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Massimiliano Visocchi *Editor*

New Trends in Craniovertebral Junction Surgery

Experimental and Clinical Updates for a New State of Art

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Editorial

Why the Craniovertebral Junction?

Massimiliano Visocchi

Abstract The craniovertebral junction (CVJ) has unique anatomical bone and neurovascular structure architecture. It not only separates the skull base from the subaxial cervical spine but also provides a special cranial flexion, extension and axial rotation pattern. Stability is provided by a complex combination of osseous and ligamentous supports, which allow a large degree of motion. Perfect knowledge of CVJ anatomy and physiology allows us to better understand instrumentation procedures of the occiput, atlas and axis, and the specific diseases that affect the region. Therefore, a review of the vascular, ligamentous and bony anatomy of the region, in relation to all possible surgical approaches to this anatomically unique segment of the cervical spine, appears to be absolutely mandatory in order to preview and to overcome possible anatomy-related complications of CVJ surgery; moreover, knowledge of the basic principles of instrumentation and of the kinematics of the region, since they interact with the anatomy, seems to be strategic in preoperative planning.

Historically considered a no man's land, CVJ surgery, or the CVJ specialty, has recently attracted strong consideration as a symbol of challenging surgery as well as selective top-level qualifying surgery.

Although many years have passed since the beginning of this pioneering surgery, managing lesions situated in the anterior aspect of the CVJ still remains a challenging neurosurgical problem. Many studies are available in the literature, aiming to examine the microsurgical anatomy of both the anterior and posterior extradural and intradural aspects of the CVJ, as well as the differences in all possible surgical exposures obtained by the 360° approach philosophy. In this paper the author provides a short but quite complete at-a-glance tour of personal experience and publications and the more recent literature available.

Keywords Instrumentation and fusion · Endoscopy · Transnasal approach · Transoral approach · Extreme lateral approach · Far lateral approach · Craniovertebral junction

This special *Acta Neurochirurgica* supplement is entirely dedicated to the craniovertebral junction (CVJ). Anatomy, physiology, embryology, malformations, microsurgery and endoscopic surgery, as well as instrumentation and fusion procedures of the CVJ, are the main topics of the supplement.

Many national and international contributors have been invited to provide their experience and to highlight tips and tricks in their surgical practice according to the trends at their own schools and the worldwide interest in this special topic.

At present, if we do keyword searches in PubMed we can find 837 papers on 'craniovertebral junction', 558 on 'craniovertebral junction surgery', 311 on 'craniovertebral junction abnormalities', 470 on 'craniovertebral junction anatomy', 102 on 'craniovertebral junction transoral approach', 40 on 'craniovertebral junction transnasal approach', 27 on 'craniovertebral junction far lateral approach' and 5 on 'craniovertebral junction extreme lateral approach.'

Obviously such a search must be considered unreliable since it does not reflect the exact number of specific papers dealing with the topic. Nevertheless, the proliferation of special issues on CVJ in different journals, as well as local societies and the international CVJ society (i.e. the Craniovertebral Junction and Spine Society), demonstrate that such a scientific topic deserves strong consideration in the neurosurgical and spine community.

Schematically, we can divide the topic into direct and oblique approaches.

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Direct Approaches

These are the transnasal and transoral anterior approaches, and the suboccipital midline approaches for decompression, instrumentation and fusion of the CVJ.

Endoscopically Assisted Endonasal Procedures

The introduction of video-assisted procedures in pituitary surgery and for treatment of skull base lesions *latu sensu* has allowed the publication of many anatomical and clinical studies dealing with the endoscopic endonasal approach to different areas of the midline skull base (from the olfactory groove to the CVJ) in recent years. Historically, the very first cadaver and clinical study publication line started in 2002 with Alfieri and then continued with Jho; by means of one or two nostril routes, both of them performed a totally transnasal endoscopic odontoidectomy [1]. Most important, Rodlens endoscopes were used: 2.7 or 4 mm in diameter and 18 cm in length, with 0°, 30° and 70° lenses. The surgical landmarks leading to the CVJ were considered the inferior margin of the middle turbinate, the Eustachian tubes and the nasopharynx. To identify the nasopharynx, the inferior margin of the middle turbinate was assumed to be a reference point. The juncture between the clivus and atlas was indicated by a line drawn between the Eustachian tubes. Alfieri et al. [2] concluded that "contrary to a conventional transoral approach, this endoscopic endonasal approach provides unlimited access to the midline clivus and a potential of carrying out surgical decompression at the ventral craniocervical junction without adding C1–2 instability". Three years later, Cavallo confirmed such an observation in a cadaver study [3, 4]. The first surgical case of fully transnasal endoscopic resection of the odontoid was performed by Kassam in 2005; the patient was a 73-year-old woman affected by rheumatoid arthritis. Kassam's equipment consisted of (1) a 0° endoscope; (2) a navigation system; (3) an ultrasonic aspirator; (4) a long angled endonasal drill; and (5) bayoneted handheld microinstrumentation. Soon after his very first experience, Kassam reported that "the defect created by transnasal approach is above the level of soft palate and should not be exposed to the same degree of bacterial contamination" [5]. Nevertheless he concluded that the transoral approach still remains the gold standard.

Although some studies have concluded that the endoscopic endonasal approach provides less morbidity than the transoral approach, this matter is still under discussion, which is consistent with our personal experience [6, 7].

The 'nasopalatine line' (NPL) concept was conceived in 2009 by Kassam's group in order to predict possible surgical problems related to anatomical limitations. The line was created by connecting the most posterior point on the hard palate in the midsagittal plane with the most inferior point on the nasal bone.

The intersection of the so-called Kassam line with the CVJ is measured relative to the inferior aspect of the body of C2 along its posterior surface. A preoperative radiological study of the possible anatomical limitations of the endonasal approach is mandatory in predicting the maximal extent of inferior dissection and the efficacy of odontoid surgery. Again, Kassam's conclusions were that the transnasal approach is indicated in *selected* cases as a valid alternative to the transoral microscopic approach for dealing with CVJ and should be performed only by dedicated endonasal surgeons after consistent cadaver anatomy training [8].

Endoscopically Assisted Transoral Procedures

In order to avoid full soft-palate splitting, hard-palate splitting, or an extended maxillo/mandibulotomy, use of a 30° endoscope has been proposed for the transoral approach. The introduction of the endoscope in such challenging surgery has changed the modern transoral philosophy; the operator gains access in all directions by rotating the instrument, thereby avoiding more destructive manoeuvres. In more detail, abnormalities, as in the high midclivus, can be visualized with the aid of an endoscope, without extensive soft- or hard-palate manipulation [5, 8]. Superior illumination can be obtained by the light source being located at the level of the abnormality.

One of our cadaver studies comparing the transnasal and transoral approaches demonstrated that the surgical exposition obtained using the endoscope in the transoral approach appears to be wider than that obtained using the transnasal approach in both the anterior and lateral projections: (1) in the coronal plane, both in coronal exposition (transnasal inferior to transoral from 50.77% to 83.88%, average 70.34%) and in the coronal surgical angle (from 65.58% to 86.71%, average 76.70%); and (2) in the sagittal plane, both in the vertical surgical angle (from 22% to 77.42%, average 56.53%) and in vertical exposition (transnasal inferior to transoral from 5.89% to 76.48%, average 35.89%). The surgical domain in the sagittal plane was found to span from the middle third of the clivus to the NPL with the transnasal approach and from the inferior third of the clivus to C3 with the transoral approach. Our conclusions were, and still are, that (1) with the aid of the endoscope and image guidance, it

is possible to approach the ventral CVJ *completely*; (2) the inferior third of the clivus overlapped the surgical domain areas of the transnasal and transoral approaches; and (3) transorally it is possible to gain access to the CVJ with minimal tissue dissection, no palatal splitting and no compromise of surgical freedom, thanks to angled-lens endoscopes, which significantly improve the exposure of the clivus without splitting of the soft palate [9–12].

Endoscopically Assisted Transcervical Procedures

In 2007 an alternative endoscopic route to the anterior CVJ with the endoscopic transcervical approach was described by Wolinsky [13]. The choice of this option is strictly related to the limitation of the transpharyngeal approaches. With use of the transnasal and transoral (i.e. transpharyngeal) approaches, in the case of dura laceration, contamination with oral flora is likely to occur, tremendously increasing the risks of infection, meningitis and poor pharyngeal healing. Moreover, transcervical exposure is quite familiar to neurosurgeons and is mainly used in screw fixation of traumatic odontoid fractures. Such a 'revised' strategy has been proposed by the author for the surgical treatment of down-seated CVJ lesions. Since the postoperative recovery time is shorter, it is possible to allow patients to consume food orally shortly after removal of the endotracheal tube. Moreover, in this surgery there is no need for a tracheostomy or a gastric or duodenal feeding tube as a result of the procedure in patients without preoperative dysphagia. On the other hand, CVJ decompression is too oblique and insufficient because there is very little control of the C1 ring, making this surgery uncomfortable and challenging. In fact the unfavourable angle of the C1 ring matching makes it difficult or impossible to dissect this portion, which needs to be removed to gain access to the lower clivus, increasing the risk of damage. Basilar impression and other high pathologies do not seem suitable for this approach [13].

The Navigated Transoral Approach

The surgeon's ability to visually reconstruct the imaging-magnified three-dimensional anatomy is greatly improved by the use of image guidance, allowing a thorough inspection in multiple reconstructed views of the anatomical images before and during the surgical procedure. In order to better plan the surgical technique, preoperative and intraoperative use of neuronavigation is helpful and allows fluoroscopy to be avoided. It takes no more than 5–10 min to perform the registration procedures. The calculated accuracy is less than 1 mm, but the error associated with a spinal shift is not completely eliminated. There is increasing acceptance of robotic surgical technology, which holds promise for use in a growing number of applications. Nevertheless, its indications in head and neck surgery are limited so far. Use of robotics along with neuronavigation allows (1) performance of two-handed surgery through small openings; (2) use of articulated instruments; and (3) improved fine motor control with a tremor filter [9–11].

Sublaminar Instrumented Wiring Versus Lateral Mass Screw Implants

In 1939 Gallie reported his method of laminar wiring. Bone grafts from the patient's iliac crest were put over the C2 spinous process against the C1 posterior arch. In 1971 Barbour first introduced anterior transarticular screw fixation of C1–C2, and it was used to stabilize the lateral atlantoaxial joints in cases of odontoid fractures. In 1975 Tucker introduced the concept of applying interlaminar clamps to the atlantoaxial joint, and in 1988 Holness described the first case of it. In 1979 Magerl introduced the transarticular screw technique, which, for the first time, enabled complete obliteration of the rotational movement and could be used when the posterior axis was damaged. In 1978 Brooks and Jenkins offered an alternative to address this rotational instability problem; two iliac crest grafts were placed between the C1 and C2 arches and stabilized with two wires, one on each side. In 1984 Goel introduced C1 lateral mass screws and C2 pedicle screws (C1LMS, C2PS); subsequently Harms and Melcher substituted the plates used by Goel with rods and contributed to popularization of the method [14]. In 1991 Dickman and Sonntag published a modification of the wiring techniques in an attempt to solve the disadvantages of the two previously described techniques by using only one self-locking titanium cable [15]. In 2002 Benzel et al. published a different entry point: through the posterior arch. In the same year Wright developed the translaminar screw (TLS) for C2; such a procedure is less stable biomechanically and poses a very small risk to the vertebral artery. Several clinical studies have been reported, stating that the results achieved by lateral mass screw implants are better than those achieved by sublaminar instrumented wiring [16]. However, complications reported at the very beginning of the experience (e.g. 30% of screws pulling out in the suboccipital area and a mortality rate of up to 9% after complex spine decompression and fixation) discouraged mainly paediatric spine surgeons [17].Very interestingly, a 100% rate of fusion and a 10.4% rate of

complications, including vertebral artery injuries, have been reported with lateral mass screw implantations in a paediatric population [14]. Otherwise, both lateral mass screw implantations and sublaminar instrumented procedures have apparently resulted in the same 100% rate of fusion in more recent experiences [17]. According to our personal experience, good mechanical properties with low complication rates are offered by sublaminar instrumented wiring procedures, and so far they can be considered an excellent simple method for stabilizing the upper cervical spine and the CVJ. Wiring, augmented with external immobilization, produces good stiffness, according to the number of vertebrae enclosed in instrumentation, and can be associated with a 100% rate of bone fusion. Despite clear advantages of screwing techniques in terms of blood loss, surgery duration and postoperative immobilization, the complication rates in terms of infection seem to be similar [18].

Oblique Approaches

These are the anterolateral (extreme lateral) and posterolateral (far lateral) approaches.

In 1972 Hammon first described the *far lateral approach* (posterolateral approach) (FLA) for vertebral artery aneurysms; this approach has since undergone numerous modifications, including drilling of the occipital condyle, removal of the laminas of the upper cervical vertebrae and so on. Also, the range of indications has increased exponentially. George described a vertebral artery (VA) medial mobilization from C2 to its dural entrance point, with ligation of the sigmoid sinus and without condyle drilling [19]. The FLA, as originally described, is a surgical approach through a lateral suboccipital route, directed behind the vertebral artery and the sternocleidomastoid muscle, just medial to the atlas, the occipital condyles and the atlanto-occipital joints. The standard suboccipital approach can be considered the precursor of the FLA, which was conceived in order to maximize the exposure of the CVJ, mainly in the lateroventral aspect. Bone removal improves the angle of view by removing the posterior arch of C1 and the most lateral part of the inferior occipital squama, following a basic principle of cranial base surgery. Various portions of the occipital condyle are drilled, thus increasing the exposure. On rare occasions the vertebral artery is transposed in order to improve tangential lateroventral cervicomedullary area exposition for an unobstructed view of the CVJ more consistent with better management of the huge and heterogeneous spectrum of different pathological patterns involving this area.

The transcondylar, supracondylar and paracondylar approaches are considered expressions as well as variants of the FLA, all devoted to more effective surgical control of intradural, anterior and anterolateral CVJ lesions. Exposure of the upper cervical spine, the foramen magnum and the lower third of the clivus can be obtained without a significant decrease in the stability of the CVJ. Minimization of morbidity is possible because of watertight dural closure, far from contaminated regions.

Excellent exposure of extradural lesions located in the ipsilateral anterolateral aspects of the CVJ and anterior to the extradural region is provided by the far lateral atlanto-occipital transarticular approach. On the other hand, the so-called extreme lateral atlanto-occipital transarticular approach provides excellent surgical exposure extending across the midline to the medial aspect of the contralateral atlanto-occipital joint and the lower clivus.

The *extreme lateral approach (anterolateral approach)*, as originally described, is a direct anterolateral approach behind the internal jugular vein along the front of the vertebral artery and deep to the anterior part of the sternocleidomastoid muscle. This term dates back to 1990 when Sen and Sekhar depicted an alternative way to handle meningiomas and schwannomas located anteriorly at the cervicomedullary junction [20].

The extreme lateral approach (ELA) is generally considered a more aggressive extension of the FLA. By drilling the condyles at the atlanto-occipital joints also in the ELA, it is possible to widen the surgical view but with a different exposure angle because of the differences in the direction of the approach.

The FLA and ELA variants of the atlanto-occipital transarticular approach to the anterior extradural structures at the CVJ provide an effective alternative to the classic transoral approach. In fact, with both of the latter approaches described here, it is possible to (1) avoid the contaminated nasopharynx; (2) reduce the incidence of cerebrospinal fluid leakage; (3) provide a shorter surgical route; and (4) allow the lateral limitations of the atlanto-occipital joints to be overcome.

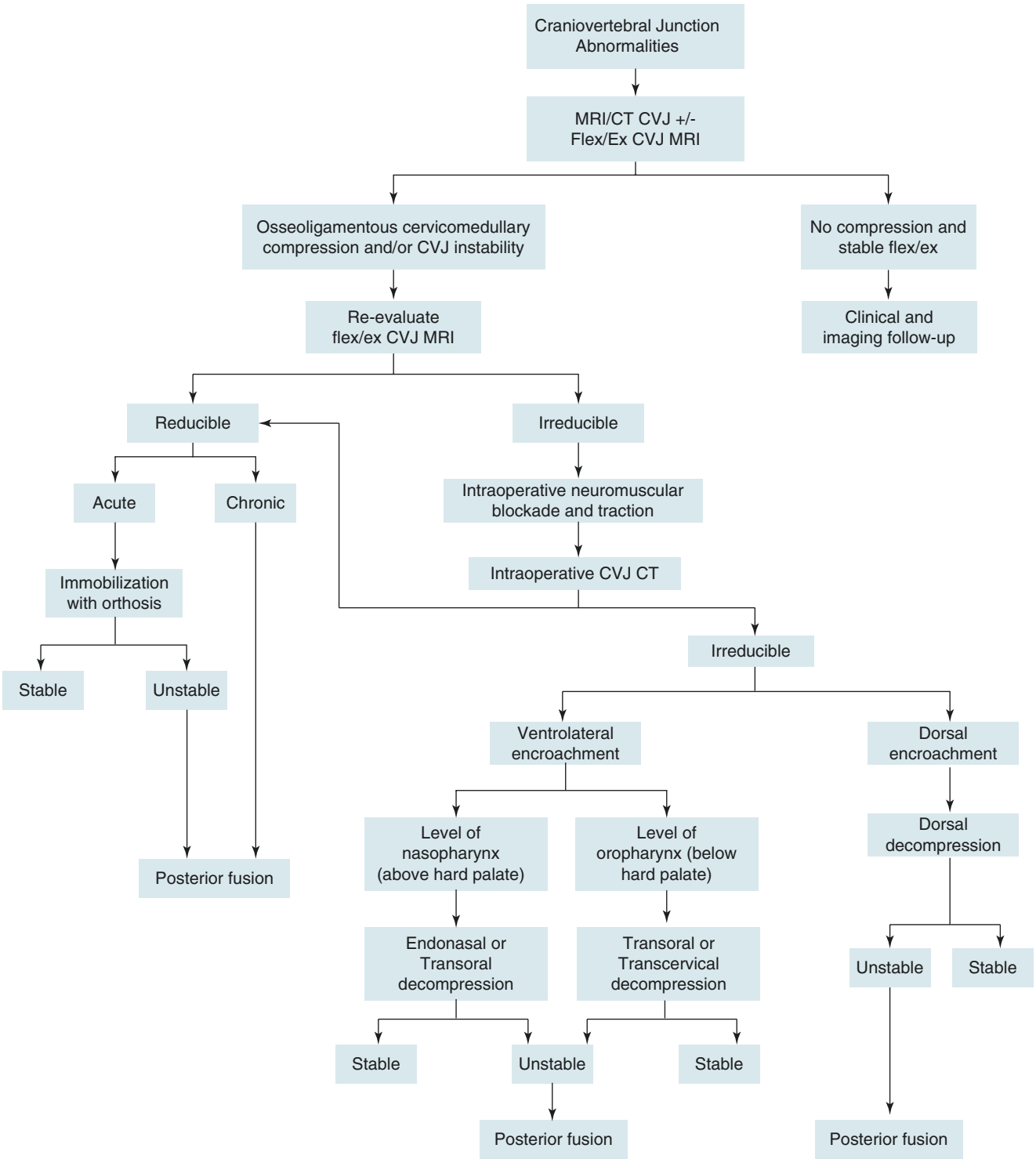
The posterolateral and anterolateral approaches and their modifications should be strongly considered in the armamentarium of all surgeons dedicated to approaching lesions of the CVJ.

Conclusion

Although it has appeared to be quite detailed, this brief tour around the world of the craniovertebral junction has provided merely a glimpse of this world, which is full of no man's lands that deserve to be better investigated and stories that need to be continued.

The craniovertebral junction is one of the most fascinating surgical fields of the future.

Menezes algorithm



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Competing Interests The author declares that he has no competing interests.

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Anatomy

The Craniovertebral Junction and Laboratory Experience: The Italian Paradox

Francesco Signorelli

Current neurosurgical training can rely on innovative methods that have been provided, especially in recent years, by the remarkable and constant evolution of surgical techniques, instruments and additional dedicated tools. Moreover, the increasingly widespread access to educational material in the forms of lectures, surgical videos and anatomical dissections represents an innovative and helpful resource, which is easily available to young surgeons. Nevertheless, the international scientific literature agrees that anatomical dissection of cadavers remains fundamental in the long process of neurosurgical education and constitutes an invaluable tool for anatomical and surgical research.

The craniovertebral junction (CVJ) is a complex anatomical region frequently affected by different pathologies, which can be either congenital or acquired. Because of its regional complexity, practical knowledge and full mastery of craniocervical anatomy—which can be mostly achieved only through direct anatomical dissection—are required to safely perform surgical procedures in this intricate area.

In this scenario, Italy nowadays represents a paradox: despite the secular tradition of anatomical dissection begun by Leonardo da Vinci and moved forward by authorities such as Vesalius and Golgi, surgical training in anatomical laboratories is an opportunity that is seldom exploited and is obstructed by the constraints of a lacking and fragmentary legislative framework and by the absence of body donation programmes for study and research purposes. Because it is so difficult to perform cadaver dissections in Italy—the cradle of anatomical studies in the sixteenth century—Italian surgeons often need to go abroad to attend rather expensive training courses in anatomical dissection laboratories.

The legal procedure to become a donor is hindered by some unresolved cultural issues such as the 'ownership' of cadavers, how consent is to be given during the donor's life and which Institution has the responsibility for conservation of the body [1]. The current legislation actually makes it impossible to use cadavers for anatomosurgical dissection. In particular, the main legal reference point dates back more than 80 years [2]. The article regulates religious sentiment and pity for the deceased, punishing anyone who dissects or, in any other way, uses a corpse or part of it for scientific or didactic purposes in cases not allowed by law. An almost contemporary decree (Regio Decreto of 31 August 1933) establishes that only bodies not recognized by the deceased person's relatives within six degrees of consanguinity can be used for scientific purposes. Such a regulation, which is still in force, is never applied in actual practice. Moreover, the legitimacy of using unclaimed bodies has exposed vulnerable groups to dissection without their consent [3]. In the absence of a consistent legislative framework, a number of centres for body collections and programmes for body donations have only recently been established [4].

