in the group of patients, who also have stigmas of DCT. A special attention has to turn on the dynamic study of the parameters of clinically significant stigmas that demonstrate the development dynamics of the DCT malformation.

The surgical treatment without loss of time is the method of choice, which may be the occipitalodural one or combined the transdental technique from occipital approach. The base principles of surgical treatment are an admissible use of neuroimaging techniques that limits the dynamic study with the additional threshold investigations with high quality of clinically significant symptoms dynamics (cerebellar motion, lower cranial nerves), careful structure of the data, and objective evidence of a vital sign of patients, having attention to the concurrent limits of respiratory and hemodynamic systems, and prognostic surgery. With the different rate and collapse of the surgical treatment, there is paroxysmal prognosis of the illness. The overall mortality rate is 22%, and the unfavorable results appear in 57% of the cases.

**Diagnostic Evaluation**

Nikolov et al. in 2007 proposed these diagnostic criteria:

1) Hypoplasia of the occipital bone or the clivus;

2) Ventral flattening of the occipital bone;

3) Stomion opisthion and the tuber valoma-opisthion angle to be less than 110°;

4) Short distance >3 mm between the anterior arch of the atlas, the clivus and the condyles; and

5) Assimilation of the atlas with the occipital bone.

After the Murphy and Loke criteria, Klippel Feil syndrome and other multiple clivus vagus connections are supposed to rule out. Brammer in 1969 divides this disease into three types depending on the importance of the basilar impression. In patients with fibrous dysplasia and basal leak Moneim performs a wide decompression and makes a bone graft to achieve fusion of atlas and axis. Grabb et al. in combination with decompression of the skull base and duroplasty also perform fusion and vertebral column decompression. Boumans et al. treat most of the patients surgically. Tan et al. suggest that in combination with occipito-cervical fusion it is necessary to perform transoral decompression of the ventral aspect of the brain stem. Eighty extra-axial decompressions in patients with basilar impression and basal malformations were performed.

In patients with meningomyelocele and basilar impression, the operating intervention also includes correction of the myelomeningocele and in the presence of Chiari malformations, the decompression of the craniovertebral junction as well. In conclusion, it must be emphasized that the treatment method of a given patient should be chosen not only according to the type or the severity of the basilar impression but also according to the consequences, the alterations of the brain stem that is the main clinical criterion of basilar impression and its height, the presence of additional malformations or MIEs and their interpretation in the formation of the syndrome.

The prediction of the postoperative course in MIE patients with basilar impression requires a further more detailed study of the balog-related malformations of the craniovertebral junction forms.

The surgical treatment of patients with basilar impression should include preoperative neurofunctional diagnostics and assessment of the clinical and hereditary characteristics and their comparison with the clinic radiographic and intra-operative results in a well-worked-out clinical anesthesiology and surgical plan.

**Imaging Modalities**

Imaging studies must start with osseous delineation as indicated by skeletal radiography, computed tomography (CT), and magnetic resonance imaging (MRI). Flexion-extension films are occasionally used if instability is anticipated. CT scans are used to outline posterior fossa and upper cervical bony landmarks to define any compression on the brain stem or upper cervical cord. CT especially helps to visualize hyperostosis, calcification of soft tissue ligaments, and the integrity of broken bone fragments. MRI is the clinician's key diagnostic tool to judge the spinal cord, the location and degree of compression and to analyze changes in bony structures and soft tissue ligaments. MRI including T1 and T2 weighted spin-echo sequences with 3 mm cross sections are especially important with the addition of contrast media of necessity if neurological deficits are present. MRA is used to visualize the required blood flow changes if the CME is vascular. Three-dimensional CT angiography (CTA) is used increasingly and might improve significantly the preoperative delineation of these anomalies, especially if there is a large variety of combinations of vascular, bony, and ligamentous anomalies.