A craniovertebral junction surgery research centre was established in March 2015 at the Catholic University School of Medicine in Rome (Rector's Decree No. 1674). The research centre and its laboratory are located at the Section of Legal Medicine and Insurance, Institute of Public Health, at the University. In this setting, to maximize the dissection experience and to overcome the lack of anatomical specimens, we have used—in addition to fresh cadavers—injected head and neck specimens:

1. *Fresh cadavers of individuals who have died between 24 and 48 h beforehand in non-traumatic circumstances, only in those cases in which a diagnostic examination is required and in which there is a specific indication to dissect the neck for forensic–legal reasons:* Special authorization has already been obtained from the ethics committee (protocol no. P663/EC/2010, approved on 28 July 2010; subsequent amendment no. P437/CE 2012, approved on 2 May 2012). Fresh cadavers have mostly been used to perform an anterolateral (extreme lateral) approach, transcervical retropharyngeal approach, and transoral or transnasal approaches to the CVJ.

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2. *Fresh-frozen specimens that are later injected with a silicone solution to perfuse the venous and arterial system, purchased from private companies:* With the aim of cost reduction, we have carried out all phases of specimen preparation (i.e. thawing, irrigation, fixation, perfusion and storage) according to a protocol developed at our research centre. On these specimens, posterior and posterolateral approaches to the CVJ, as well as other keyhole approaches, have been performed with the aid of neuronavigation, visual magnification of 3.5× offered by binocular lenses, a microscope and an endoscope.

To conclude, anatomical dissections play an irreplaceable role in the training of residents and young specialists, especially when they are approaching an anatomical region that is among the most complex in the entire body, such as the craniovertebral junction. Accordingly, promotion of donation programmes is crucial to allow Italian universities to keep up with the training provided by the most prestigious European and American higher education centres.

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Competing Interests The author declares that he has no competing interests.

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Mastering Craniovertebral Junction Surgical Approaches: The Dissection Laboratory Experience at the Catholic University of Rome

Francesco Signorelli, Vittorio Stumpo, Antonio Oliva, Vincenzo Lorenzo Pascali, Alessandro Olivi, and Massimiliano Visocchi

Introduction

The craniovertebral junction is an intricate anatomical region frequently affected by neoplastic, vascular, traumatic, congenital and degenerative pathology. Because the topography of this region is complex, direct knowledge and full mastery of craniocervical anatomy is mainly obtained through anatomical dissections performed in neuroanatomical laboratories.

In neurosurgery, scientific knowledge and manual skills must converge; therefore, anatomical dissection of cadavers remains the cornerstone of neurosurgical education and represents an invaluable tool for research.

Nowadays, in Italy—despite the secular tradition of anatomical dissection practised by pioneers such as Mondino de Luzzi, Leonardo da Vinci, Andreas Vesalius, Giovanni Battista Morgagni, Antonio Pacchioni, Luigi Rolando, Camillo Golgi and many others—surgical training in anatomical laboratories is rarely performed, being halted by the restrictions of disconnected legislative regulations and the absence of donation programmes for bodies to be used for study, training and research purposes.

In this paper we outline our experience in establishing an equipped craniovertebral junction laboratory for anatomical dissection, with a specific focus on some aspects concerning the preparation of specimens.

Methods

Laboratory Environment

Most of our equipment has been purchased with funds from the Craniovertebral Junction Surgery Research Centre, established in March 2015 at the Catholic University School of Medicine in Rome (Rector's Decree no. 1674), and from the "Master on Surgical Approaches to the Craniovertebral Junction" fund of the University. All phases of the studies are conducted at the Section of Legal Medicine and Insurance, Institute of Public Health, at the University. The Institute has two dissection stations, which mimic the operating room environment, and a dedicated computed tomography (CT) station. Each dissection station houses a scialytic lamp with a video capture unit, which allows recording of both still images and video for archival purposes. The laboratory is equipped with the following instruments: binocular lenses (visual magnification 3.5×), 420 mm; 0° and 30° rod lens endoscope (Storz, Tuttlingen, Germany); neuronavigation (Medtronic StealthStation Treon Plus); high-speed drill (Storz); vacuum aspirator (Super Vega Battery); digital camera (EOS 7D telescopic lens image stabilizer ultrasonic macro 100 mm; Canon, Tokyo, Japan); operating microscope (Zeiss, Oberkochen, Germany); microsurgical instruments; and stainless steel headholder.

Cadaver Specimens

With the aim of providing the best possible dissection experience, two kinds of anatomical specimens are used: fresh cadavers and injected 'head and neck' specimens.

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Fresh Cadavers

These are cadavers of individuals who have died in non-traumatic circumstances between 1 and 2 days beforehand and for which a diagnostic examination is needed, with a specific interest in the neck area. Special authorization has already been obtained from the ethics committee (protocol no. P663/EC/2010, approved on 28 July 2010; subsequent amendment no. P437/CE/2012, approved on 2 May 2012). The surgical approaches (all targeting the CVJ region) that are performed on the fresh cadavers are the anterolateral (extreme lateral) approach, transcervical retropharyngeal approach, and transoral or transnasal approaches.

Injected 'Head and Neck' Specimens

Fresh-frozen specimens have been purchased from private companies, but, to reduce the costs involved in the necessary specimen preparation, we have directly performed all of the processing—including the phases of thawing, irrigation, fixation, perfusion and storage—according to a protocol developed at our research centre. The thawing solution and the phases of preparation of the specimen are summarized in Table 1 and in Figs. 1 and 2.

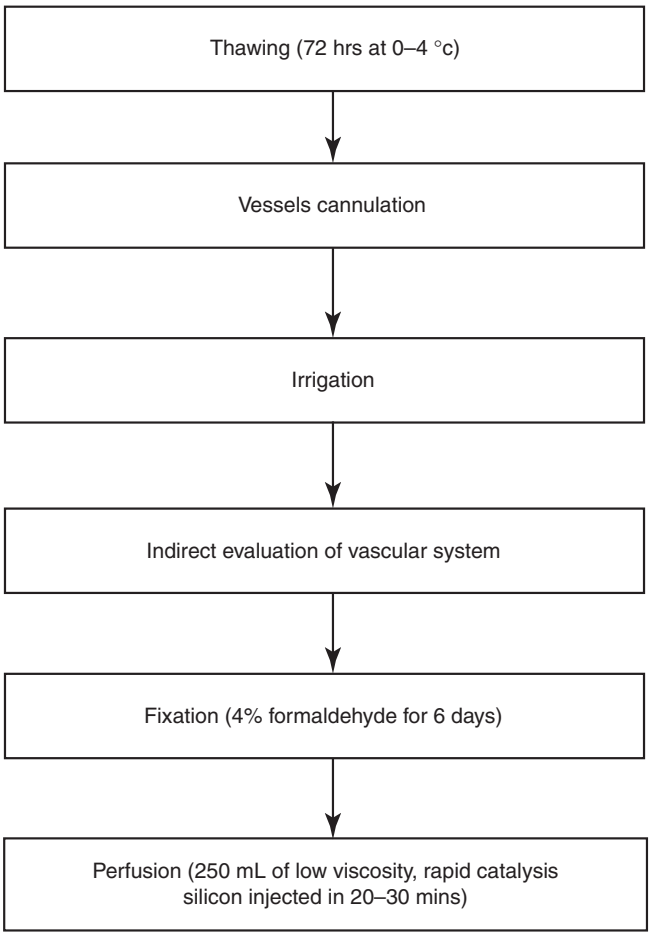

Fig. 1 Phases of preparation of anatomical specimens

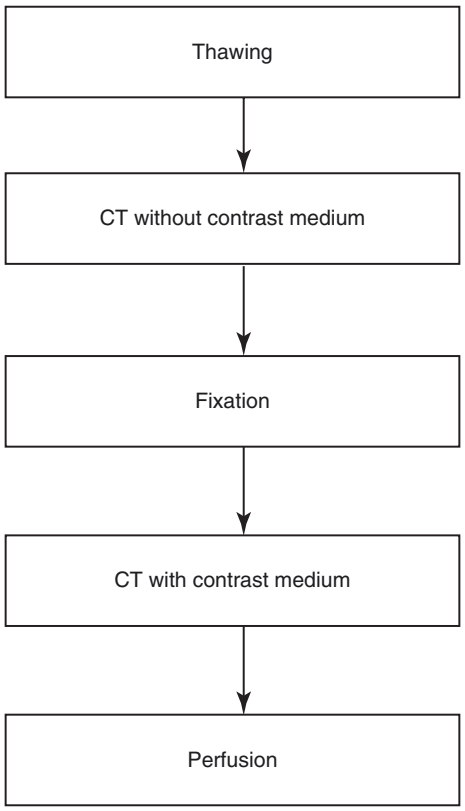

Fig. 2 Timing of execution of computed tomography (CT) scan of anatomical specimens

The purchased specimens have been serologically tested and certified for human immunodeficiency virus (HIV)-1, HIV-2, hepatitis B and hepatitis C; death certificates and informed consent from the donors' families have previously been obtained in accordance with the Uniform Anatomical Gift Act and in compliance with all national and international laws governing the recovery and distribution of anatomical specimens.

We perfuse the vascular system with coloured silicone solutions: blue-coloured for the venous system and red-coloured for the arterial system. Before being perfused, the formalin-fixed specimens undergo a high-definition CT scan with Iomeprol (Iomeron®) 375 mg/mL—a monomeric contrast medium iodinated, diluted at a ratio of 3:1 and injected at an approximate rate of 1 mL/s. The imaging data (saved in DICOM [Digital Imaging and Communications in Medicine] format) are stored on compact disc (CD) and imported into the neuronavigation workstation (Medtronic Treon) to perform three-dimensional reconstructions.

Then—with the aid of neuronavigation, the visual magnification of 3.5× offered by the binocular lenses, the microscope and the endoscope—posterior and posterolateral approaches to the CVJ, as well as other keyhole approaches, are performed.

Discussion

Although they were probably introduced for cultural and religious reasons, embalming techniques have ultimately been shown to have huge scientific and educational value. Embalming prevents putrefaction of corpses, and dissection can be done in a methodical way and shown to other people. In our studies the use of a solution of formaldehyde, ethanol and glycerol for thawing preparations and for conservation after fixation, together with a low concentration of formaldehyde (4%), results in an optimal state of preservation and hydration of the specimens. The injection of silicone solutions with low viscosity and quick hardening allows optimal vascular perfusion, extending also to the microcirculation.

Anatomical dissections have always provided a useful tool for neurosurgeons to enhance their anatomical knowledge and microsurgical skills [1–3].

The addition of a surgical skills laboratory can augment the surgical training experience, as trainees are able to gain better understanding of surgical anatomy and approaches without the constraints of the operating room atmosphere. Moreover, by exploiting adjunctive image guidance systems such as the endoscope and neuronavigation, which are now consolidated instruments of the neurosurgical armamentarium, the quality and learning outcomes of anatomical dissection can be enormously enhanced. Different points of view provided by the microscope and the endoscope can help in acquiring better spatial orientation, especially in a deep and curvilinear space such as the CVJ [4].

In addition, neuronavigation can provide real-time positional information, which allows precise localization, thus dramatically improving the reliability of the approach [5]. An essential condition for neuronavigation is execution of a high-quality CT scan of the anatomical preparations; for the sake of better identification of vascular structures—that is, preliminary to the execution of the surgical approach—we perform a study with contrast medium by injecting a mixture of water and iodized water-soluble monomeric low-viscosity contrast medium, with satisfactory impregnation not only of the vascular tree but also of the subarachnoid cisterns. Few studies have reported the use of contrast medium for cerebral anatomical preparations, and in many cases they have used solutions—generally with high viscosity—that were detrimental for the impregnated preparation, which was thus eventually no longer available for anatomical dissection [6]. This event has not occurred in our experience.

Conclusions

Our studies confirm the paramount importance of anatomical dissections in the training of residents and young neurosurgeons, with particular reference to complex spinal surgery at the craniovertebral junction level because of its inherent anatomical complexity.

Further studies are needed to develop an optimal impregnation contrastographic protocol for the study of cadavers or anatomical specimens for which a diagnostic examination or anatomical dissection is planned.

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Competing Interests The authors declare that they have no competing interests.

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Surgical Highways to the Craniovertebral Junction: Is It Time for a Reappraisal?

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Abstract *Background:* The craniovertebral junction (CVJ) can be affected by a variety of congenital or acquired anomalies. Because of its complexity, a careful evaluation of bones and ligamentous structures in all three planes is required. This can be achieved by studying the CVJ in terms of several anatomical and radiological lines that have been visualized to facilitate understanding of its surgical anatomy. In this study we aimed to review the state-of-the art craniometric CVJ lines and approaches.

Methods: In December 2016 a PubMed search was performed, including the search terms 'CVJ surgical approach/line', 'cervical approach', 'craniometric measurement', 'CVJ anatomy' and 'ventral/dorsal/far-lateral approach'. Anatomical and radiological lines and angles evaluated on traditional radiography, computed tomography (CT) scanning or magnetic resonance imaging (MRI) in the axial/sagittal/coronal views were included and described.

Results: Several measurements and radiological landmarks were included to evaluate the anatomy of the CVJ. They were fully described and categorized on the basis of the anatomical plan and the surgical or diagnostic purpose they are used for.

Conclusion: Among the numerous radiological measurements described, it has been shown that McRae's line, Chamberlain's line, McGregor's line, the Redlund-Johnell method and Ranawat's line are the most widely used and reliable ones for evaluating skull base craniometry. Secondly, the hard palate line (HPL), nasoaxial line (NAxL) and palatine–inferior dental arch line (PIA) are used to preoperatively assess the ventral endonasal or transoral surgical approaches. Thirdly, the C7 slope has been demonstrated as a reliable predictor of occipitocervical and spinopelvic alignment in CVJ fusion.

Keywords Cranio-vertebral junction · Surgical approach · Radiological lines · Skull base craniometry · Surgical angles

Introduction

The craniovertebral junction (CVJ) defines a complex anatomical area where the occiput, atlas and axis interrelate with each other. This area contains neural and vascular structures and gives motion to the skull and the upper segment of the spine [1, 2]. Its unique range of motion is ascribed to the articulation between segments and their anatomical properties. At the atlanto-occipital joint (C0–C1), the atlas joins the occiput through the occipital condyles and surrounds the foramen magnum. At the atlantoaxial junction (C1–C2), the atlas, lacking a vertebral body, joins the odontoid process of the axis. Thus articulated by the CVJ, the cervical spine range of motion consists of 33–47° in flexion/extension, 90° in rotation and up to 12.2° in lateral bending [1].

The CVJ can be affected by a variety of congenital or acquired anomalies that result in its instability; given its involvement in wide angles of neck movement, those conditions pose a challenging problem [3]. Among these anomalies, congenital bone malformations, intradural and extradural tumours, inflammatory diseases such as rheumatoid arthritis or ankylosing spondylitis, and traumatic injuries still represent a surgical challenge because of their high mortality and morbidity [4–6]. Since the onset of neurological symptoms related to the cervical spinal cord, brainstem, cerebellum, cervical nerve roots, lower cranial nerves, and vasculature represents a poor prognostic factor for those conditions, early surgery is advocated to safeguard the vital structures housed in the CVJ [7].

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Because of its complexity, the surgical anatomy of the CVJ and the skull base has attracted increasing interest in order to gain better access to lesions affecting this region [8]. For this reason, careful evaluation of bones and ligamentous structures in all three planes is required, through use of conventional radiography of the skull and upper cervical spine, and through computed tomography (CT) and magnetic resonance imaging (MRI). On the basis of imaging it is possible to measure several craniometrical indexes to estimate the best surgical approach [9].

Over the decades, several surgical routes and approaches have been proposed in order to gain functional and safe exposition of the CVJ. They can be divided into ventral approaches (transoral, transnasal, transmaxillary, transpalatal, transmandibular and extended open-door maxillotomy), the endoscopic endonasal skull base approach, the endoscopic endonasal transclival transodontoid approach [10], the far/extreme lateral approach (paracondylar, transcondylar) [8] and the dorsal approach (midline and dorsolateral) [11].

To diagnose and classify CVJ anomalies, to assess the severity of disease, to verify the feasibility of surgical approaches and to evaluate new routes for newer and more sophisticated surgical devices, several anatomical and radiological lines have been visualized and compared to facilitate understanding of CVJ surgical anatomy. In this study we aimed to review the state-of-the art craniometrical CVJ lines evaluated in surgical, anatomical and radiological studies.

Materials and Methods

The aim of this review was to identify and describe the most widely used and reliable radiological and surgical lines and angles for analysing CVJ anatomy. It was intended to provide a synoptic view of the craniometric landmarks and measurements that can be evaluated on conventional radiography, CT scanning and MRI, and on the surgical field, in order to provide an up-to-date diagnostic instrument for preoperative assessment in CVJ surgery.

In December 2016 a PubMed (http://ncbi.nlm.nih.gov/pubmed) search was performed, including the search terms ‘CVJ surgical approach/line’, ‘cervical approach’, ‘craniometric measurement’, ‘CVJ anatomy’ and ‘ventral/dorsal/far-lateral approach’. Cadaver studies, radiological retrospective studies and reviews were included. The criteria for exclusion were papers in which only surgical procedures or anatomical dissection were described without mentioning lines and landmarks, papers in which no anatomical line or craniometric landmarks were described, and papers not published in English. For papers that were considered potentially eligible, the full-text versions were sought. When the full-text versions had been obtained, the papers were further investigated for eligibility: only anatomical and radiological lines and angles evaluated on traditional radiography, CT scanning or MRI in the axial/sagittal/coronal views were included and described.

Results

A wide variety of anatomical landmarks and radiological lines were found. The most clinically useful and frequently employed ones in radiological and neurosurgical practice are discussed below.

Assessment of Craniovertebral Junction Anatomy and Surgical Approaches

Sagittal Plane

McRae’s Line

This is a line drawn across the foramen magnum from the tip of the anterior margin (basion) to the tip of the posterior margin (opisthion), and it determines the anteroposterior dimension of the foramen magnum [7, 9, 12–15].

Chamberlain’s Line

This is a line drawn from the posterior edge of the hard palate to the posterior border of the foramen magnum (opisthion). Usually the odontoid process is located ±3 mm around this line. When it extends more than 6 mm above Chamberlain’s line, this signifies basilar invagination [2, 7, 9, 12, 14, 15].

McGregor’s Line

This is a line drawn from the posterior pole of the hard palate to the lowest point of the occipital curve on the midline. This makes it easier to identify on a standard lateral radiograph than Chamberlain’s line. The mean distance between McGregor’s line and the odontoid tip is 1.45 mm in males and 0.44 mm in females; a distance greater than 4.5 mm signifies a diagnosis of basilar impression [2, 7, 12, 14, 15].

Ranawat’s Line

This is a perpendicular line drawn from the centre of the C2 sclerotic ring (representing the radiographic projection of the pedicles) to the transverse axis of the atlas, measured along the axis of the odontoid process on conventional radiographs. The mean length is 17 mm in males and 15 mm in females. The midpoint of the C2 base is used as a landmark on CT scanning since the C2 sclerotic ring is not applicable (the modified Ranawat’s line) [7, 14, 15].

Modified Ranawat's Line

This is a perpendicular line between the midpoint of the base of the C2 end plate and the transverse axis of the atlas, drawn from the centre of the anterior arch to the centre of the posterior arch of C1 [7, 14].

Wackenheim's Clivus Base Line

This is a line drawn along the clivus and extending inferiorly into the upper cervical canal. It should be tangential to the posterior aspect of the tip of the odontoid process. When it intersects with the body of the odontoid process, this signifies anterior cervical dislocation [2, 7, 9].

Redlund-Johnell Method

This method measures the minimum distance between the midpoint of the base of the C2 end plate and McGregor's line [7, 14].

Odontoid Length

This is the distance from the midpoint of the base of the C2 end plate to the odontoid tip [7].

Clivus–Canal Angle

The clivus–canal angle (CCA) is the angle formed by Wackenheim's line and a line drawn along the posterior surface of the odontoid process and the inferodorsal portion of the C2 body. This angle ranges from 150° in flexion to 180° in extension. If this angle is less than 150°, ventral spinal cord compression may occur [2, 9, 16].

Occipitocervical Angle

This is an angle measured at the intersection of a line drawn along the inferior end plate of C2 and the occiput line, or alternatively using Chamberlain's line, McRae's line or McGregor's line. It has been shown that the occipitocervical angle is most reliably reproduced when McGregor's line is used [17].

Effective Canal Diameter

The effective canal diameter (ECD) is the distance from the posterior surface of the odontoid process to the nearest posterior bony structure (the foramen magnum or the posterior arch of the atlas). The ECD is measured on CVJ CT scans in a neutral view and represents the space occupied by the buffer space (which permits neck movements without compromising neural structures) and the cord itself [18].

Welcher's Basal Angle

This is the angle formed at the intersection between the nasion–tuberculum line and the tuberculum–basion line. On average it measures about 132° and should be not more than 140°. A wider angle results from abnormal skull base flattening [2, 16].

Clivodens Angle

This is the angle formed by the intersection of a line drawn along the long axis of the clivus and one drawn along the long axis of the odontoid process on a midsagittal reconstructed CT scan. Its mean value is 135.8°, and a diagnosis of basilar invagination is suggested by an angle less than 125° [19].

Atlantodental Interval

The atlantodental interval (ADI) is the distance between the posterior border of the anterior arch of the atlas and the anterior border of the odontoid process, measured on a midline reconstructed CT scan [16]. An ADI greater than 3 mm in adults and 5 mm in children is diagnostic of atlantoaxial dislocation [9].

Posterior Atlantodental Interval

The posterior atlantodental interval (PADI) is the distance between the posterior border of the odontoid process and the anterior border of the posterior arch of the atlas. On average it is greater than 13 mm; a measure shorter than 13 mm is suggestive of canal narrowing [9].

Grabb–Oakes Method

This measures the distance from a line perpendicularly drawn from the most posterior region of the dura mater covering the odontoid process to a line drawn between the inferior surface of the basion and the posteroinferior aspect of the C2 vertebral body. It is used to measure ventral cervicomedullary encroachment by the odontoid process [16].

Transcondylar Angle

This is the angle measured between a reference line, drawn tangentially to the posterior aspect of the occipital condyle and to the posterior rim of the foramen magnum, and a line drawn from the medial edge of the nearest two-thirds condyle after theoretical drilling to the farthest point on the clivus, intersecting with the reference line [8].

Boogard's Angle

This is the angle measured between a line drawn from the dorsum sellae to the basion and McRae's line [20].

Coronal Plane

Atlanto-occipital Joint Axis Angle

This is the angle formed between lines drawn parallel to the atlanto-occipital joints, which usually intersect at the centre of the odontoid process when the condyles are symmetrical. On average it ranges from 124° to 127°, and it is wider in cases of occipital condyle hypoplasia [2].

Axial Plane

Paracondylar Angle

This is the angle measured between a reference line, drawn tangentially to the posterior aspect of the occipital condyle

and to the posterior rim of foramen magnum, and a line drawn from the posterior edge of the occipital condyle intersecting with the reference line [8].

Assessment of the Ventral Craniovertebral Junction Approach

Coronal Plane

Hard Palate (Palatine) Line

The hard palate (palatine) line (HPL) is a line drawn along the plane of the hard palate toward the craniovertebral junction on a sagittal CT image. It is used to determine the feasibility of the endonasal approach if a lesion lies above or under this line [10, 21].

Nasopalatine (Kassam's) Line

The nasopalatine (Kassam's) line (NPL) is a line drawn from the most inferior point on the nasal bone (rhinion) toward the posterior edge of the hard palate, extending to the craniovertebral junction, on the midsagittal radiological plane. It is used to assess the maximal inferior extension of dissection [10, 21–23].

Nasopalatine Angle

This is the angle created between the NPL and the plane of the hard palate (HPL), which defines the surgical exposure to the CVJ through the endonasal approach [22].

Nasoaxial Line

The nasoaxial line (NAxL) is a line drawn from the midpoint between the rhinion and the anterior nasal spine of the maxillarybone, running through the posterior edge of the hard palate to the C2 vertebral body [10, 21, 23]. It has been shown that the NAxL more accurately predicts the lowest limit of the endoscopic endonasal approach to the CVJ [24].

Mandibulopalatine Line/Palatine Inferior Dental Arch

The mandibulopalatine line/palatine inferior dental arch line (PIA) is a line drawn from the inferior dental arch to the posterior aspect of the hard palate, measured on a laterolateral conventional radiograph with a wide-open mouth [10].

Rhinopalatine Line

This is a line drawn from the point that corresponds to two thirds of the distance between the rhinion and the anterior nasal spine of the maxillary bone, to the posterior nasal spine of the palatine bone and then extends posteriorly and inferiorly to the CVJ [21].

Atlantosuperior Dental Arch Line

The atlantosuperior dental arch line (ASA) is a line drawn between the superior dental arch and the anterior base of the atlas on a laterolateral conventional radiograph with a wide-open mouth [10].

Superior Nostril Hard Palatine Line

This is a line drawn from the inferior edge of the nostril toward the posterior edge of the hard palate, extending to the craniovertebral junction, on the midsagittal radiological plane. Actually, it seems that this line is not a reliable predictor for the endoscopic endonasal approach, because it underestimates its inferior limit [24].

Coronal Plane

Inter-Eustachian Line

This is a line drawn between the Eustachian tubes, which corresponds to the atlantoaxial (C1–C2) joint. This line and the middle turbinates, on the aspect of the nasopharynx, represent surgical landmarks of the CVJ [23].

Assessment of Occipitocervical Alignment

Sagittal Plane

C2–C7 Cobb's Angle

This is an angle measured at the intersection of a line parallel to the C2 inferior end plate and a line parallel to the C7 inferior end plate [25].

C2–C7 Harrison's Angle

This is an angle measured at the intersection of a line drawn parallel to the C2 posterior border and a line drawn parallel to the C7 posterior border [25].

C7 Slope

This is the angle measured at the intersection between a line parallel to the superior end plate of C7 and a horizontal reference line measured on a midsagittal CT scan or laterolateral conventional radiograph. It has been proposed as a novel parameter for the assessment of spinopelvic alignment, since it is correlated with cervical alignment, the occipitocervical angle and the spinal slope [25].

C1–C2 Angle

This is the angle measured between a line drawn parallel to the inferior aspect of C1 and a line drawn below the inferior end plate of C2 [25].

Cervical Lordosis

This is the angle measured between a line drawn from the most inferodorsal to the most superodorsal part of C2 (the tip

of the odontoid process) and a line drawn along the posterior aspect of the C7 vertebral body [20].

Discussion

The CVJ is a complex anatomical structure, which can be involved in several primary malformations and acquired pathological alterations. Thus far, numerous measurements and craniometric landmarks have been proposed to assess the functional state of the CVJ, to plan the best surgical strategy and to follow up its anatomy after the procedure [15]. The former measurements, such as Chamberlain's, McGregor's and McRae's lines [12], used to be evaluated on plain conventional radiographs for the pathological first assessment and diagnostic purposes [15]. However, conventional laterolateral radiography makes it difficult to identify bony landmarks and to accurately evaluate CVJ measurements because of the low imaging resolution and the superimposition of several structures [7]. Therefore, multislice CT scanning and MRI have been tested for the assessment of CVJ anatomy and used instead of conventional radiography because of their superior resolution and three-dimensional views [12]. Using these techniques, the accuracy of the former radiological lines has been re-evaluated and new ones have been studied for both surgical and diagnostic purposes [7, 12].

In recent years, different studies using CT scanning and MRI have confirmed the surgical reliability of McRae's, Chamberlain's, McGregor's and Ranawat's lines, which have been demonstrated to be the most sensible and widely used ones in diagnosing CVJ anomalies and basilar impression [15]. The availability of CT and MRI allows better delineation of CVJ anatomy and better diagnosis of occipitocervical diseases, providing more detailed imaging of soft tissues and bony details [12]. In a cross-sectional imaging study by Kwong et al., McRae's line was demonstrated to be the most concordant one on both radiographic and CT measurements, although the identification of its landmarks seems difficult on plain radiographs [7]. However, McRae's line has been confirmed as the easiest measurement to use for assessing basilar invagination according to the distance of the odontoid tip (mean value 5.8 ± 1.6 mm) [7]. On the other hand, McGregor's and Chamberlain's lines (with mean values of the extension of the odontoid tip of 1.6 ± 2.8 mm and 2.3 ± 2.6 mm, respectively), which have mostly been used on plain radiographs, seem to be less reliable on CT scanning and MRI because of the frequent exclusion of the hard palate in the field of view [7]. Also, the Redlund-Johnell method (mean value 37.5 ± 4.0 mm) and the modified Rawanat's line (mean value 29.7 ± 2.6 mm) have found wide agreement on CT scanning with respect to the plain radiographic counterpart [7]. It has been demonstrated that some differences exist between CT measurement and radiographic measurement of Chamberlain's and McGregor's lines [7]. Moreover, McRae's and Ranawat's lines appear to be more reliable and easier to measure, for these reasons they should be preferred [15].

With regard to CVJ surgery, several anterior approaches have been proposed to expose the cranial base and the upper cervical spine [26, 27]. The ventral approach has been mostly carried out through the transoral route and has been improved in recent years by use of transnasal routes. The high risks of surgical complications and morbidity related to microsurgical ventral approaches have recently been overcome by the use of the endoscopic endonasal approach (EEA) and the transoral approach (TOA) to the skull base [10, 23]. The EEA provides wide access to the midline clivus and the CVJ, but entails the necessity of using surgical instrumentation with a precise range of motion in a confined space. In the preoperative assessment, the feasibility of the endonasal approach instead of the TOA can be determined by evaluation of the level of the lesion in regard to the HPL [10, 21]. Evaluation of the extension of the dissection in ventral endoscopic surgery can be performed on plain radiographs by open-mouth evaluation of the NAxL and PIA. While the NPL overestimates the caudal exposure of the CVJ and the superior nostril–hard palate line (HPL) underestimates it [24], it has been shown that the NAxL is a reliable predictor of the maximal caudal extension of the endonasal approach [23, 27]. In preoperative assessment of ventral CVJ surgery, the PIA reliably marks the cranial extension of the TOA [23].

With regard to CVJ stabilization and occipitocervical and spinopelvic interdependence, Matsunaga et al. reported a higher incidence of subaxial cervical malalignment in the case of occipitocervical fusion in an abnormal position [28]. On the basis of this evidence, a study by Núñez-Pereira et al. aimed to evaluate the range of compensatory mechanisms for successful occipitocervical fusion [25]. To identify these compensatory mechanisms and assess a reliable measurement to evaluate appropriate cervical alignment, several parameters were verified. Among these, the C7 slope was demonstrated to be a reliable and useful indicator of cervical alignment and sagittal thoracolumbar balance, since it was shown that it correlates with the C0–C2 angle, C2–C7 angle, spinal slope and C7 sagittal vertical axis, and acts as a link between the cervical and thoracolumbar spine [25].

Even if some of the relevant studies present several limitations, such as the lack of a standardized neck position [7], within the wide variety of radiological measurements and surgical landmarks, some of them deserve to be pointed out. In fact, Chamberlain's, McGregor's and McRae's lines, the Redlund-Johnell method, Ranawat's line on plain radiographs and the modified Ranawat's line on CT scans still represent the most widely used radiological measurements in the assessment of skull base craniometry and CVJ diseases,

whereas in the microsurgical or endoscopic ventral approach, the preoperative plan can be easily evaluated on plain radiographs of the skull and cervical spine using the HPL, NAxL line and surgical PIA as state-of-the-art ventral CVJ landmarks.

Conclusion

The craniovertebral junction (CVJ) is a complex anatomical area whose structures are involved in a wide angle of neck movement. It can be affected by several congenital or acquired anomalies that bring about CVJ instability. Thus, several approaches have been proposed to gain functional and safe exposition of the CVJ, and anatomical and radiological lines have been studied. Among these, McRae's line, Chamberlain's line, McGregor's line, the Redlund-Johnell method, Ranawat's line and the modified Ranawat's line are the most reliable ones for diagnosis of basilar invagination. Secondly, in the ventral approach to the CVJ, the hard plate line allows us to determine the feasibility of the endonasal or transoral approaches, while the nasoaxial line is used to predict the maximal caudal extension on the endonasal approach and the surgical palatine inferior dental arch line marks the cranial extension of the transoral approach. Thirdly, in occipitocervical fixation and in cervical spine surgery, the C7 slope can be easily evaluated and is a reliable predictor of occipitocervical and spinopelvic alignment, and of thoracolumbar sagittal balance.

Compliance with Ethical Standards

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Competing Interests

The authors declare that they have no competing interests.

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The Endoscopic Endonasal Approach to Craniovertebral Junction Pathologies: Surgical Skills and Anatomical Study

Paolo Pacca, Valentina Tardivo, Giancarlo Pecorari, Diego Garbossa, Alessandro Ducati, and Francesco Zenga

Abstract *Introduction:* Surgical anterior decompression is the treatment of choice for symptomatic irreducible ventral craniovertebral junction (CVJ) compression. Along with the classic transoral approach, the endoscopic endonasal approach has evolved and is gaining growing success.

Materials and Methods: In this work we discuss the surgical technique, give a complete step-by-step description of dissection of the craniovertebral junction and report a specific case of endoscopic endonasal odontoidectomy with use of a high-definition (HD) three-dimensional (3D) endoscope.

Discussion: The extended endonasal approach exploits an anatomical corridor to the odontoid process, involving only a small incision in the nasopharynx and sparing palate integrity. The most important limitation of the technique is 2D visualization, which hinders correct recognition of anatomical structures.

Conclusion: The endoscopic endonasal route to the odontoid process has proven to be a feasible, safe and well-tolerated procedure. Anatomical study is very important for better understanding of the 3D anatomy of the CVJ and relation of critical neurovascular structures to specific bony and muscular landmarks.

Keywords Odontoidectomy · Endoscopic · Endonasal approach · CVJ stability · Anatomical study

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Introduction

The craniovertebral junction (CVJ) is a complex transition area between the skull and the upper cervical spine, and it provides stability and motion [1–3]. Biomechanical studies have confirmed that the majority of spine flexion, extension and rotation occur at this level [4–6]. Moreover, vital neurological and vascular structures are housed in the CVJ. Different pathologies may affect the CVJ, leading to impairment/limitation of its physiological function with loss of mobility and eventually compression of neurovascular structures. Prompt surgical intervention is required when these disorders cause symptomatic high spinal cord or brainstem compression. Focusing on the pathology of the odontoid process, if such disorders are not treated, they may cause ventral brainstem and spinal cord compression with subsequent neurological deficits. In addition, if the ligaments are disrupted (especially the transverse ligament), instability and subassial atlo-occipital dislocation may occur. Rheumatoid arthritis, metastasis and congenital deformities represent the most frequent disorders involving the CVJ and the odontoid process [7]. In cases of irreducible ventral compression at the CVJ, anterior approaches offer direct access to the lesion without the need for neural tissue retraction, with a consequently low rate of lower cranial nerve injury [8]. Along with the classic transoral approach, the endoscopic endonasal approach has evolved and is gaining growing success. The latter, in fact, has been shown to be safe and effective, avoiding the need for tongue retraction, upper airway swelling, and the need for palate splitting [9]. The surgical field has anatomical limits dictated by the osseous structures of the region (the nasal and palatine bones), which form two lines: the Kassam line and the naso-axial line, which define a triangle-shaped surgical corridor [10]. The most important limitation of the technique is two-dimensional (2D) visualization, which hinders correct recognition of anatomical structures. The high-definition (HD) 3D endoscope is a new instrument that overcomes this problem, thus giving the transnasal approach enormous and growing

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importance in surgery in this region [11, 12]. This paper discusses our experience with the extended endoscopic endonasal approach (EEA) to the craniovertebral junction, using a specific case to provide a complete step-by-step description of the surgical technique used for the dissection, and discusses the advantages and limitations of this approach.

Dissection

Access to the foramen magnum and the CVJ requires a low trajectory in comparison with sellar approaches [13]. The posterior nasal septum, inferior sphenoid wall and vomer are removed so the rhinopharyngeal part of the clivus can be reached [14]. The inferior third of the clivus is drilled away to improve the exposure of anterior CVJ. The lateral limits of the exposure are determined by the carotid protuberance, foramen lacerum, vidian canal and Eustachian tube. The nasopharyngeal mucosa is removed, widely exposing the underlying basipharyngeal fascia and the median raphe covering the prevertebral muscles (Fig. 1). Two muscles cover this area anterior to the foramen magnum and extend from the clivus downward: the longus capitis and rectus capitis anterior (RCpA). The longus capitis is removed and has multiple bellies (Fig. 1). The atlantooccipital membrane (AOM) is a broad fibrous structure, which extends from the anterior edge of the foramen magnum to the superior edge of the anterior arch of the atlas (Fig. 2). The median raphe is a band of connective tissue, which is attached to the clivus and continues as the anterior longitudinal ligament (Fig. 2). It is necessary to remove the AOM and the anterior median raphe to expose the foramen magnum and the anterior arch of C1 (Fig. 3). The alar and

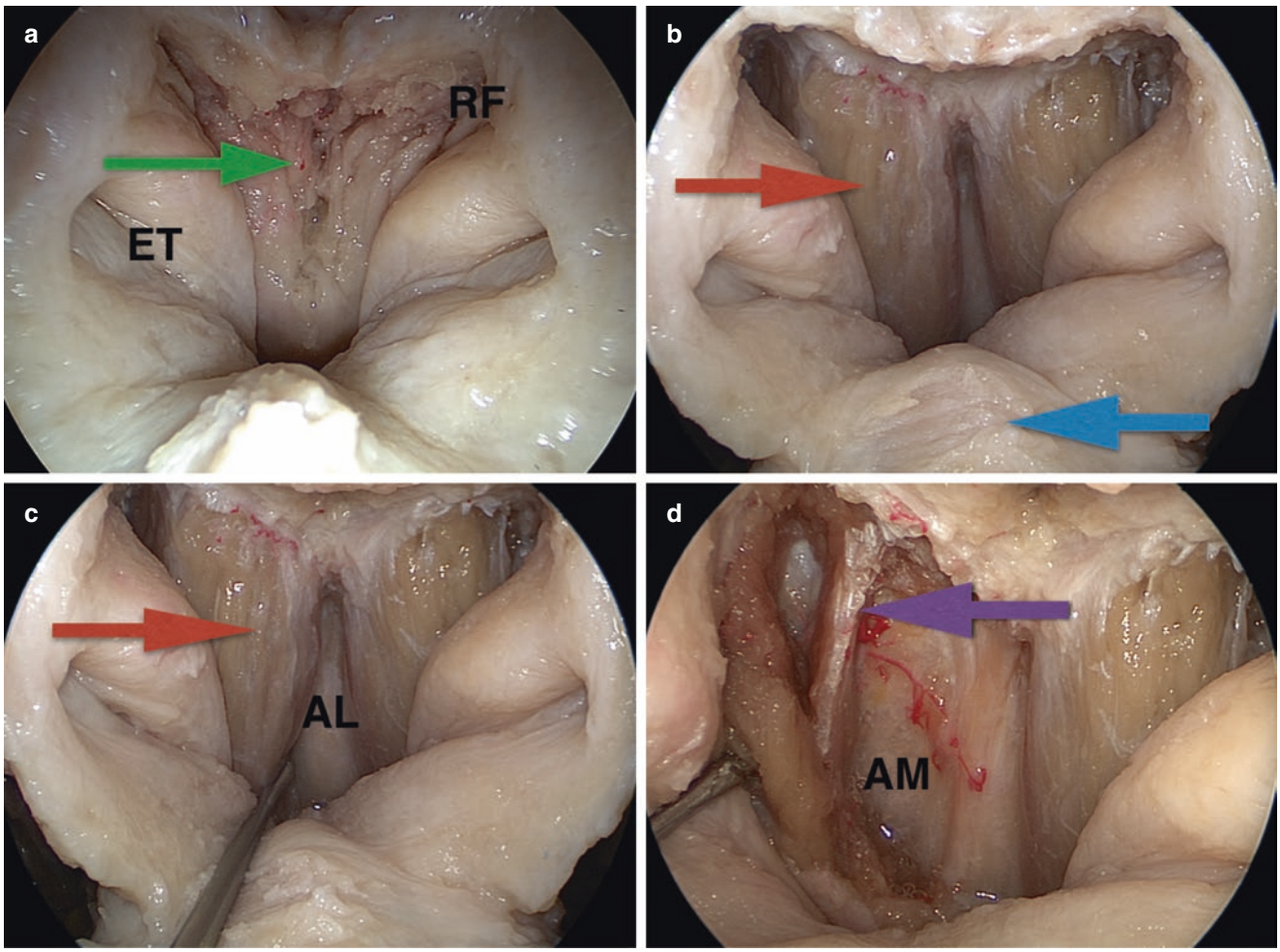

Fig. 1 Stepwise endoscopic approach to the foramen magnum and craniovertebral junction. (**a**) View of the mucosa of the rhinopharynx (*green arrow*) with the Eustachian tubes (ET) and the fossa of Rosenmüller (RF) as the lateral limits of this exposure. (**b**) Floor of the nasal cavity (*blue arrow*). The nasopharyngeal mucosa has been removed, exposing the basipharyngeal fascia overlying the longus capitis muscle (*red arrow*) and the median raphe attached to the pharyngeal tubercle on the midline (AL). (**c**) The longus capitis muscle (*red arrow*) can be seen after removal of the fascia. It is attached laterally to the pharyngeal tubercle along the superior clival line. (**d**) The longus capitis muscle has multiple bellies and is attached in layers to the clivus, as can be seen after partial removal from the right side (*purple arrow*). (AM) anterior atlantooccipital membrane

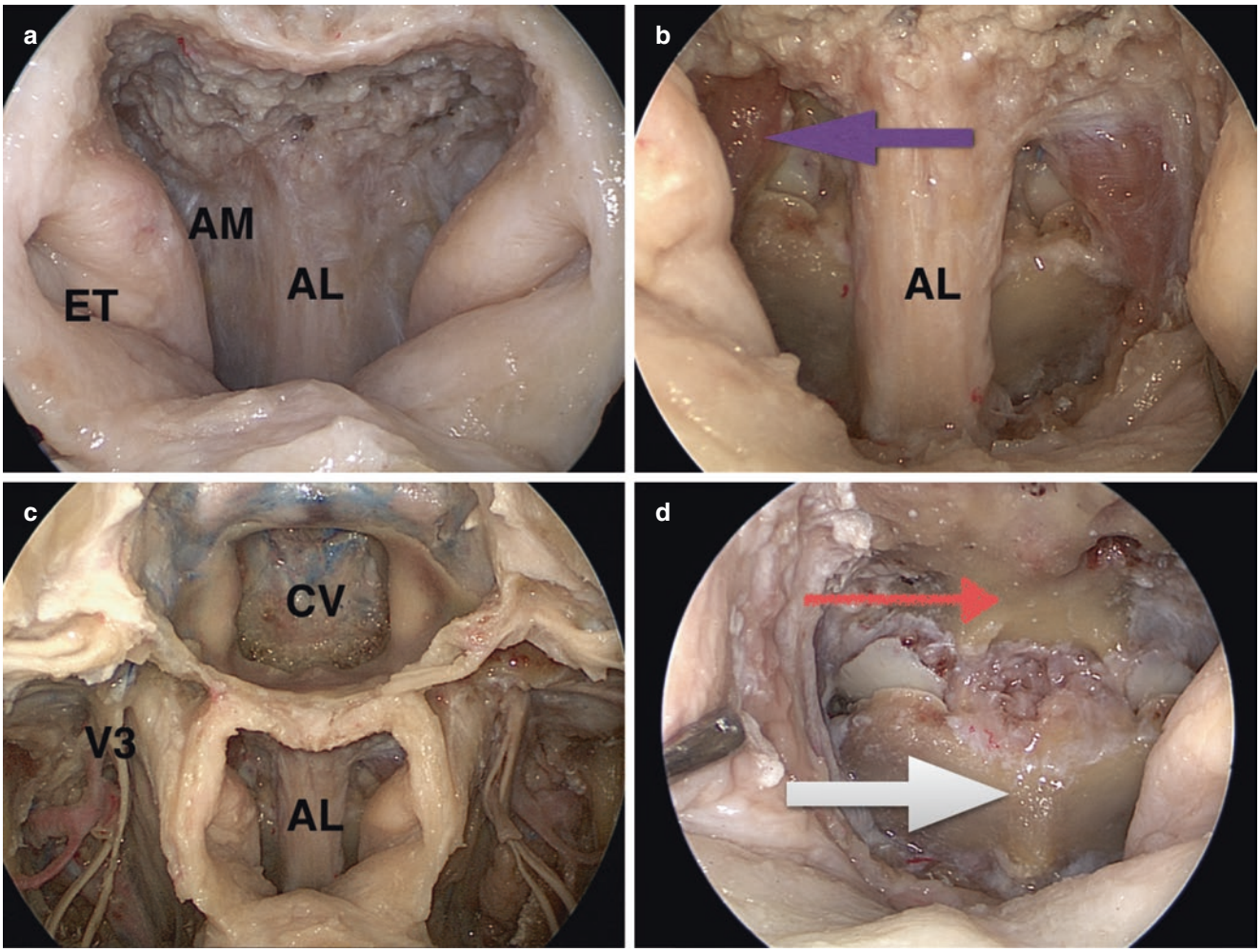

Fig. 2 Stepwise endoscopic approach to the foramen magnum and the craniovertebral junction. (**a**) Both sides of the longus capitis are removed, exposing the median raphe part of the anterior longitudinal ligament (AL) attached to the pharyngeal tubercle and laterally forming a thick, broad membrane called the anterior atlanto-occipital membrane (AM). (**b**) Both the longus capitis and the anterior atlanto-occipital membrane have been removed, exposing the anterior longitudinal ligament on the midline (AL) and the atlanto-occipital joint. The rectus capitis anterior muscle can be seen laterally attached along the clivus from the inferior clival line to the foramen magnum (*purple arrow*). (**c**) Extended endonasal approach to the clivus (CV); the sphenoid stage of dissection has been completed. On the right side, the infratemporal fossa has been dissected, showing the path of the Eustachian tube and the branches of the mandibular nerve in depth (V3). (**d**) The anterior longitudinal ligament and rectus capitis anterior have also been removed here, exposing the foramen magnum and the arch of C1 with both atlanto-occipital joints (*white arrow*). The gap between the C1 arch and the foramen magnum is filled with dense connective tissue, which also encloses the apical and alar ligaments attached to the dens of C2. The pharyngeal tubercle can be seen here (*red arrow*)

apical ligaments are located within the space between the foramen magnum and the anterior arch of C1. To view the odontoid process and the ligaments it is necessary to partly remove the anterior arch of C1 (Fig. 3). The atlas has two lateral masses: an anterior arch with a midline anterior tubercle, and a posterior surface (Fig. 3). The alar ligaments are fibrous bands arising from the lateral surface of the odontoid process and ascending toward the alar tubercle on the medial side of the occipital condyles (Fig. 3). The apical ligament of the odontoid process has a cartilaginous base arising from the apex of the dens and coming up to the anterior edge of the foramen magnum. The posterior surface of the dens also has a posterior facet, which articulates with the cartilaginous facet on the anterior surface of the cruciform ligament (Fig. 4). The anterior cortical surface and the core of the dens are drilled, leaving only a thin shell of bone, which can be removed with rongeurs. Removal of the dens exposes a thick, strong, white band called the transverse ligament, which arches posteriorly the dens and holds it in position. It extends from the tubercles on the medial side of the lateral mass of the atlas (Fig. 4). The transverse ligament is the horizontal part of the cruciform ligament, which has two upper and lower vertical bands. The tectorial membrane, a broad fibrous band that spans the area between the medial edges of the occipital condyles, is a rostral extension of the posterior longitudinal ligament,