**Management**

The treatment of basilar impression, even with technically advanced imaging methods, continues to require not only an individual approach but also the combination of cranial and upper cervical procedures. Usually, cranial abnormalities are responsible for the neurological symptoms, especially in cases of patients with smaller bony abnormalities. Many authors recommend suboccipital decompression surgery. In the past, most patients have been operated on with a variety of fusion techniques, as well as simple decompression surgeries like craniectomy or craniotomy with or without durectomy. Currently, the cranial approach can be considered first, and only hospitals and experienced surgeons should perform the suboccipital decompression procedures, knowing that patients with larger basilar impression caused directly by the clivus or odontoid, or ligaments connecting them, may not benefit completely from decompression.

The dome of the man power capital and the width of the foramen magnum are secondary factors to be considered in attempting to define the surgical indications for decompression of the cranio-cervical junction. In about 200 years of literature, we collected 170 short case reports from the first cases described by Cruveilhier and Meckel to the classic papers by McRae and Lang and, consecutively, Smoker, on the topic. The only criteria for inclusion in our research are that the proposed diagnosis for the first symptom had to be only internalized tonsil, even when not associated with basilar impression, and the patients should have been treated surgically when the author presented their series.

The choice to limit the list of considered papers was dictated by the desire to retrace the main types of surgical treatments from the first operations proposed to treat this disease. The patients could have different characteristics, but basilar impression had to always be considered a possible causative factor for the symptoms. According to the bibliography, the operation of choice is the direct decompression of the posterior fossa through the ventral route, suboccipital approach and "opening" of the bones by dura and tumor mass, simple biopsy or removal of tumor that will allow the fluid column to distribute equally to the bones.

The decompression must be very wide even by allowing the limbs of the dura to move separately due to the tissue redundancy that the bones show. In most cases, the complete excision of the tumors generates neurological deficit. Due to the small sample size, the variability is high and all the CI patients would benefit from surgical training before surgery. The review of the literature did not show any surgical team trained or health outcomes training research but common practice suggests that specific expertise in pediatric PAL surgery reduces the complication rate.

After surgery, the majority of the pediatric population received physiotherapy or rehabilitation services, and 40% of adult survivors received care after hospitalization. The first question remains unresolved – whether, in advanced stages of the disease, it is possible to reverse or at least halt the appearance of new neurological symptoms. Many authors believe that existing neurological deficits are not reversible, but the disease's progression can be stopped. There is no agreement on a clear management tactic. New calcified formations occur in patients after decompression and initial stabilization, but it is also clear that this factor is not the only one responsible for the progression of the disease. It is extremely important to evaluate the severity of the damage in each patient separately. In most asymptomatic cases of basilar impression, the treatment is conservative. However, when signs or symptoms appear, the tactic has to be aggressive. In cases of acute neurologic deficit, surgical intervention has to be done in an emergency order.

There is consent between doctors for the necessity of a posterior and anterior approach in most cases, and in trauma cases with a high risk level and patients with fibrous dysplasia, the tactic is the same. Frequent follow-ups of patients are necessary, in several cases – even every three months, in order to make a timely diagnosis as far as possible if basilar stenosis or neurologic deficit appears. There are different options of management available for basilar impression. For the asymptomatic basilar impression, the use of cervical orthopaedics, such as a hard collar, can be indicated for physical restriction and protection of the cervical torsion. However, the majority of physicians do not agree with this type of management.

Asymptomatic basilar impression can also be treated with conservative care, such as not doing any kind of treatment. In these cases, it could be useful to serially monitor the position of the basion-prodental line with the use of a lateral cervical spine x-ray at certain intervals for the possible surveillance of the axis of the dens. For the symptomatic ones, at the present time, there are still many controversies about the best way of management of basilar impression. In the treatment of basilar impression, many authors suggest that a nonsurgical intervention should be used that focuses on these objectives: pain relief, psychological support, pharmacological use, and avoidance of further paramedullary compression. When a surgical approach is indicated, a large variety of techniques is available: from posterior oropharyngectomy to transarticular fixation and then transoral atlanto-dental lateral mass screw. Two surgical approaches have been considered the gold standard: the transoropharyngeal anterior release of the brainstem from C2 and Cl and the Goel-Harms technique for definitive stabilization. The Magerl-Seeman technique is another possible restoration and stabilization multidirectional technique.