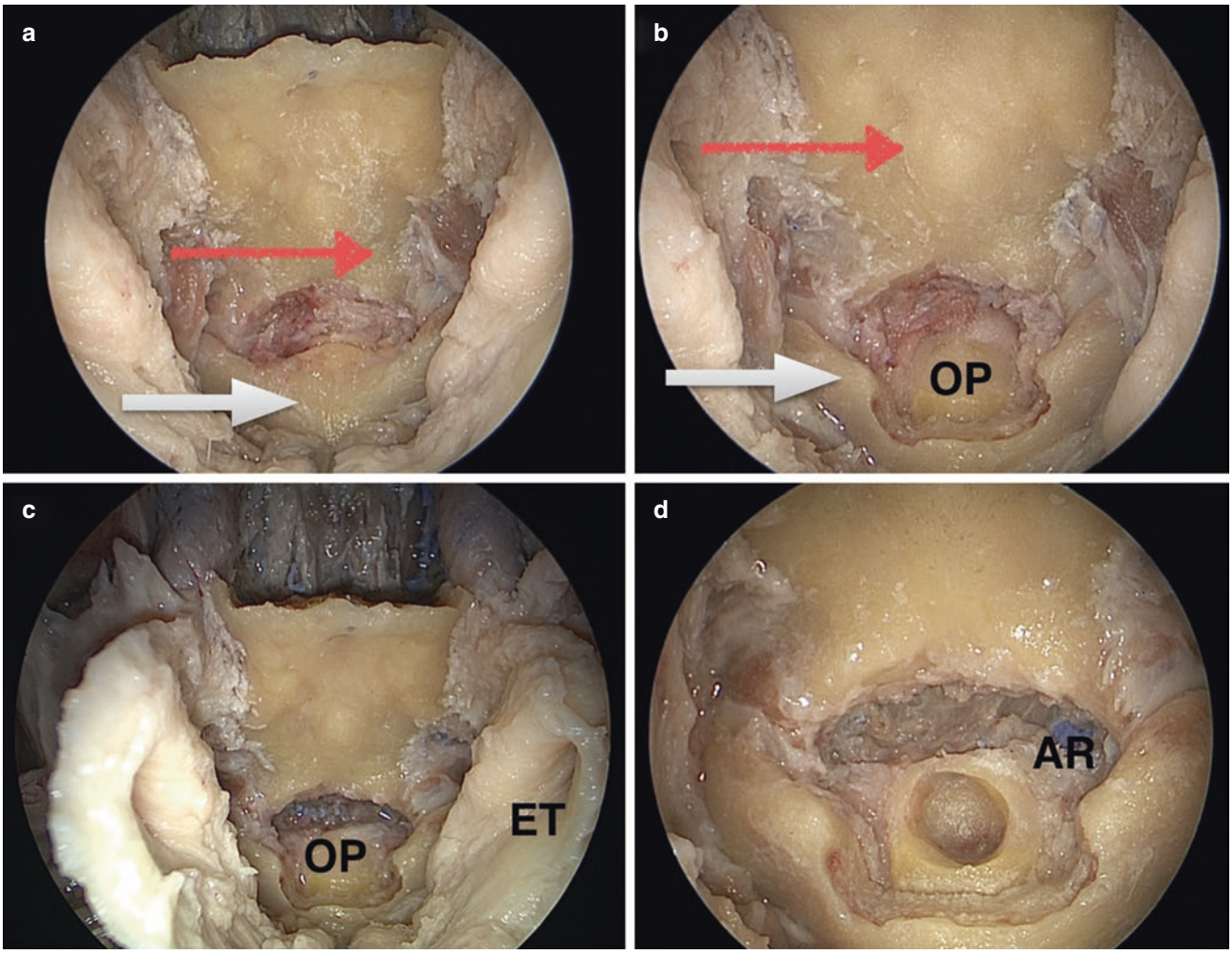

Fig. 3 Stepwise endoscopic approach to the foramen magnum and craniovertebral junction. (**a**) The foramen magnum after removal of the anterior longitudinal ligament and the anterior atlanto-occipital membrane. The supracondylar groove can be identified laterally with the rectus capitis anterior muscle (*red arrow*). (**b**, **c**) The clivus after removal of the muscles, showing the superior and inferior clival lines (*red arrow*). The inferior clival line corresponds to the supracondylar groove laterally. The anterior arch of C1 is partly removed (*white arrow*), exposing the odontoid process (OP). (**d**) The alar ligaments (AR), which are thick fibrous bands that attach to the posterolateral roughened surface of the odontoid process and ascend obliquely and laterally to attach the alar tubercles located on the medial side of the occipital condyles. Odontoidectomy begins with drilling of the central core, as seen here

which attaches to the axis inferiorly and the clivus superiorly (Fig. 4). It is separated from the dura by the epidural venous plexus. Once the dura is opened, the junction of the spinal canal with the medulla, which is defined as being at the level of the origin of the C1 ventral root, is exposed.

Case Illustration

The patient was a 71-year-old woman affected by an odontoid lesion. Because of radiological evidence of C1 myelopathy associated with both lower and upper limb weakness, she had undergone posterior C1–C2 laminectomy in November 2010 at another hospital.

After some months of disease stability she experienced worsening of limb weakness, with development of spastic paraparesis and gait impairment. Her reflexes were overreactive. She also complained of paraesthesia. Brain–cervical spine magnetic resonance imaging (MRI) (Fig. 5a, b) and computed tomography (CT) scanning (Fig. 6a, b) were performed. The radiological exams showed worsening of myelopathy without lesion growth.

Therefore, the patient underwent endoscopic endonasal odontoidectomy. The surgical procedure was uneventful, and postoperative CT scanning confirmed the success of the procedure. After 1 week a posterior C0–C2 arthrodesis was performed with an occipital plate and a C2 transcortical screw. Another cervical spine CT scan was performed to check for correct screw positioning.

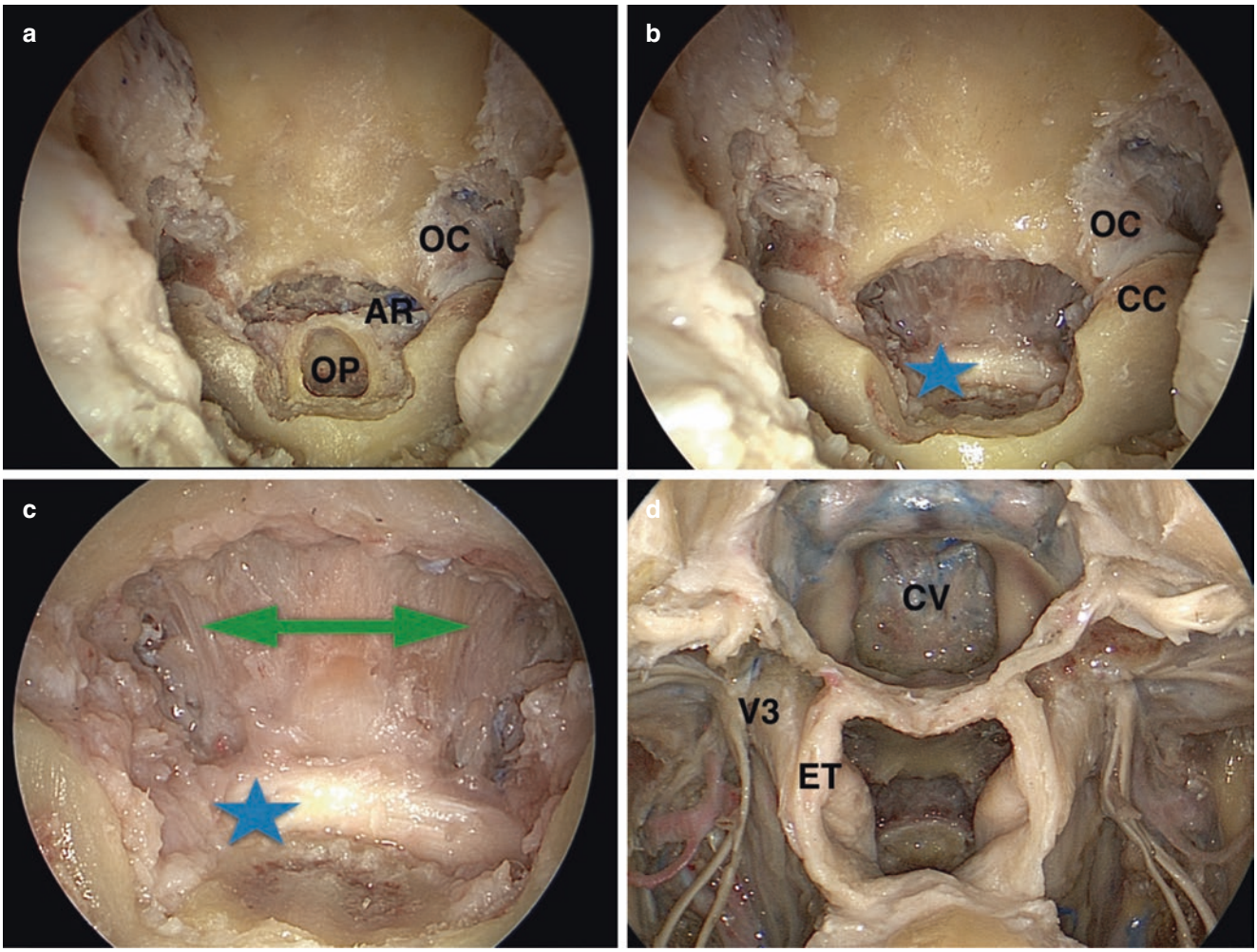

Fig. 4 Stepwise endoscopic approach to the foramen magnum and craniovertebral junction. (**a**) The alar ligaments (AR), which are thick fibrous bands that attach to the posterolateral roughened surface of the odontoid process and ascend obliquely and laterally to attach the alar tubercles located on the medial side of the occipital condyles (OC). Odontoidectomy begins with drilling of the central core, as seen here (OP). (**b**, **c**) Once the dens is removed, the transverse ligament (*blue star*) can be seen extending between the transverse tubercles on the medial side of the C1 lateral masses (CC). The tectorial membrane (*green arrow*) can be seen extending from the transverse ligament (*blue star*) to the foramen magnum. (**d**) Final image of the extended endonasal approach to the clivus (CV) when the sphenoid stage of dissection has been completed. The right and left infratemporal fossae have been dissected, showing the path of the Eustachian tube (ET) and the branches of the mandibular nerve in depth (V3). Complete dissection of the craniovertebral junction has been performed

The hospital stay was uneventful, and the patient was discharged after the second surgery. The histological diagnosis was an odontoid G2 chondrosarcoma. An intensive rehabilitation programme was useful to help the patient to reduce her gait impairment. MRI (Fig. 5c, d) and CT scans (Fig. 6c, d) were performed postoperatively, showing complete decompression of the dura. After 6 months a large area of arthrodesis was evident.

Surgical Technique

The procedure was performed with the assistance of neurophysiological monitoring. Somatosensory and motor-evoked potential data were collected for the patient. The surgical team was composed of a neurosurgeon and an ear, nose and throat surgeon. The surgery was done with a Visionsense III, 3D endoscope (Visionsense Ltd, Petach Tikva, Israel). Everyone in the surgical theatre wore passive polarized glasses to obtain a stereoscopic view [15, 16].

The patient was placed in a supine position with the head fixed in a three-pin head holder (Mayfield Infinity Skull Clamp; Integra LifeSciences Corporation. We positioned the head slightly tilted to the left on the coronal plane and slightly flexed to slide up the dens. All positioning manoeuvres were performed under neurophysiological monitoring. The nasal cavities were prepared preoperatively with 2 mL of 10% carbocaine (mepivacaine) with adrenaline, which was applied topically with patties. A binostril 4-hand technique was used routinely. Once the surgeons entered the

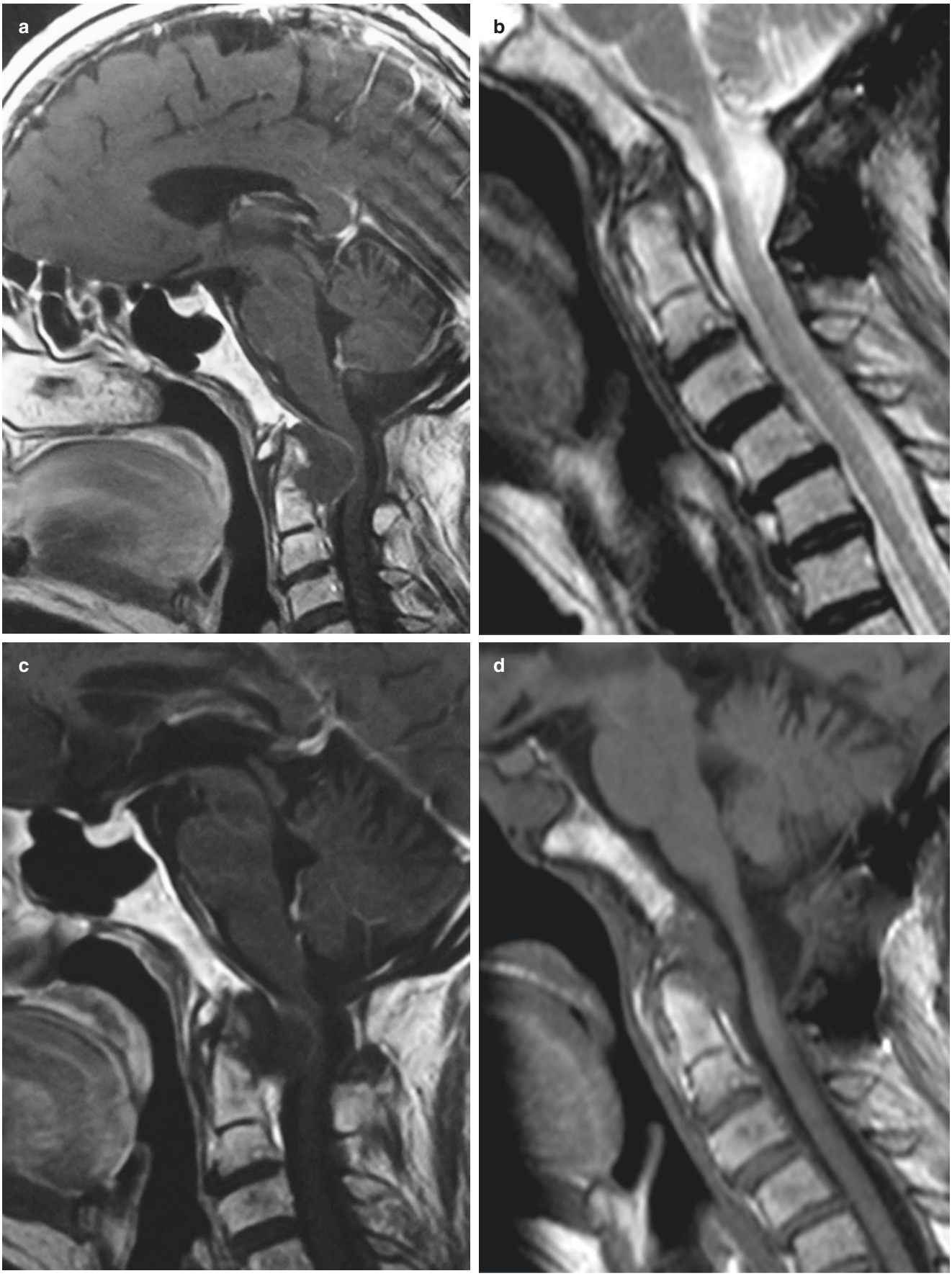

Fig. 5 (**a**, **b**) Preoperative magnetic resonance image (MRI) showing a complex craniovertebral junction malformation with a lesion of the odontoid process. (**c**, **d**) Postoperative MRI showing good spinal cord decompression

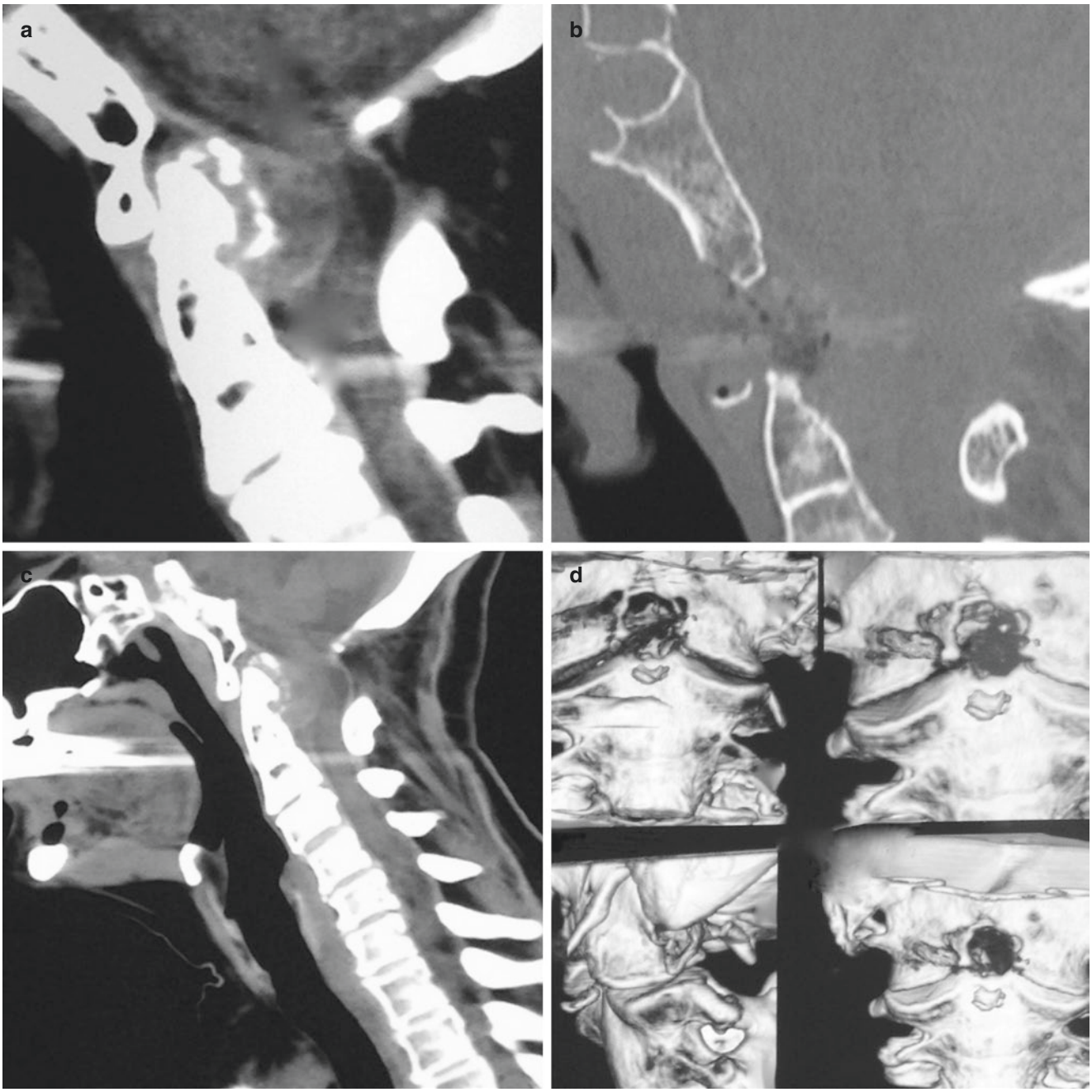

Fig. 6 (**a**, **b**) Preoperative computed tomography (CT) scan showing a complex craniovertebral junction malformation with a lesion of the odontoid process. (**c**, **d**) Postoperative CT scan showing good bone decompression

right nostril, the inferior and middle turbinates were displaced laterally to widen the surgical corridor (Figs. 7 and 8). The inferior margin of the middle turbinate, the nasopharynx, and the Eustachian tubes were the surgical landmarks that led the surgeons to the craniocervical junction. We usually do not remove the turbinates to preserve physiological airflow. The inferior margin of the middle turbinate was then followed until the nasopharyngeal cavity was entered. To enable use of both nostrils, the posterior third of the nasal septum was removed (Fig. 9). The junction between the clivus and the atlas was grossly defined by a line connecting the Eustachian tubes. The anatomical boundaries of the surgical field were defined by the floor of the sphenoidal sinus superiorly, the upper part of the oropharynx inferiorly, and the Eustachian tubes and the fossa of Rosenmüller laterally. The neuronavigation system provided significant help for identification of the anatomy at this point, especially concerning the vertical tracts of the internal carotid arteries. An inverted U-shaped flap of nasopharyngeal mucosa and muscular layers was harvested with the laser (CH fibre laser; Dornier MedTech, Munich, Germany) (Fig. 9). The Eustachian tubes had to be considered the most lateral margin of the incision because they identified the parapharyngeal segment of the carotid artery. The craniocaudal extension of the flap involved

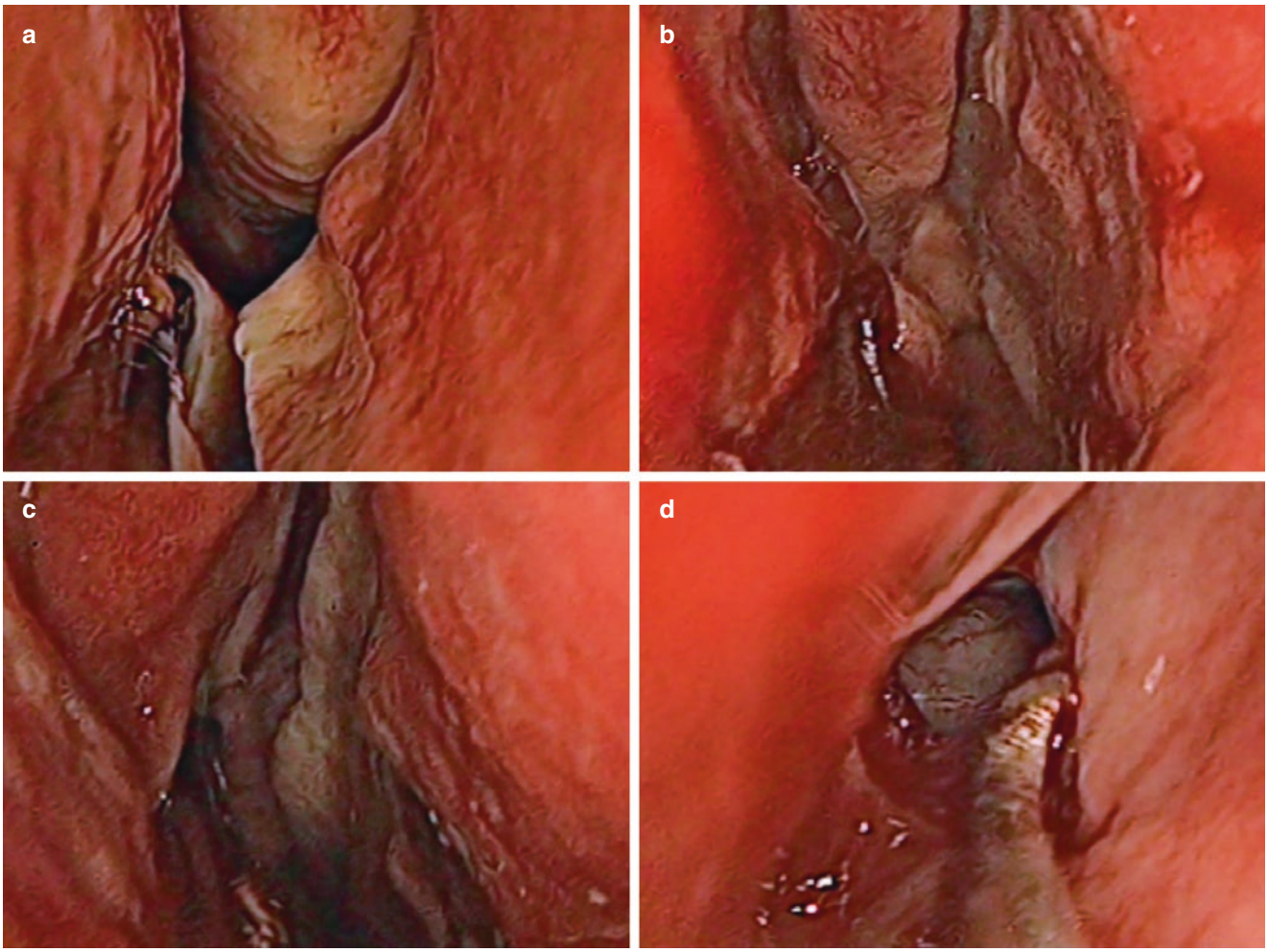

Fig. 7 (**a**, **b**) In the right nostril, the inferior and middle turbinates are displaced laterally to widen the surgical corridor. (**c**, **d**) Identification and initial opening of the ostium of the sphenoid sinus

the inferior third of the clivus superiorly and the C2 vertebral body inferiorly, and the lateral margin of the surgical exposure included the lateral masses of the C1 vertebra. Skeletonization of the anterior arch of C1 and the odontoid process was then carried out in a subperiosteal fashion. The lowest part of the clivus and the C1 anterior arch were then removed [17].

We checked the correct surgical position with the navigation system. A 3-mm coarse diamond burr was used to enter the anterior cortex of the odontoid process (extra-long high-speed microdrill; The Anspach Effort, Inc., Palm Beach Gardens, FL, USA). We proceeded with excision of pathological tissue between the odontoid process and the dura of the pontomedullary junction. An ultrasonic bone curette was then used to remove the tip of the odontoid process (Sonopet Omni ultrasonic surgical system; Stryker, Inc., Kalamazoo, MI, USA). Finally, the residual shell of the odontoid process could be removed by sectioning of the apical and alar ligaments, and separation of the process from adhesions to surrounding tissues [18].

Exposition of the dura anterior to the pontomedullary junction that was covered by inflammatory tissue. At the end of the procedure, image guidance was helpful for assessment of the effectiveness of the decompression. Furthermore, the integrity and the pulsatility of the dura were checked to rule out the risk of cerebrospinal fluid (CSF) leakage.

After the procedure, the inverted U-shaped mucosal nasopharyngeal flap that had been harvested at the beginning of the procedure was retrieved to cover the surgical cavity and fixed with fibrin glue. A Foley catheter was used to hold it in place for 2 days to promote the healing process. Both nostrils were packed with nasal swabs.

Discussion

'Craniovertebral junction' is a collective term that refers to the occipital bone, atlas and axis with supporting ligaments and membranes that provide stability and mobility to this

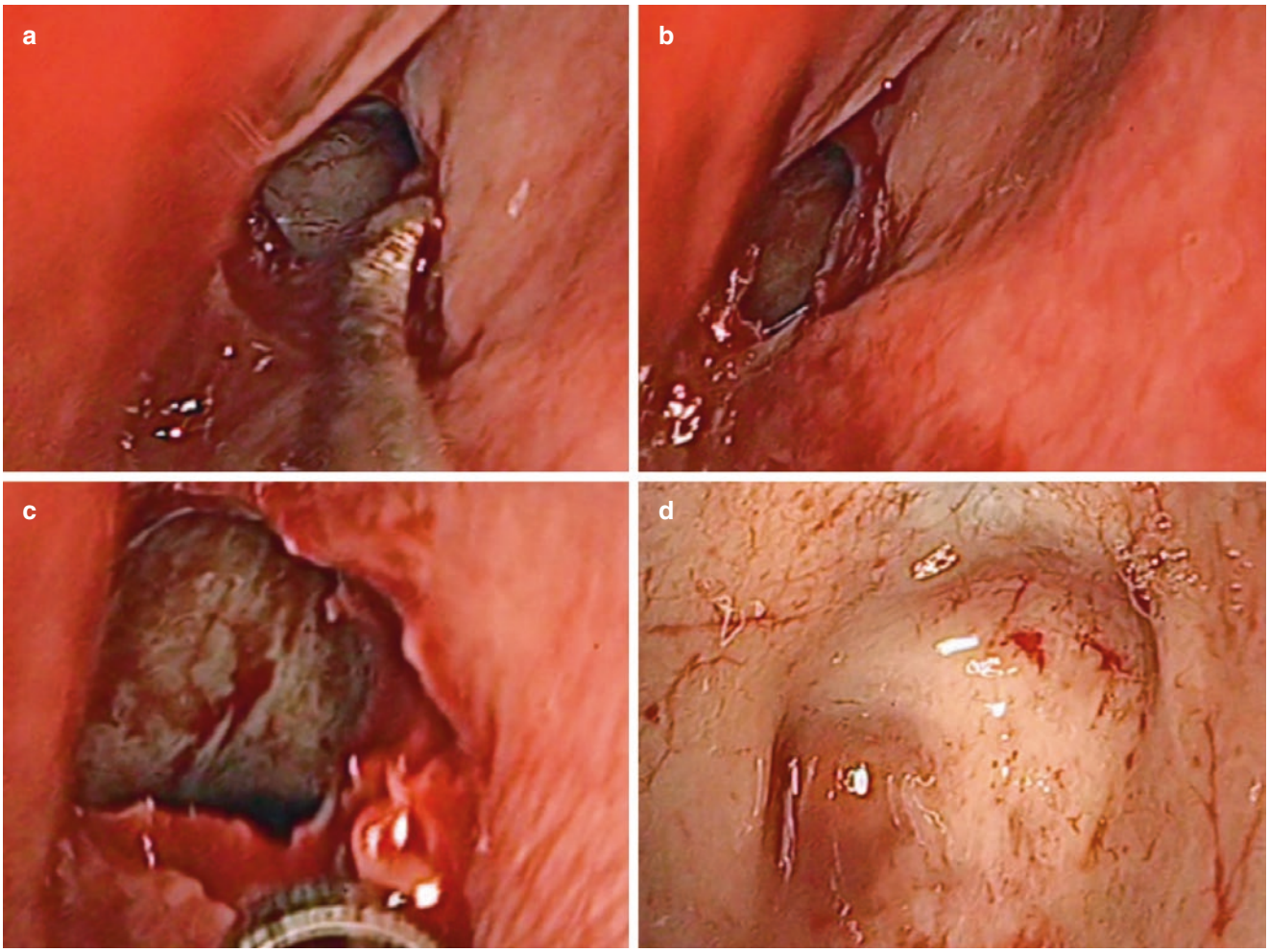

Fig. 8 (**a**, **b**) Opening of the ostium of the sphenoid sinus. (**c**, **d**) Identification of the sphenoid sinus and clivus

critical crossroads of the central nervous system at the brainstem transition to the spinal cord. Different surgical approaches have been described for exposing the CVJ, depending on the location of the pathology. If irreducible ventral compression of the brainstem occurs, an anterior route is the gold standard for obtaining straightforward access to the lesion without the need for neural tissue retraction [19]. The transoral, endonasal and transcervical approaches represent the three main options for performing odontoidectomy [4, 5, 8, 20]. The first approach to be reported was the transoral–transpharyngeal route to the ventral CVJ, described by Kanavel in 1917 [21]. Despite the wide surgical exposition (from the clivus to C2–C3) provided by this approach, it carries the risk of bacterial contamination, prolonged postoperative intubation, nasogastric tube feeding, tongue swelling and nasopharyngeal incompetence after transoral surgery [22]. The refinement of transsphenoidal endoscopic approaches to midline skull base lesions has led to the introduction of an endonasal route as a less invasive approach to the CVJ [23–25]. The extended endonasal approach exploits an anatomical corridor to the odontoid process, involving only a small incision in the nasopharynx and sparing palate integrity, without exposure to saliva and oropharyngeal bacteria, and therefore with a theoretically lower infectious risk [24, 26]. The endoscopic approach can provide access to the entire skull base, extending from the anterior cranial fossa to the body of C2 on the midline. The lateral exposure is limited here by the Eustachian tube, medial pterygoid plates and paraclival internal carotid arteries (ICAs). The muscles of the ventral CVJ are thin and have an avascular plane on the midline that can be used to retract them laterally or remove them as a U-shaped flap for later reconstruction. The anterior tubercle of C1 is a useful midline landmark, which can be confirmed with image guidance. A cadaver study by Baird et al. [27], comparing the endonasal, transoral and transcervical endoscopic approaches, showed that the EEA offers a shorter distance to the surgical target. Moreover, with the endonasal approach, the trajectory is entirely top down; thus, it is favourable to achieve better control of the drill and advantageous for detaching the ligaments while performing clivus and odontoid resection [27]. In addition, the endoscope provides supe-

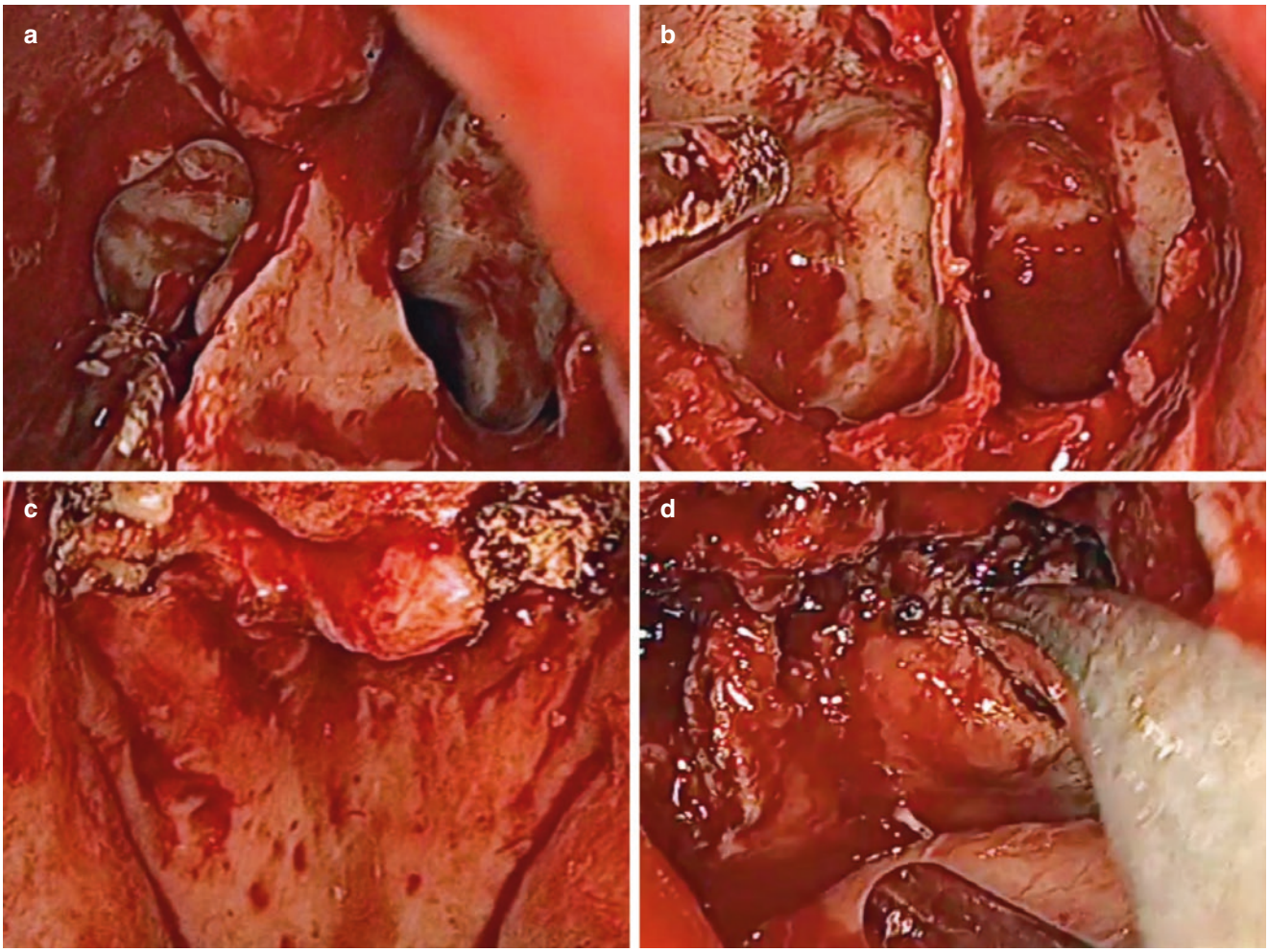

Fig. 9 (**a**) Rostrum of the sphenoid bone. (**b**) Clivus and paraclival carotid. (**c**, **d**) Inverted U-shaped flap of nasopharyngeal mucosa and muscular layers harvested with the laser

rior visualization and a larger optical field than the microscope does, which has turned out to be particularly advantageous in this deep surgical field. As highlighted by Ponce-Gómez et al. [3] in the odontoidectomy setting, the views differ between the endoscope and the microscope, and in such a narrow working area, movements are more fluent and faster with an endoscope. 3D endoscopes, which provide depth perception, may further improve the precision and safety of surgical manoeuvres [11, 15].

Maintenance of CVJ stability is another major issue to consider while performing surgical procedures at this level. Preservation of anterior C1 arch continuity by resection of only the odontoid process and pathological tissue—when feasible—reduces the risk of spinal biomechanical instability and the need for posterior fusion [9, 15]. One of the main criticisms of the EEA to the upper cervical spine is the limited exposure inferiorly. The K line, also known as the nasopalatine line (NPL), provides a window of caudal exposure inferior to the horizontal plane of the hard palate. As De Almeida et al. [26] demonstrated in their work describing extrapolation of the NPL to the vertebral column, the most inferior limit of dissection with endoscopic instruments is about 8.9 mm above the base of the C2 vertebral body. These findings suggest that while removal of the C2 body cannot be performed with the EEA, odontoidectomy usually is feasible with the EEA. It is currently thought that the surgical approach has to be tailored to the unique anatomical setting of each patient and the features of the lesion. On the basis of our own surgical experience, drilling of the inferior nasal spine, located between the soft and the hard palate, can be useful to widen the route of access to the CVJ, to gain more caudal access [16]. Further inferior dissection may require the use of angled instruments or a transoral approach [26]. Another drawback of the EEA may be the steep and long learning curve that this complex approach requires. Last—but not least—for a skull base neurosurgeon, being familiar with the EEA means not only being able to perform it but also being able to deal effectively with the complications that may derive from this approach.

A recent review by Shriver et al. [6] stated that postoperative CSF leakage rates are higher after the EEA than after the transoral approach. This systematic review, comparing complications associated with transoral and transnasal odontoidectomy, reported intraoperative CSF leakage rates of 0.3% and 30%, respectively, and postoperative CSF leakage rates of 0.8% and 5.2%, respectively. CSF leaks, if not properly treated, may lead to meningitis. Therefore, a skull base neurosurgeon dealing with the EEA should be prepared to deal with complex CSF leaks.

We performed the approach using a Visionsense III 3D endoscope. The relative importance of the 3D view depends on the relative increase in the depth of field it provides in comparison with a 2D endoscope. In 2012, Castelnuovo et al. [28] described one case of transnasal resection of an anterior skull base malignancy with a 3D endoscope. They reported that the surgeons were able to recognize and manage anatomical structures, and control bleeding easily, thanks to use of a bimanual technique and 3D visualization. Probably the most fascinating potential of 3D vision is the ability to control the anatomical structures that are present but not usually and easily visible. The addition of depth perception allows us to overcome the limitations of 2D, making the new 3D endoscopic system an ideal tool for a wide range of procedures. Finally, the accuracy of the visualization of the anatomical structures and their relationships makes the 3D endoscope a versatile tool, which entails a shorter learning curve than the traditional 2D endoscope [12].

Conclusion

The endoscopic endonasal route to the odontoid process has proven to be a feasible, safe and well-tolerated procedure. Anatomical study is mandatory for better understanding of the three-dimensional anatomy of the craniovertebral junction (CVJ) and relation of critical neurovascular structures to specific bony and muscular landmarks. Endoscopic relation of CVJ anatomy is difficult and complex, and cadaver studies help to ensure safety and control during surgical procedures. Anatomical knowledge and dissection in the cadaver laboratory will help future neurosurgeons to develop approaches, facilitate safe surgery and reduce the current limitations of the endoscopic endonasal approach to the CVJ: the caudal exposure limited by nasal and palatine bony and soft tissues. The advantages of the endoscopic endonasal approach are the location of the incision (in the nasopharynx rather than in the oropharynx) and the wider, closer and brighter view provided by the endoscope. High-definition three-dimensional endoscopes, lasers and ultrasound bony curettes have been shown to be useful tools for this approach.

Compliance with Ethical Standards No financial support was received for this work.

Competing Interests The authors declare that they have no competing interests.

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Endonasal and Transoral Approaches to the Craniovertebral Junction: A Quantitative Anatomical Study

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Abstract *Background:* The endoscopic endonasal approach has recently been added to the surgical armamentarium to access the anterior craniovertebral junction (CVJ). Comparative analyses with the transoral approach are scarce. The aim of this study was to provide a quantitative anatomical analysis of both approaches.

Methods: In four specimens the endoscopic endonasal approach (before and after sphenoidectomy) and the transoral approach (without and with a soft palate split) were performed. ApproachViewer—part of GTx-UHN (Guided Therapeutics software, developed at University Health Network, Toronto, ON, Canada)—was used to quantify and visualize the working volume, as well as the exposed area, of each surgical approach. Different modalities (crossing and non-crossing) were used to quantify the exposure of the deep surface, providing an indirect quantitative value of the 'surgical freedom'. The lowest point exposed by the endonasal approaches was compared with that predicted by preoperative radiological lines. Non-parametric Welch analysis of variance (ANOVA) was used for statistical analyses.

Results: The working volume was significantly larger and the distance to the target was shorter with the transoral approaches than with the endonasal approaches. Clival exposure was better with the endonasal approaches than with the non-crossing transoral approach without a soft palate split; areas below C1 were better exposed with the transoral routes. The nasoaxial line best predicted surgical exposure with the endonasal approaches.

Conclusion: Endoscopic endonasal and transoral approaches to the anterior CVJ provide optimal exposure of different areas that overlap at the level of C1 when no anatomical anomalies are present. A split of the soft palate is not necessary during the transoral approach if it is combined with an endoscopic endonasal approach.