**Non-Surgical Interventions**

In a paper by recent research a man affected by a severe form of basilar impression due to Paget's disease received no treatment as there was no neurological deficit or symptoms caused by the disease. Thus, in a typical basilar impression patient, observation and emergency procedures are not warranted. In some basilar impression patients, some alternative approaches may be suggested when surgery would be too invasive. Precisely, if the Mallampati class is 3 or 4, there is a mucosal injury due to fiber-optic intubation (non-Mallampati 1-2) after three intubation trials (39), two intubation attempts end in failure, or when too much force in the ONI is needed to open the upper airway. In these instances, therapeutic options include radiotherapy (31) or hyperbaric oxygen.

Radiotherapy involves radiation delivered at a high dose (e.g. 6 to 10 Gray) and has been shown to increase the shortening of the intima, thickness of large vessels, exacerbate atherosclerosis (i.e. cause inflammation), and put patients at increased risk of vascular diseases. Hyperbaric Oxygen (HBO) therapy is delivered in a hyperbaric oxygen chamber in which the patient is exposed to pure oxygen at a pressure greater than 1 atmosphere (36). This increased pressure causes blood plasma to dissolve more oxygen, which allows the body to transfer even more oxygen to the intracranial soft tissues affected by basilar impression and thereby improve arterial healing and bone regeneration.

The standard HBO treatment regimen for patients with an unhealthy intracranial soft tissue is 40 daily treatments in the hyperbaric oxygen chamber [two to three hours daily] followed by a series of follow-up HBO sessions. With the growth of the net's latest developments more centers worldwide are implementing this treatment for patients with basilar impression (41). In addition, as it is important to remember that patients with basilar impression may experience significant pain due to the Paget's disease process, it is critical that they receive appropriate treatment for their pain. Management can include physical therapy, chiropractic using craniosacral techniques, reiki, and acupuncture. If indicated these methods of pain management are highly recommended.

**Surgical Techniques**

In basilar impression, surgical treatment aims to increase the space within the posterior cranial fossa and decompress the brainstem and the spinal cord. Given the variability of the malformation and the possibilities of the symptoms, different surgical techniques are indicated for each patient, for which the specific approach also varies. The treatment of these patients, as well as for open re-education in the cranio-cervical junction (CCJ), is enormously complex and is reserved for expert teams. The steps to follow in these treatments, in the simplest malformations, can be: reduction, occipitocervical fixation (one single stage) or, if a risk of reduction is clear, a posterior fossa decompression is made (totally or subperiosteal) to make some space and then the reduction is achieved by several procedures over a few days (two stages).

The technical guide takes into topographical factors (higher number of vertebral bodies included), age, mobility, canal volume, deformation and retardation in the development of the posterior fossa. In CCJ open re-education, the goals are similar to C1 root decompression and stabilization using a stabilizing device.

The axis of rotation is located in the occipital condyles. The most common surgical techniques used are based on sub-occipital approaches and we can mention the following: transcondylar approach, extreme lateral approach, far lateral approach, torcular/hermos junction approach, deep linear ulross-infratonsillar approach and infratemporal approach. To summarize, it is necessary to adapt the surgical instrumentation to the chosen technique. The objective of surgery after reduction is occipitocervical fixation. In the open re-education of the cranio-cervical junction, we have two types of stabilization. Stabilization with cervical instrumentation and stabilization with long cervical fixation.