Keywords Anatomy · Craniovertebral junction · Endonasal Quantitative · Transoral

Introduction

Endoscopic endonasal approaches to the anterior craniovertebral area have recently been added to the surgical armamentarium for the treatment of pathologies that involve this anatomically complex region [1–3].

Detailed anatomical descriptions of the endoscopic endonasal approach are available [1, 2, 4], but only a few papers have attempted a comparative analysis of this approach and the transoral approach [5–9]. Limited clinical comparative data are available [10].

The aim of this paper is to systematically analyse the anatomical features of the endonasal and transoral routes to the anterior craniovertebral junction (CVJ), using a recently

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developed research method that allows visualization and quantification of the surgical pyramid that defines a surgical approach [11, 12].

Materials and Methods

Specimens

Four lightly embalmed specimens underwent computed tomography (CT) scans, using a 1 × 1 frame with contiguous slices, at both 1 and 3 mm. CT was performed using a gantry of 0°, with a scan window diameter of 225 mm and a pixel size of more than 0.44 × 0.44 (University of Toronto Research Ethics Board (REB) approval no. 23,849). The CT scan files were saved in DICOM (Digital Imaging and Communications in Medicine) format.

Surgical Approaches

Surgery was simulated at the University of Toronto Surgical Skills Centre at Mount Sinai Hospital. Endoscopic dissections were performed using a high-definition head camera with 0° and 30° rod-lens scopes (Karl Storz®, Tuttlingen, Germany). Microsurgical dissections were performed under microscopic visualization at ×4 to ×18 magnifications. A Crockard retractor (Codman®, Randolph, MA, USA) was used for the transoral approaches.

The *endoscopic endonasal approach to the anterior CVJ* was performed. Using a 0° rod-lens scope. The inferior turbinate was out-fractured to visualize the choana bilaterally. A rubber band was introduced into the nostril and pulled out of the oral cavity bilaterally, so as to retract the soft palate. A right middle turbinectomy and maxillary antrostomy were performed, and a nasoseptal flap was harvested on the right and positioned in the maxillary sinus. A posterior septectomy was performed, taking special care to drill the septum flush with the posterior portion of the hard palate. A linear incision was performed at the level of the rhinopharynx, down to the prevertebral fascia, and the muscles were dissected subperiosteally, exposing the anterior tubercle of the atlas. Then a wide sphenoidectomy was added to the approach, drilling the sphenoid floor to the clival plexus posteriorly and laterally to both the vidian and hypoglossal nerves.

The *microsurgical transoral approach* was also performed. The mouth opening was checked and, if it was abnormal because of the partial fixation of the specimens, it was normalized using an incision at the level of the masseter insertion into the zygoma. Once self-retaining retractors were positioned, a rubber catheter was placed through the nose and then sutured to the uvula to retract it out of the field. A midline incision was performed at the level of the oropharynx, elevating a single myomucosal flap from the anterior longitudinal ligament.

The soft palate was then divided at its midline from the junction with the hard palate to the side of the uvula, deviating off the midline to preserve it.

Quantifications

A dedicated software package called ApproachViewer—part of GTx-UHN (Guided Therapeutics software, developed at University Health Network, Toronto, ON, Canada)—was used for the anatomical quantifications [11, 12].

ApproachViewer allows for real-time evaluation and comparison of surgical approaches, as well as postdissection analyses of collected data. It was developed to visualize, in three dimensions (3D), the surgical approach inside the head in which it was performed, as well as quantifying its anatomical features. The quantitative comparison of approaches was therefore based on their anatomical features, defining them as 'truncated pyramids', as described by Andaluz et al. [13]. A truncated pyramid is defined by its superficial surface (or 'surgical window'), deep surface (or 'area of exposure'), height (the distance between the previous areas) and pyramid volume (the surgical space that is available for straight instruments) (Fig. 1).

Commercially available navigation hardware was used (Northern Digital Imaging®, Waterloo, ON, Canada) and included a passive rigid body, a passive probe (pointer) with four markers, and the Polaris Vicra® optical tracking system. The Polaris optical camera emits infrared (IR) light and captures IR reflections off sphere markers attached to the pointer, whose geometry allows reproduction of its position and orientation with a multiplanar reconstruction.

DICOM files were uploaded into GTx-UHN and a registration tolerance of <2 mm was accepted. In addition to providing neuronavigation during the dissections, ApproachViewer allows collection of deep and superficial surfaces, using the pointer to track their perimeters, and provides real-time visualization and quantification of the surgical pyramid in axial, coronal and sagittal sections, and as a 3D rendering (Fig. 4) [11, 12]. The quantification procedure was repeated twice for each approach, tracking both 'non-crossing' and 'crossing' pyramids. A non-crossing pyramid is obtained by keeping the pointer in corresponding positions on both deep and superficial surfaces, thus obtaining the widest obstacle-free working space, defined by a straight instrument. A crossing pyramid is obtained by keeping the pointer in opposite positions on

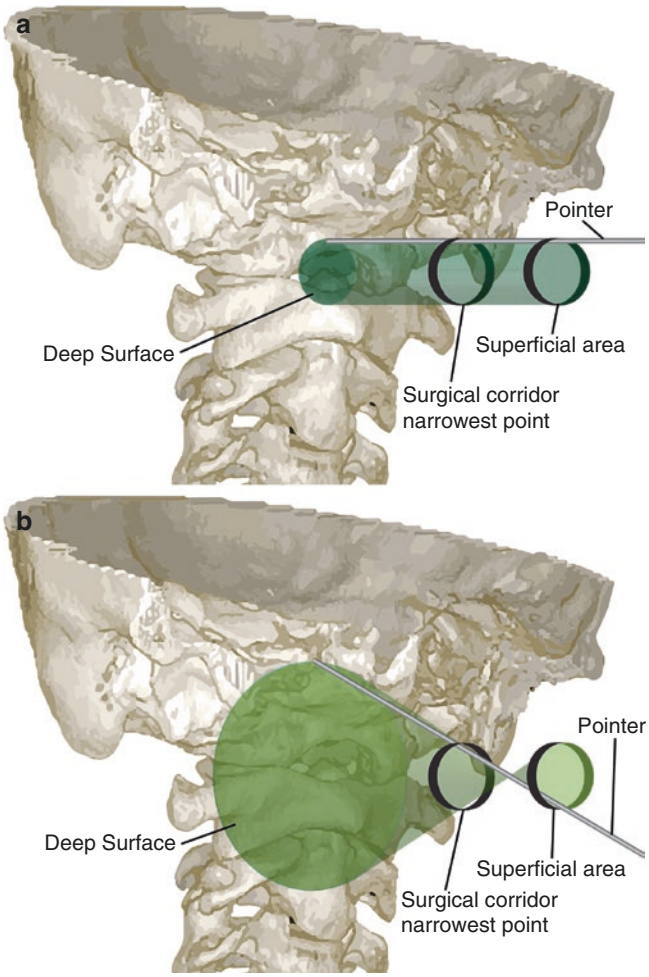

Fig. 1 Surgical pyramid with 'non-crossing' and 'crossing' collection modalities. Two different modalities are used to evaluate the working volume of each approach. (**a**) Non-crossing modality: when the pointer is positioned at the level of the deep area, at the anterior craniovertebral junction (CVJ), special care is taken to touch the superficial area at the same level (i.e. the pointer touches the 12 o'clock point on both surfaces). (**b**) Crossing modality: the pointer positioned on the deepest surface is positioned at the opposite level in the superficial area (i.e. the pointer is positioned at 12 o'clock on the deep surface and touches the 6 o'clock point on the superficial area). The aim of the non-crossing registration is to record the widest volume in which surgical manoeuvrability is maximal, as each point on the deep surface can be reached by all points in the superficial area. The aim of the crossing modality is to record the widest possible deep area; its periphery is made up of points that can be reached only through contralateral points on the superficial area (i.e. for bimanual surgery, the surgeon has to have an angled instrument in one hand to control that point with two instruments)

deep and superficial surfaces (i.e. at 3 o'clock at the pyriform aperture and at 9 o'clock at the posterior wall of the sphenoid sinus), defining the largest deep surface that can be reached with a straight instrument (Fig. 1).

ApproachViewer also allows a postdissection analysis to be performed by drawing areas of interest on the specimen CT scans [11, 12]. The drawings were done by tracing lines extending from one side to the other side of the desired surface in each consecutive axial CT slice, which were automatically assembled to generate the surface. The following five areas were traced for this study: the lower clivus, the C0–C1 space, the anterior arch of the C1 vertebra, the odontoid process and the body of the C2 vertebra (Fig 2). The lower clivus area was defined by the floor of the sphenoid sinus as the superior border, by the lower end of the clivus as the inferior border and by the vertical projections of both hypoglossal canals as the lateral borders. The C0–C1 space area was the surface between the lower clivus and the anterior arch of C1, bound laterally by the occipital condyles. The anterior arch of the C1 vertebra was drawn from the upper border of the arch down to its lower edge, the lateral border being the junction between the arch and transverse processes. The odontoid process area corresponded to the space located between the anterior arch of C1 superiorly, the base of the odontoid process inferiorly and the medial border of C1–C2 articulation laterally. The body of the C2 vertebra area was defined as the anterior surface of the body of C2; its superior border corresponded to the base of the odontoid process, its inferior border was the inferior edge of the vertebra itself, and the lateral borders consisted of the lateral edges of the vertebra inferiorly and the junctions between the body and transverse processes superiorly (Fig. 2). Matching the areas of interest and the surgical pyramids, ApproachViewer provided absolute and percentage values of the surface exposed by each approach.

The height of each approach was calculated by measuring the distance between the midpoint of the superficial surface of the pyramid and the closest point on the odontoid process.

Radiological and Surgical Lines for the Endonasal Approach to the Craniovertebral Junction

The nasopalatine line (NPL) [14] and the nasoaxial line (NAxL) [15] were drawn on CT scans (Fig. 3). The distance between the projections of the NAxL and NPL on the odontoid process was divided into five segments (Fig. 3). Subsequently, the segment reached by the most caudal point of the endonasal approaches was identified and recorded (Fig. 3).

Statistical Analyses

Statistical analyses comparing the percentages of exposure provided by the endonasal and transoral approaches were

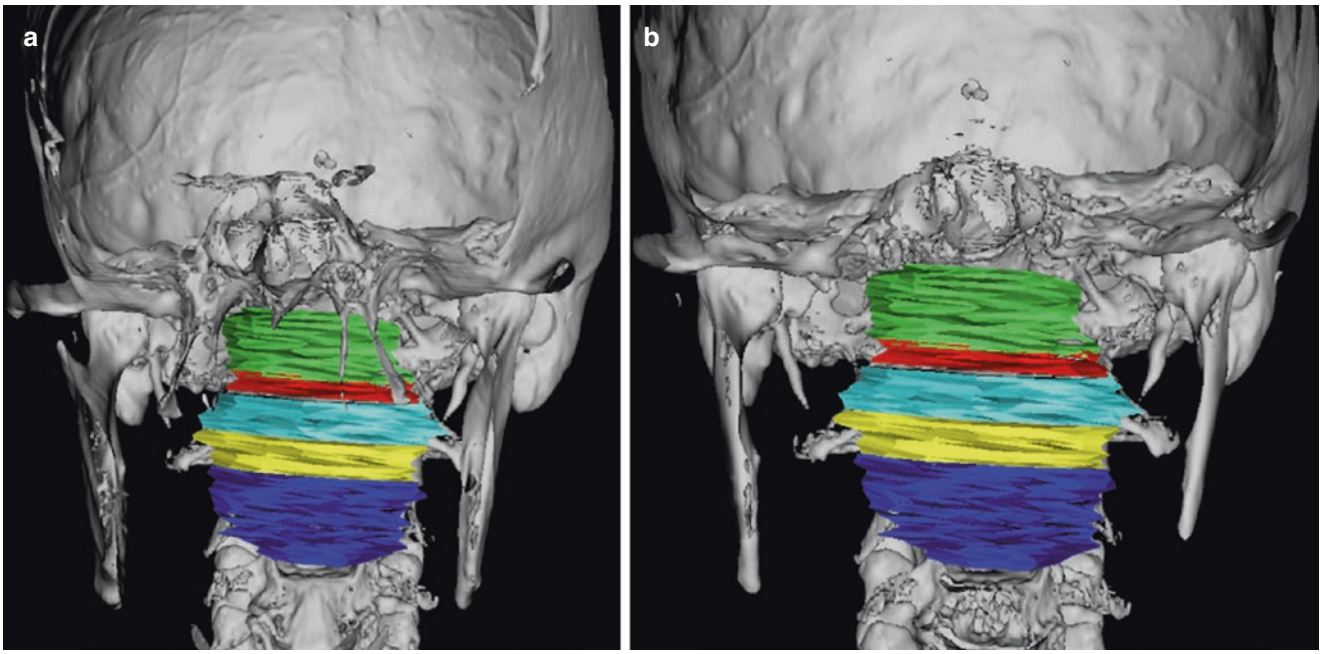

Fig. 2 Anterior craniovertebral junction (CVJ) segmentation. In ApproachViewer, five different areas are drawn: the lower clivus (*green*), the C0–C1 space (*red*), the anterior arch of C1 (*light blue*), the 'odontoid process' (*yellow*) and the body of C2 (*dark blue*) (see text for further details). The same areas are shown with part of the sphenoid and pterygoid processes still visible (**a**) or removed (**b**)

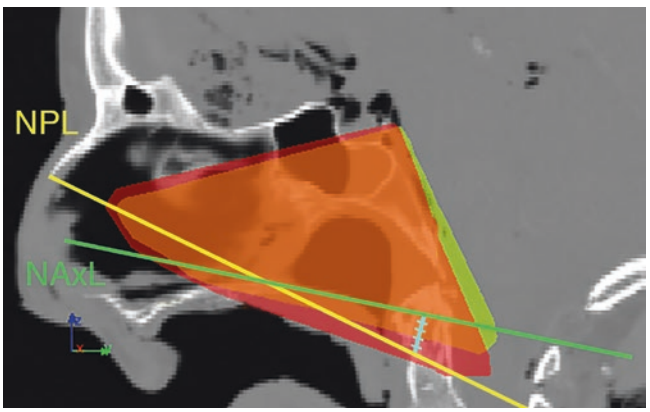

Fig. 3 Predictive lines for endonasal approaches. The nasopalatine line (NPL; *yellow*) and the nasoaxial line (NAxL; *green*) are drawn on computed tomography (CT) images on the midsagittal plane. The NPL is defined by the line that connects the inferior margin of the nasal bones anteriorly and the posterior border of the hard palate posteriorly [14]. The NaXL is the line that starts from the midpoint of the distance from the rhinion to the anterior nasal spine of the maxillary bone and ends at the C2 vertebra, tangential to the posterior nasal spine of the palatine bone [15]. The distance between the two lines at the level of the anterior craniovertebral junction (CVJ) is divided into five segments (*blue line*). The lowest point at the level of the anterior CVJ included in the non-crossing (*yellow*) and crossing (*red*) endonasal approaches is recorded (see text for further details)

performed for each area of interest. The same analyses were performed also for the heights and volumes of the approaches, but the crossing volumes were excluded, as they do not define a real working space.

The predictivity of the NAxL and NPL was analysed by recording where the lowest point of the surgical pyramids was for all endonasal approaches; crossing and non-crossing approaches were compared.

The normality and homogeneity of the variances were tested and demonstrated to not be present for each group of results. Therefore, non-parametric Welch analyses of variance (ANOVAs) were performed.

Results

The mean operative volume of the endonasal approaches was 27.4 cm^3 before the sphenoidectomy, which led to a statistically significant increase to 53.3 cm^3 ($p = 0.00005$) (Table 1). This gain of volume was directed superiorly, as shown in Fig. 3, and not relevant to surgical exposure of the odontoid process.

The mean operative volumes of the transoral approaches, without and with a soft palate split, were 82.2 and 111.5 cm^3, respectively (with a gain of 26.37%); the difference between the two was not statistically significant (Table 1).

The approach height was significantly greater with the endonasal approaches than with the transoral approaches, with mean distances of 90.2 and 80.4 mm, respectively ($p = 0.008$) (Table 1).

The analyses of the anterior craniovertebral areas exposed by each approach are summarized in Table 2. The lower

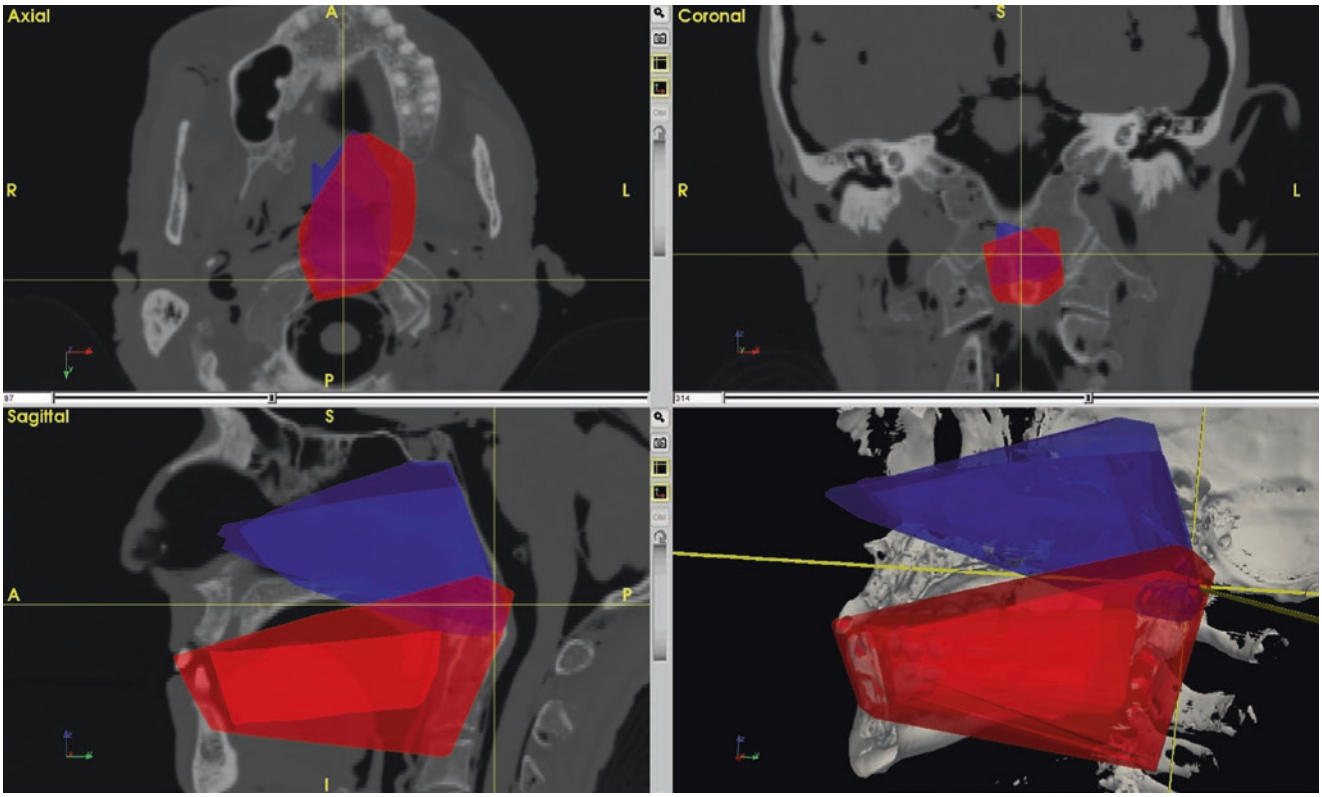

Fig. 4 Areas shared by the endonasal and transoral approaches. The endonasal and transoral approaches share a common area at the level of C1, as shown on the three axes and with the reconstructed surgical volumes in ApproachViewer. An endonasal approach after sphenoidectomy is shown in *blue* (*lighter blue and larger volume:* crossing; *darker blue and smaller volume:* non-crossing). A transoral approach without a soft palate split is shown in *red* (*lighter red and larger volume:* crossing; *darker red and smaller volume:* non-crossing)

Table 1 Heights and volumes of transnasal and transoral approaches (see text for further details)

	Transnasal approach		Transoral approach	
Height [mm (range)]	90.22 (79.4–103.57)		80.35 (63.73–96.91)	
Volume [cm^3 (range)]	Without sphenoidectomy 27.42 (14.96–37.43)	After sphenoidectomy 53.30 (34.36–69.92)	Without soft palate split 82.16 (42.7–133.85)	After soft palate split 111.47 (57.68–147.5)

clivus exposures obtained with the crossing and non-crossing endonasal approaches (47.67% and 38.02%, respectively) were significantly larger than those obtained with the non-crossing transoral approach without a soft palate split (5.93%; $p < 0.00001$).

The only statistically significant difference in C0–C1 surface exposure was between the crossing and non-crossing endonasal approaches (76.71% versus 55.91%; $p = 0.01$).

A similar result was demonstrated for the C1 surface (62.38% versus 22.90%). The non-crossing endonasal approaches showed also significantly less exposure of the C1 surface than the crossing transoral approaches without a soft palate split (80.33%) and with a soft palate split (87.32%). The crossing transoral approaches with a soft palate split provided more exposure than the crossing endonasal approaches ($p < 0.00001$).

The odontoid process surface (i.e. between the inferior margin of C1 and the body of C2; see Fig. 1) was significantly less exposed by the non-crossing and crossing endonasal approaches (0% and 10.54%, respectively) than by the non-crossing and crossing transoral approaches without a soft palate split (65.42% and 83.67%, respectively) and with a soft palate split (78.61% and 90.48%, respectively; $p < 0.00001$ for the comparison between the endonasal and transoral approaches). Similarly, the C2 surface was more exposed by the transoral approaches than by the endonasal approaches ($p < 0.00001$) (Table 1).

The Welch ANOVA performed on the segments reached by crossing versus non-crossing approaches showed that the mean segment reached by non-crossing approaches was the first (mean value 1.19) and the segment reached by crossing approaches was the third (mean value 2.89; $p = 0.01$).

Table 2 Percentages of the different areas of interest exposed by each approach (see text for further details)

	Value [% (range)]					
	TN	TN X	TO	TO X	TO-S	TO-S X
Lower clivus	38.02 (3.63–74.66)	47.67 (20.48–81.45)	5.93 (0–20.58)	35.53 (2.31–69.27)	22.23 (0–49.85)	55.77 (0–94.86)
C0–C1	55.91 (34.57–69.41)	76.71 (51.26–99.32)	29.52 (0–72.37)	69.86 (23.85–96.84)	57.32 (0–85.61)	72.44 (25.95–99.32)
C1	22.90 (0–51.61)	62.38 (43.69–83.01)	53.35 (0–80.88)	80.33 (51.6–96.71)	63.01 (17.42–79.62)	87.32 (77.14–94.3)
Odontoid	0 (0–0)	10.54 (0–45.61)	65.42 (0.91–88.32)	83.67 (64.68–100)	78.61 (70.61–94.86)	90.48 (76.22–100)
C2	0 (0–0)	0 (0–0)	35.09 (0.47–78.4)	71.71 (16.64–99.54)	51.79 (20.34–80.37)	86.26 (51.95–99.74)

TN transnasal, *TN X* transnasal, crossing, *TO* transoral, *TO X* transoral, crossing, *TO-S* transoral after soft palate split, *TO-S X* transoral after soft palate split, crossing

Discussion

As the endonasal endoscopic approach to the CVJ has recently been added to the surgical armamentarium and the pathologies for which it is indicated are rare, clinical data are relatively scarce, as documented by a recent systematic review and meta-analysis that attempted a comparative analysis between transoral and endonasal approaches and was able to include only 92 patients who had undergone transnasal odontoidectomy [10].

Some authors have performed an anatomical comparative analysis of transoral and endonasal approaches to the anterior CVJ.

In this study we applied a new research method, based on the software ApproachViewer (part of GTx-UHN), which allows visualization and quantification of the surgical pyramid that defines an approach, as well as the absolute and percentage values of exposed areas of interest. The volumetric analysis confirmed what seems obvious to the experienced surgeon and has been qualitatively or indirectly demonstrated by others: the endonasal approach provides a significantly smaller working volume than the transoral approach. The height of the surgical pyramid proved to be significantly greater with the endonasal approach. These data are novel and do not confirm previous radiological analyses [16]. Together, these data describe what is qualitatively evident in the volume visualization: the endonasal approach is a significantly narrower corridor to the odontoid; it can be significantly augmented by sphenoidectomy, which provides a further cranial working volume (Fig. 4).

The exposed area analyses well documented that the endonasal approach provides less working space than the transoral approach: the crossing modality, which signifies the extreme surface touched by a straight instrument, almost always significantly led to an increase in exposure when compared with the non-crossing modality in the endonasal approach. These data, though, were documented for each nasal cavity: in a standard odontoidectomy, a bi-nostril technique is used; furthermore, angled instruments might be used to augment the working space.

The comparative area analysis proved that there is a significant overlap between the endonasal and transoral approaches, as reported by others [8, 16–18]. In this study, the area was at the level of the anterior arch of C1. In a previous study of fresh cadavers with the aid of x-rays and CT scans, Visocchi et al. described the different angles that defined both approaches, and they defined the inferior third of the clivus as the shared surgical domain area [8]. The slight difference in these studies' data was most probably due to the difference in the specimens that were used.

As for the preoperative lines that predict the most inferior point exposed by the endonasal approach, we defined both the nasopalatine line (NPL, or 'Kassam' line) [14] and the nasoaxial line (NAxL) [15]. Our findings confirmed that the nasoaxial line best predicts the lowest point of the 'straight' working volume, i.e. the one with the greatest possible surgical manoeuvrability. The NPL was instead closer to the lowest possible point, exposed only by the crossing modality, i.e. the lowest point reached with an endonasal approach with a straight instrument. The inferior margin of the endonasal approaches, as measured by us and defined by the preoperative lines, does not take into account the possibility of using curved drills and instrumentation, which have already been developed for use in endonasal odontoidectomy; furthermore, we did not drill the posterior portion of the hard palate, which is another well-known procedure that can reduce the exposure obtained with an endonasal approach.

Though all of these data might seem obvious to the surgeon who is experienced in both approaches, they validate some clinical impressions and have clinical implications: (1) the transoral approach might be easier to perform because of the single and wider working space, but it will not effectively expose the clivus in most cases without a

soft palate split; (2) the endonasal approach requires specific training and angled instruments to fully take advantage of a narrower surgical corridor; and (3) a combination of these approaches eliminates the need for a soft palate split in the case of a transoral approach.

Limits of the Present Study

The use of lightly embalmed rather than fresh specimens probably led to underestimation of both endonasal and transoral approaches due to relatively greater stiffness of the tissues.

We did not compare use of a microscope and an endoscope in the transoral approach. The aim of the study was to document the working volume of each approach, which is not influenced by the visualizing tool: as shown by the group led by Ammirati in an anatomical study [19] and by others in clinical reports [17, 20, 21], use of an endoscope can certainly increase visualization, but specific angled instrumentation might be needed to fully take advantage of endoscopic visualization and to optimize surgical manoeuvrability.

Conclusion

The endoscopic endonasal and transoral approaches to the anterior craniovertebral junction provide optimal exposure of different areas that overlap at the level of C1 when no anatomical anomalies are present. A split of the soft palate is not necessary during a transoral approach if it is combined with an endoscopic endonasal approach. Detailed knowledge of the non-crossing volume that defines an approach might contribute to the development of optimal angled instrumentation.

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Competing Interests The authors declare that they have no competing interests.

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Ventral Brainstem Anatomy: An Endoscopic Transoral Perspective

Oreste de Divitiis, Alfredo Conti, Teresa Somma, Flavio Angileri, and Paolo Cappabianca

Abstract In recent years the use of the endoscope through the transclival route has gained new attention as a minimally invasive operative method to successfully treat numerous clival pathologies such as chordomas, meningiomas, haemangiopericytomas, enterogenous and epidermoid cysts, and metastasis(Cappabianca et al. Neurosurgery 55:933–940, 2004; Cappabianca et al. Childs Nerv Syst 20:796–801, 2004; Cappabianca et al. Adv Tech Stand Neurosurg 33:151–199, 2008; Cappabianca et al. Neurosurgery 49:473–475, 2001; Cappabianca et al. Surg Neurol 62:227–233, 2004; Dehdashti et al. Neurosurgery 63:299–307, 2008; Kerschbaumer et al. Spine (Phila Pa 1976) 25:2708–2715, 2000; Saito et al. Acta Neurochir (Wien) 154:879–886, 2012; Stippler et al. Neurosurgery 64:268–277, 2009). Here we describe the endoscopic anatomy of the region reached through an endoscopic transoral approach. Fresh and formalin-fixed cadaver specimens were used to demonstrate both the feasibility of an endoscopic transoral–transclival intradural approach and its potential exposure. The transoral approach was performed using a clival opening of 20 × 15 mm. This smaller access point through the clivus, which allowed insertion of the endoscope and its instruments, did not limit the complete exposure of the cisternal spaces and permitted reconstruction of all anatomical layers.

This endoscopic approach thus provides excellent exposure of some of the most dangerous and inaccessible territories of the brain, respecting the anatomy and remaining a minimally invasive approach. Further extensive clinical experience is necessary to prove its safety. The endoscopic transoral–transclival approach will presumably be selected to gain access to lesions of the lower ventral brainstem and the surrounding cisternal spaces, with development of new and more efficient surgical strategies for dural and bone defect repair.

Keywords Transoral · Endoscopy · Brainstem · Vertebrobasilar system · Anatomy · Brain tumour

Introduction

A wide spectrum of neoplastic and malformative lesions can affect the ventral portion of the brainstem and the vertebro-occipital junction. The approach for surgical resection of tumours and vascular lesions of the petroclival junction and anterior brainstem still remains one of the major challenges of contemporary neurosurgery, although many different possible routes have been proposed to reach this critical area. With a conventional skull base approach it is possible to achieve a good view of the anterior brainstem through a posterolateral route, but it is aggressive and requires wide bone removal and a high degree of cerebral and vascular manipulation, in contrast to the modern concept of minimally invasive surgery [10, 16–18, 28, 32–34, 37, 38, 41]. The less deconstructive and direct route to reach the anterior surface of the brainstem is through the pharynx and the clival bone, which offers a direct view of the anterior brainstem and vertebrobasilar junction without requiring dislocation or manipulation of any noble structure.

The transoral approach has been used extensively for surgical removal of extradural lesions and is considered effective to reach ventral tumours and also non-neoplastic lesions of the clivus and the craniovertebral junction (CVJ) [1, 9, 10, 12, 19, 20, 22–25, 35].

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This approach for treatment of an extra-axial tumour was described for the first time by Mullan et al. [26] in 1966; since then the approach has been used to treat midbasilar or vertebrobasilar junction aneurysms [13, 14, 18, 30]. In 1991, Crockard and Sen [10] reported surgical resection of seven intradural lesions, comprising meningiomas and neurofibromas, using this access route.

Despite their innovative efforts, these authors reported various postoperative complications such as meningitis and cerebrospinal fluid (CSF) fistula. Obtaining a watertight closure of the clival dura mater is actually extremely difficult. Therefore, all patients experienced CSF leaks, which required positioning of lumbar drainage or a multilayer reconstruction. Because of this set of problems, use of the transoral approach for intradural pathology was mostly abandoned. In recent years, use of an endoscope through the transclival route has gained new attention as a minimally invasive operative method to successfully treat numerous clival pathologies such as chordomas, meningiomas, haemangiopericytomas, epidermoid cysts and metastasis [2–6, 11, 21, 31, 36]. Here we describe the endoscopic anatomy of the region reached through a transoral approach.

Endoscopic Anatomy of the Anterior Brainstem and Surrounding Structures

Methods

This anatomical study was performed at the Institute of Anatomy of the University of Vienna in Austria. For the study we used ten heads from fresh cadavers and five from formalin-fixed cadavers. The arteries and veins were injected under pressure with coloured silicone rubber (Dow Corning, Midland, MI, USA) via the internal carotid arteries and internal jugular veins. We used rigid endoscopes with 0°, 30°, 45° and 70° rod lenses, 2.7 mm or 4 mm in diameter and 11 cm or 18 cm in length (Karl Storz, Tuttlingen, Germany).

A retractor system was used to keep the mouth open. The soft palate was split on the midline. The pharyngeal mucosa was incised and the mucoperiosteal layer was retracted, exposing the clivus. On the clival surface the pharyngeal tubercle was identified, and a clival craniectomy with an average diameter of 20 mm in length and 15 mm in width was performed just above it, with a high-speed drill. The dura mater was visualized and opened with a linear incision. A video-capture system (Sony digital still recorder) was used for digital acquisition of endoscopic images.

Results

Through the opened dura mater we directly visualized the premedullary, the prepontine, and the lateral cerebellomedullary cisterns (Fig. 1a, b). Following the cisternal course of the vertebral arteries we reached the vertebrobasilar junction at the pontomedullary junction. In a lateral view, coming from the vertebral arteries, the origin of the two posteroinferior cerebellar arteries and the anterior spinal artery were visible from the cerebellomedullary cistern and the premedullary cistern, respectively. At this level we also observed the fibres of the hypoglossal nerve arising from the preolivary sulcus.

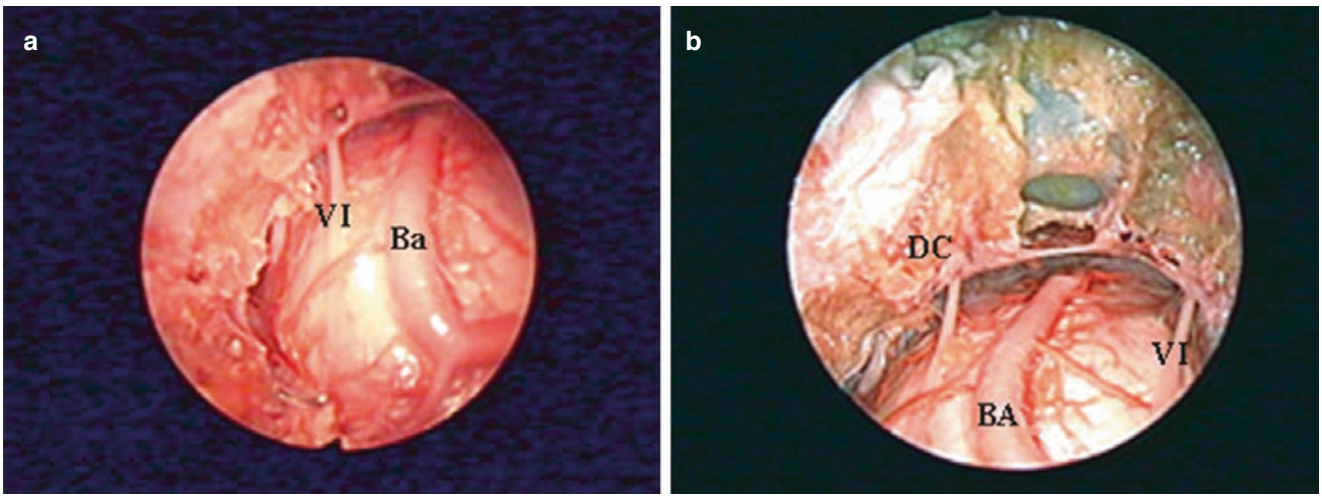

Fig. 1 With a 0° optical lens, the premedullary cistern (**a**) and prepontine cistern (**b**) were visualized. After the opening of the dura mater in the centre of the endoscopic field we could visualize the basilar artery (BA) and the vertebrobasilar junction. Lying over the pyramid decussation the two vertebral arteries (VA) were visible and could be followed until the spinomedullary junction. Along the course of the basilar sulcus to the upper part of the basilar artery, at the border of the interpeduncular cistern, the origin of the anteroinferior cerebellar artery and the perforating branches could be visualized. The entire free course of both abducens nerves (VI) could be followed in the prepontine cistern, from their origin in the pontomedullary sulcus to Dorello's canal (DC)

The basilar artery—with its typical variability in diameter, length and course—could be visualized in the lower two thirds of the field in the prepontine cistern [29]. In this cistern the abducens nerve was recognized and followed along its course toward Dorello's canal. It was possible to visualize the cerebellopontine cistern from a premeatal route by using a 30–45° optical lens and lateral inclination of the endoscope. The acoustic–facial bundle was identified coming after the anteroinferior cerebellar artery, and we were able to follow it along its free cisternal course to the internal acoustic channel (Fig. 2a).

The anatomical characteristics of the internal acoustic channel that can be visualized are very dependent on the optical properties of the endoscope. In fact, it was also possible to identify the labyrinthine arteries in their course toward this channel (Fig. 2a). Using the same angled optical lens at 30° and 45° rotated rostrally, we were able to explore the upper part of the cerebellopontine angle. The most important structure visualized under this view was represented by the trigeminal nerve along its course from the pons toward Meckel's cave (Fig. 2b). With the same angled optical lens and with the endoscope turned laterally and downward, it was possible to reach the posterior part of the lateral cerebellomedullary cistern (Fig. 2c, d).

The interpeduncular fossa was also reached with this approach, using an angled optical lens at 45° or 70°. With the endoscope directed upward with an inclination of approximately 45° it was possible to follow the basilar artery to its

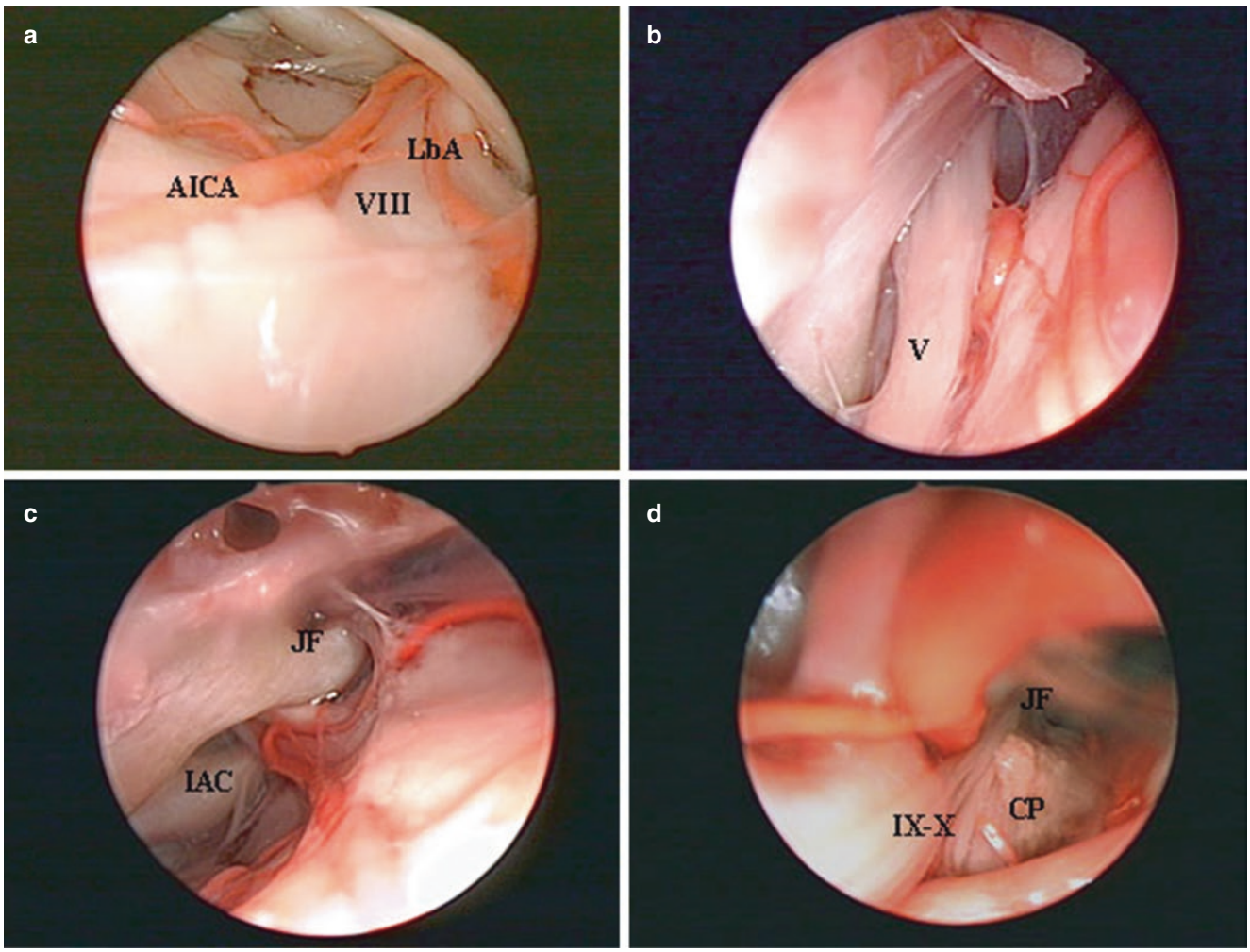

Fig. 2 With lateral inclination of the endoscope and a 30–45° optical lens it was possible to enter the cerebellopontine cistern via a premeatal route along the anteroinferior cerebellar artery (AICA) course. The acoustic–facial bundle (VII–VIII) was identified coming after the anteroinferior cerebellar artery, and we were able to follow it along its free cisternal course to the internal acoustic channel (IAC). At this level it was possible to visualize the labyrinthine arteries (LbA) in their course toward the internal acoustic channel (**a** and **c**). Using an angled optical lens at 30° and 45° rotated rostrally, we were able to explore the upper part of the cerebellopontine angle. The most important structure visualized under this view was represented by the trigeminal nerve (V) along its course from the pons toward Meckel's cave (**b**). By using the same angled optical lens and turning the endoscope laterally and downward, we could reach the posterior part of the lateral cerebellomedullary cistern. It was possible to visualize the IX and X nerves (IX–X) running laterally and posteriorly from the retro-olivary sulcus to the jugular foramen, covered in their anterior portion by a tuft of the choroidal plexus (CP) exiting from the foramen of Luschka and by the variable looping of the posteroinferior cerebellar artery (**d**)

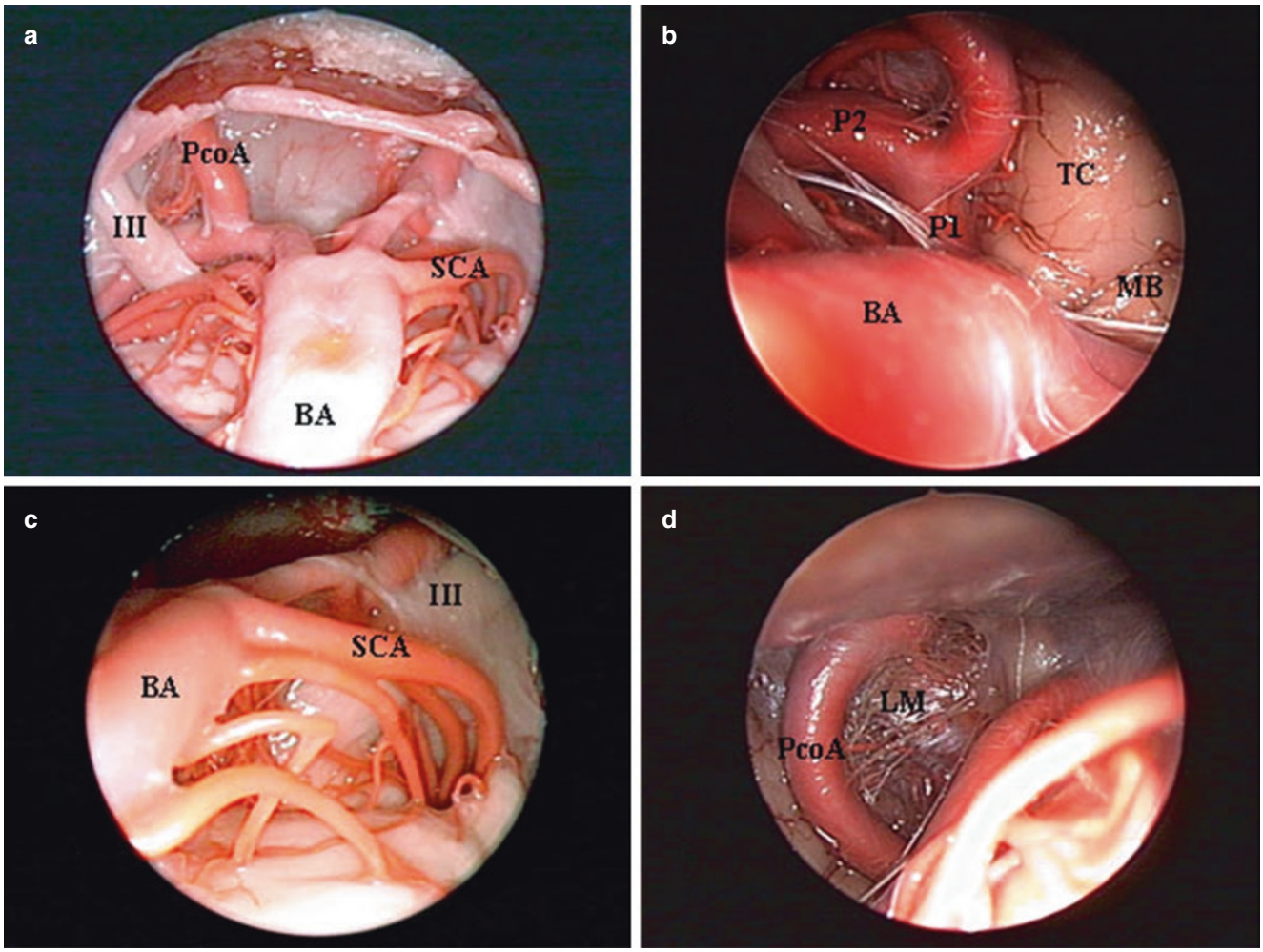

Fig. 3 We reached the interpeduncular cistern by using a 70° optical lens and upward inclination of the instrument, following the basilar artery (BA) to its superior third. In this cisternal space the visual field was limited superiorly by the tuber cinereum (TC), anterolaterally by Liliequist's membrane (LM) and posterolaterally by the optic tracts. The basilar tip, the basilar bifurcation, the superior cerebellar arteries (SCA) and the P1 tracts of the posterior cerebral arteries were completely visible. The perforating branches of the basilar tip and of P1 were visible in detail and could be followed while entering the posterior perforated substance. The posterior communicating arteries (PcoA) appeared in the anterolateral part of the surgical field, where they crossed Liliequist's membrane with a lateral deflection to reach the posterior communicating arteries. Above the posterior perforated substance and posterior to the tuber cinereum, mammillary bodies (MB) were also visualized. Identification of the oculomotor nerves (III) completed the exploration of the cistern. They coursed from the interpeduncular fossa, passing between the superior cerebellar arteries and the posterior communicating arteries in an anterior and superior direction toward the tentorial edge

superior third, which was hidden by the border of the clival craniectomy. It was thus possible to reach and widely explore the interpeduncular cistern (Fig. 3a). The oculomotor nerves could be clearly identified (Fig. 3a–c). In the anterolateral part of the surgical field the posterior communicating arteries appeared, and they crossed Liliequist's membrane to reach the posterior cerebral arteries with a lateral deflection (Fig. 3d). The mammillary bodies and the tuber cinereum were also visualized (Fig. 3c). The perforating branches of the basilar tip and of P1 were visible in detail and could be followed while entering the posterior perforated substance (Fig. 3d).