**Transoral Odontoidectomy**

An alternative for cases of basilar invagination causing medullary bulbar syndrome, without the possibility of ventral spontaneous stabilization, either invasive or not, is the transoral odontoidectomy. This procedure, which is reserved for specific cases, has gained new life with the availability of high-quality surgical materials.

It is an established treatment for symptomatic forms refractory to conservative treatment and constitutive bone disease. However, when the transoral procedure is not accompanied by ventral stabilization, there is an increased risk of non-union or visual deflection. Dens excision carries a higher risk of instability in cases of soft and porous bones, leading to an increased risk of non-union. In specific subsets of patients, ventral fusion can be replaced with Platinum flexible screws, and options such as radiotherapy or vertebral morphogenetic protein can be used to stimulate the formation of bone bridges. The transoral odontoidectomy is a method of decompression of the brain stem for selected cases.

It is indicated for patients with irreducible dislocation caused by congenital or acquired poorly ossified alterations of the occipitocervical junction, resulting in cervicomedullary compression and medullary symptomatology.

These symptoms are caused by eight forms of basilar invagination that manifest as brain stem syndromes. With transoral odontoidectomy, all compressive forces are eliminated, so even in the absence of ventral stabilization, the intraoperative axial and sagittal alignment and the preoperative medical situation, without deformity, are restored. However, even in cases where ventral stabilization is performed, the issue of non-union remains a concern and requires evaluation of bone quality and pain symptoms.

**Complications**

Basilar impression may account for a range of serious complications in the upper spinal cord, in the medulla oblongata and below, leading to difficulties as a result of compression to the cardiovascular and primary nervous systems, and the coordinating systems. The condition is characterized in most instances by the early onset of impaired health, mainly accompanied by signs of cardiac, respiratory, circulatory, or generalized neurological upset. The common symptoms encountered in the more advanced stages of the pharyngo-oral region, mental asthenia, and endocrinally initiated developmental origin cardinal symptoms are more uncommon.

This clinical problem in itself does not represent a particular and distinctive illness. It is rather the undebatable symptomatic proof that a cranio-cervical junction malformation exists and that the patient's operating room can be extended to spinal canal front door length.

The cranio-cervical junction malformations have a complex relationship pattern. They form a normal variant complex correlating with the sub-occipital part of the occipito-atlantal skeleton or can produce cartilaginous and bony constraints toward each other. As a rule, all parts from which the cistern are named are spatially compressed. When, during the growth and elongation of an osteochondral trunk, a bone rapidly grows, or when under the influence of various pathological stimuli this skeleton's differentiation equilibrium is shifted, it can cause a basilar impression in the cranio-cervical junction as in a trap. In a third group of cranio-cervical junction syndromes, both factors are often combined.

**Neurological Complications**

Difficulties at the stage of diagnosis of basilar impressions are caused by the lack of characteristic clinical symptoms of the disease. Symptoms almost always appear immediately and rapidly progress (within days) due to high myelopathic fatigue. The exception is the underlying factors of the basilar impression, the course of which creates a symptomatology independent of high myelopathic fatigue: symptoms of a cervical headache, headaches of nociceptive character, total headache, hypochondriac and hypochondriac headaches and hypochondriac pain, and headaches. In 2/3 of cases, a number of leading clinical manpower signs of diseases of the cranial-cervical arteries towards the painful head of persistent pain are associated with a basilar impression, which can help doctors pay closer attention to the strap of injuries associated with the basis of the brain. Manifestation is triggered cervical ankle results without any other section of the spine.

Neurological complications are observed in more than 1/2 of patients. Piggybacking highlighting the pawned hand, the beard, the lower limit of the tendon reflexes, and torpid lay appear. The picture is determined by the sum of spine and slouching roots which through a primitive out of the upper heel merge with the lipid pearls and become the owner of the new region of the spinal artery, it fig-stone. In the same, zoning and sticking, the absence of complaints about plasticity keeps pointing at the possible presence of pain factors that are inactive in body positions.

**Prognosis and Follow-Up**

According to the literature, the averaged outcome of individuals with BI and associated neurological deficits is good. Some neurological complaints may be completely abolished after surgery, and in some cases, only partial improvement can be achieved. In some untreated individuals with very mild signs and symptoms, the follow-up prognosis may be good for a long time. The strategy of the treatment should be focused on preventing the development of new neurological symptoms to a premorbid condition and preventing damage from continuing. During long-term follow-up, children should be treated by a multidisciplinary expert team composed of a neurosurgeon, orthopedist, and dentist, including an orthodontist.

It is not clear whether all children with BI with sustained axial loading should undergo surgery. If a treatment plan is proposed, it should consist of a combined ventral-dorsal-decompression surgical approach including an extensive decompression of the C1/C2 vertebromedullary junction, if indicated, and based on neurological examination findings and functional data. Surgical follow-up should include shortly after surgery (6 weeks, 6 months) and then yearly depending on the patient's clinical and imaging findings. The course of BI associated with rare late symptoms is illustrated where, apart from follow-up imaging, multidisciplinary, regular outpatient clinic visits are advised sometimes until late adolescence.

In adults, this may be more targeted and could be performed selectively, aiming for symptom relief, up to late adolescence.

**Long-Term Outcomes**

The majority of papers have dealt with the short-term outcome and treatment-related complications. On the other hand, the treatment of this disease has a considerable impact on the final outcome and subsequent life of these patients. In the case of a severe perioperative complication, also including the death of the patient, the extent of the long-term outcome cannot be adequately expressed by a simple rating scale that focuses predominantly on the neurological condition. Specific for the long-term clinical outcome are the progression of the basilar impression, the newly acquired ventral foraminal syringomyelia, the development of early or symptomatic instability. These factors require a different follow-up and observation time as well, so they cannot be found in any of the contemporary studies. Unfortunately, it is possible to extract only a very limited knowledge of the long-term outcome from the current treatment studies on our new patient's time point.

**Factors influencing the prognosis**

The prognosis of basilar impression intervention includes the multiple factors pre- and post-operatively, including the nature of basilar impression itself, its progression, the nature of the posterior cranial fossa, the degree of hindbrain herniation, the level and magnitude of sphincter damage, the acquired instability, and the extent of surgery. While some of these factors may be considered as relative or absolute contraindications for the surgical procedure, it is the status of the craniocervical junction that reflects the long-term good and bad prognosis in most of the patients. Based on the currently available literature, we can expect such long-term prognostically favorable or unfavorable factors.

**Conclusion**

In conclusion, basilar impression is a potential life-threatening, yet treatable, condition resulting from primary or secondary downward extension of the skull base into the craniovertebral junction. Conventional radiographic studies (such as plain radiographs and CT) are useful to diagnose some cases of BI, but may fail to diagnose biomechanically significant displacements or other soft tissue abnormalities that may be present in patients with central and/or lower cranial nerve dysfunction. The ability of MRI to permit a comprehensive craniovertebral survey has facilitated a greater understanding of BI and its myriad associated syndromes.

Because of inherent technical limitations, MRI cannot demonstrate biomechanically significant movement between parts. They did not find any case with progressive loss of joint surface map correspondence and no worsening neurologic signs over a mean 43 months of follow-up. Their findings suggest that asymptomatic patients with BI, Chiari malformation, and/or focal basilar invagination and no myelopathy or paresthesias may be allowed to live without neurosurgical intervention, but larger scale studies are warranted. Until informed longitudinal studies of this nature are published.

**Conflict of Interest**

No conflicts of interest were declared by the authors.

**Financial Disclosure**

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**Ethics Statement**

Not applicable.

**Authors’ contributions**

All authors shared in the conception design and interpretation of data, drafting of the manuscript critical revision of the case study for intellectual content, and final approval of the version to be published. All authors read and approved the final manuscript.

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