Discussion

An endoscopic minimally invasive approach for the treatment of intradural lesions located in or around the anterior brainstem offers several theoretical advantages. In our anatomical study we confirmed that this approach allows the operator to obtain full access to the anterolateral brainstem and to the cisternal space that surrounds it, from the spinomedullary junction to the interpeduncular cistern, offering a thorough vision of the vertebrobasilar arterial system and of cranial nerves III–XII. This endoscopic approach thus

provides excellent exposure of some of the most dangerous and inaccessible territories of the brain, remaining a minimally invasive approach.

Furthermore, the majority of the technical problems related to the microscope-assisted transoral approach could be solved by use of endoscopic instrumentation. In fact, in the microsurgical transoral procedure proposed by Crockard and Sen [10] the clival opening needed for identification of intradural lesions was 2–3 cm; in our study the opening sufficient for the endoscopic view was limited to 20 mm in length and 15 mm in width, and it was located in a 'safe' entry zone through the clivus. This smaller access point through the clivus did not limit the complete exposure of the cisternal spaces and could reduce the rate of postoperative instability by minimizing the risk of condyle injuries. Moreover, the use of the endoscope for this approach offers exposure superior to that of the traditional open transoral route, as was confirmed in a recent anatomical study performed by La Corte et al. [23]. Furthermore, the endoscopic transoral approach permits better preservation of velopharyngeal function; a wider clival defect created with the microscopic approach is responsible for incompetence between the posterior pharyngeal wall and the soft palate, resulting in postoperative deficits in swallowing and in phonation. In fact, as reported by Chan et al. [8], the endoscopic endonasal approach could be reliably used to gain access to the CVJ, avoiding the dissection of soft palate tissue associated with a palate-splitting technique or with an extended endonasal approach. Another possible complication related to the microscope-assisted transoral approach, recently confirmed by Visocchi et al. [39], is the need for postoperative tracheostomy placement due to postoperative respiratory dysfunction.

The dural opening was minimized with the endoscope-assisted transoral technique because it was sized to allow introduction of only the endoscope and the instruments, and it is possible to close the dura mater by use of multilayer reconstruction. In comparison with the transsphenoidal approach, the wider entrance area and the shorter distance make the reconstruction of the bone plane easier and allow repositioning of the mucoperiosteal flap.

Both of these aspects—the limited size of the entry point and the possibility of easier reconstruction—may be the basis for the reduced occurrence of postoperative cerebrospinal leakage and meningitis, which represent the main postoperative complications of the standard approach.

Nowadays the endoscopic transoral approach is not the standard technique. However, clinical findings on the use of an endoscopic transoral–transpharyngeal approach to treat craniocervical junction abnormalities have been reported in the neurosurgical literature, and endoscopes have also been used to assist in the removal of brainstem cavernous angiomas and clival tumours [7, 9, 15, 27, 40, 42]. The endoscopic transoral–transclival approach will presumably be selected to gain access to lesions of the lower ventral brainstem and the surrounding cisternal spaces, with development of new and more efficient surgical strategies for dural and bone defect repair. For appropriately selected lesions near the palatal line, the endoscopic transoral approach appears to be the preferred approach [8].

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Transoral Approach to the Craniovertebral Junction: A Neuronavigated Cadaver Study

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Introduction

More than 100 years after the first description by Kanavel of a transoral–transpharyngeal approach to remove a bullet impacted between the atlas and the clivus [1], the transoral approach (TOA) still represents the 'gold standard' for surgical treatment of a variety of conditions resulting in anterior craniocervical compression and myelopathy [2, 3]. Nevertheless, some concerns—such as the need for a temporary tracheostomy and a postoperative nasogastric tube, and the increased risk of infection resulting from possible bacterial contamination and nasopharyngeal incompetence [4–6]—led to the introduction of the endoscopic endonasal approach (EEA) by Kassam et al. [7] in 2005. Although this approach, which was conceived to overcome those surgical complications, soon gained wide attention, its clear predominance over the TOA in the treatment of craniovertebral junction (CVJ) pathologies is still a matter of debate [3]. In recent years, several papers have reported anatomical studies and surgical experience with the EEA, targeting different areas of the midline skull base, from the olfactory groove to the CVJ [8–19]. Starting from these preliminary experiences, further anatomical studies have defined the theoretical (radiological) and practical (surgical) craniocaudal limits of the endonasal route [20–25]. Our group has done the same for the TOA [26, 27] and compared the reliability of the radiological and surgical lines of the two different approaches. Very recently, a cadaver study, with the aid of neuronavigation, tried to define the upper and lower limits of the endoscopic TOA [28].

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The purpose of the present study, whose preliminary data were published in 2015 [27], is to exploit the accuracy provided by neuronavigation in order to further compare operative craniocaudal extensions of the transnasal and transoral routes.

Materials and Methods

Materials

Two adult formalin-fixed cadavers were examined after computed tomography (CT) scanning (multidetector, 128 layers) and with the aid of neuronavigation (Medtronic StealthStation Treon Plus) and use of the following instruments: a high-speed drill (Storz, Tuttlingen Germany); vacuum aspirator (Super Vega Battery); digital camera (EOS 7D telescopic lens image stabilizer ultrasonic macro 100 mm; Canon, Tokyo, Japan); microsurgical instruments; stainless steel headholder; and jaw block.

Methods

All four phases of specimen preparation (thawing, irrigation, fixation and perfusion) were performed at our centre, following a research protocol developed by our group. Before perfusion, the formalin-fixed specimens underwent high-definition CT scanning with the iodinated monomeric contrast medium Iomeprol (Iomeron®) 375 mg/mL. The imaging data (saved in DICOM [Digital Imaging and Communications in Medicine] format) were stored on compact disc (CD) and imported into the neuronavigation workstation (Medtronic Treon), and three-dimensional reconstructions were obtained. A jaw block was used to achieve maximal opening of the oral aperture.

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Results

The examination of the CT scan of the two specimens did not reveal any CVJ pathology. With the aid of neuronavigation, accurate measurements were made in both cadaver heads. The results for both specimens in terms of craniocaudal and lateral exposures are summarized in Tables 1 and 2 and shown in Fig. 1.

Discussion

The TOA, spanning ventrally from the inferior third of the clivus to the C2–C3 interspace, allows shorter, wider and more direct access to the CVJ than other approaches, including the anterior, lateral and posterior approaches [29, 30]. Because of these anatomical and surgical considerations, this approach has been considered the preferred route to treat irreducible extradural ventral lesions causing cervicomedullary compression [4, 31–33]. Extensions of the approach with palatotomy, labiomandibulotomy or osteotomy, which are sometimes required to expose lesions located more rostrally, carry high risks of various types of permanent damage, including velopharyngeal insufficiency, malocclusion, neural deficits, temporomandibular joint (TMJ) dysfunction, swallowing and speech difficulties, and need for a tracheostomy and nasogastric feeding tube [33, 34]. The need to overcome the occurrence of these comorbidities of considerable clinical significance led to the development of alternative and potentially less invasive techniques to address ventral CVJ pathology, such as the EEA. Extensive literature has demonstrated through comparative anatomical and clinical studies that an endoscope—in addition to providing increased rostral exposure, brighter illumination and closer visualization of the lesion to be treated [35, 36]—can be used during the TOA as a valid complementary tool in a combined procedure. Nevertheless, though a recent systematic review and meta-analysis [37] demonstrated a statistically significant increased risk of postoperative tracheostomy after the TOA in comparison with the EEA, it also showed a slight trend toward an increased morbidity/mortality prevalence with the EEA in comparison with the TOA (mortality 4% versus 2.9%; intraoperative cerebrospinal fluid [CSF] leakage 30% versus 0.3%; postoperative CSF leakage 5.2% versus 0.8%; meningitis 4% versus 0–4%; reoperation 5.1% versus 2.5%; velopharyngeal insufficiency 6.4% versus 3.3%; sepsis 7.7% versus 1.9%), although none of these differences were statistically significant. These data have prompted us to reconsider the presumed clear-cut superiority of the EEA over the endoscopic TOA, demonstrating the need for further comparative studies to better define and quantify real advantages and disadvantages of these techniques that are useful for the surgical decision-making process.

Table 1 Craniocaudal exposure: comparison between transoral and transnasal approaches

Craniocaudal exposure	Transoral	Transnasal	Percent superiority of transoral to transnasal (%)
Specimen A	45 mm	30.1 mm	33.12
Specimen B	44.9 mm	20.2 mm	55.02

Table 2 Lateral exposure: comparison between transoral and transnasal approaches

Lateral exposure	Transoral	Transnasal	Percent superiority of transoral to transnasal (%)
Specimen A	50 mm	29.8 mm	40.4
Specimen B	58.6 mm	25.8 mm	55.98

To clearly define the limits of the TOA, our research group devised a radiological 'theoretical' line—the palatine inferior dental arch line (PIA), a conceptual analogue of the nasopalatine line (NPL)—as a reliable predictor of the maximal superior extension of the TOA, and we then compared the reliability of the radiological and surgical lines of the two different approaches.

A recent cadaver study by LaCorte et al. was also performed with the aid of neuronavigation, with the aim of defining the upper and lower limits of the endoscopic TOA [28].

In the wake of our previous experimental volumetric studies [26, 27] and other recent contributions, we tried to exploit the accuracy provided by neuronavigation, to further compare operative sagittal and axial extensions of the transnasal and transoral corridors. Our observations were consistent with a relevant advantage of the TOA over the EEA in terms of craniocaudal and lateral extension in both specimens. It is worth noting that our measurements were performed in the setting of a minimal oral aperture as a consequence of the jaw block. Considering that this setting was suboptimal, we speculate that the actual advantage of the TOA is even greater than that reported in our study. We also conclude that even in cases in which wide opening of the mouth is not achievable, as in the case of paediatric patients, the TOA still offers a significant gain in terms of sagittal and axial exposure.

This study has limitations that are inherent to many cadaver studies: the specimens had normal cranial base anatomy, and the findings in this study may not be applicable in cases where the cranial base or oropharyngeal anatomy is abnormal as a result of disease or congenital variation. Moreover, the CVJ is a 'moving target', with great variability even among individuals without CVJ pathologies, as recently reported by Burke et al. [38]. In their study the CVJ was positioned below the palatine line (PL) in two thirds of

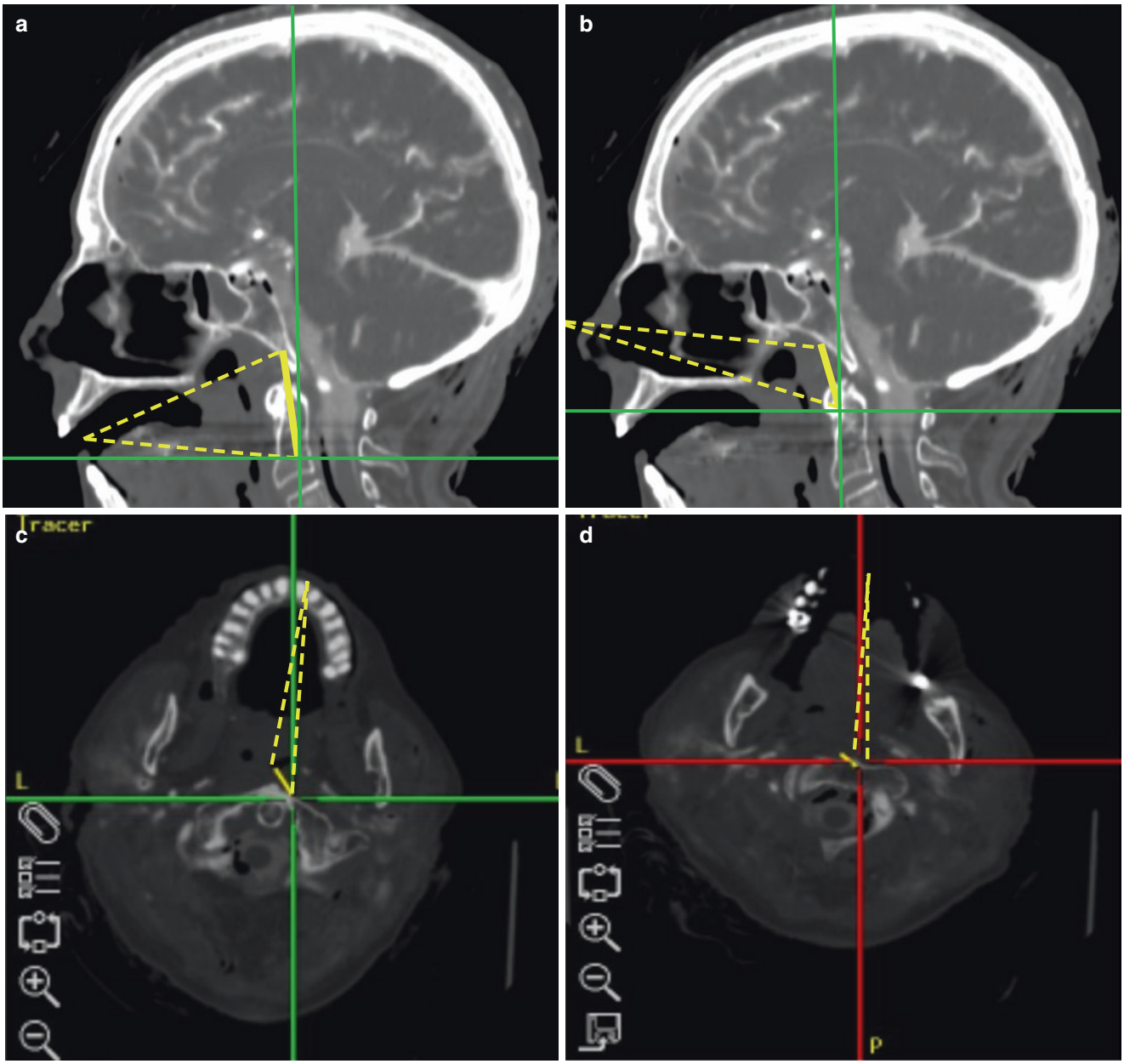

Fig. 1 (**a**, **b**) Sagittal and (**c**, **d**) axial neuronavigated computed tomography (CT) scans with contrast medium showing (**a**) craniocaudal and (**c**) lateral exposures of the transoral approach and (**b**) craniocaudal and (**d**) lateral exposures of the transnasal approach

the control group and above it in one third of the group. Furthermore, because of the small number of specimens, our findings require validation in larger studies.

Conclusion

Our experimental study, conducted with the aid of neuronavigation, confirms that the transoral approach (TOA) offers a wider surgical working channel than the endoscopic endonasal approach (EEA), even in conditions in which the oral aperture is suboptimal. These findings, along with recent observations that the EEA can produce complications similar to those seen with the TOA in craniovertebral junction surgery—including velopharyngeal insufficiency and severe infections—suggest that the presumed superiority of the EEA over the TOA needs to be re-examined.

Compliance with Ethical Standards No financial support was received for this work.

Competing Interests The authors declare that they have no competing interests.

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Pathophysiology

Atlantoaxial Instability: Evolving Understanding

Atul Goel

The atlantoaxial joint is the most mobile joint in the body. The physical architecture of the joint is characterized by a uniformly round and approximately flat surface, which allows a wide range of unobstructed movements. The standing human posture and lifelong heartbeat like uninterrupted activity of the atlantoaxial joint, and its ability to facilitate saying both 'yes' and 'no' necessarily requires smooth and 'fluid' movements that are supported by strong yet supple ligaments. The magnificent architectural structure that is 'magically' designed and carved by nature to provide both stability and mobility and to allow a smooth and safe transit passage for the most critical neural and vascular structures can only be admired in awe and appreciated.

All movements in the atlantoaxial joint occur at the large and strong facet joints. The odontoid process acts to guide and direct the movements. The odontoid process and intervertebral discs are designed to act like opera conductors for musicians, symbolized by the facets. The odontoid process and intervertebral discs are similar in function and act as the 'brain' of the movements, while the 'brawn' of the movements is at the facets [1, 2].

In general, as they say, there is no terror of error in nature. However, considering the remarkable and flawless activity that needs to be performed, any kind of disuse or misuse of muscles, physical injury, and chemical or genetic disorganization leading to abnormal ligamentous laxity can make the joint unstable. Tuberculosis, rheumatoid arthritis and tumours can also destroy the fabric of the bone construction, making the joint unstable. Poor child delivery practices and protein calorie malnutrition can initiate and establish atlantoaxial instability early in life.

Conventionally, atlantoaxial instability is diagnosed by an abnormal increase in the atlantodental interval on flexion of the neck [3, 4]. In general, on plain radiographs, an atlantodental interval of more than 3 mm on flexion of the head is considered to be abnormal. In children, because of the suppleness of the tissues, the atlantodental interval may normally be a little larger than that in adults. For several decades, when plain radiography formed the prime investigative modality, this parameter constituted the sole mode of diagnosing atlantoaxial instability. With the introduction of computer-based imaging, the range of parameters that suggested instability increased, but the abnormal increase in the atlantodental interval and its consequent neural effects formed the basis of diagnosis. Computed tomography (CT) examination showing a reduction in spinal canal dimensions and neural compression provides clear evidence of instability. Magnetic resonance imaging (MRI) has brought clarity to imaging and shows cord compression and pressure effects on the neural structures vividly. Compression of the subarachnoid space anterior to the cord and posterior to the odontoid process is also suggestive of atlantoaxial instability.

As our understanding of the region and radiographic imaging have improved over the years, other forms of atlantoaxial instability have also been included in the range and scope of clinical diagnosis. The previously labelled *'irreducible' atlantoaxial dislocation* is now considered to be a pathologically mobile and unstable clinical entity [5]. The understanding of this has revolutionized surgery for basilar invagination, which has now moved on from decompression via the anterior transoral route or posterior foramen magnum decompression to atlantoaxial stabilization [6–8]. *Vertical atlantoaxial dislocation* is identified by superior migration of the odontoid process in the presence of maintained atlantodental alignment. It results in basilar invagination. Vertical atlantoaxial dislocation can be mobile and reducible on dynamic imaging. Incompetence of the facets and the joint appears to form the basis of vertical atlantoaxial dislocation [9]. *Lateral facetal instability* occurs in situations where the ring of the atlas is broken following trauma or infection, or in association with bone-destructive tumours or bifid anterior

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or posterior arches of the atlas [10–12]. The facet of the atlas is dislocated laterally in relation to the facet of the axis. *Rotatory dislocation* generally occurs in young children. In this condition the facet of the atlas rotates anteriorly in relation to the facet of the axis on one side and the facet of the atlas is posterior to the facet of the axis on the contralateral side [13].

Apart from diagnosis of instability on the basis of an increase in the atlantodental interval, atlantoaxial dislocation or instability can also be diagnosed on the basis of the alignment of the facets of the atlas and axis [14, 15]. On a lateral profile image, sagittal CT scan or MRI, *type A facetal dislocation* is when the facet of the atlas is positioned or dislocated anteriorly in relation to the facet of the axis. The alignment of the facets in this type of dislocation simulates the alignment of vertebral bodies in cases of lumbosacral listhesis [16]. We have labelled this situation as *spondyloptosis*, when the entire facet of the atlas is dislocated and positioned anteriorly in relation to the facet of the axis. The dislocation of facets results in posterior and superior migration of the odontoid process, resulting in basilar invagination and atlantoaxial dislocation. The odontoid process compresses the spinal cord. We recently hypothesized that more than neural deformation or compression, it is the repeated microtrauma related to instability that produces clinical symptoms [17]. As the neural compression occurs relatively early in the course of the disease and the joints are clearly unstable, the symptoms are relatively acute in this type of facetal instability. Type A facetal instability is associated more frequently with mobile atlantoaxial dislocation and group A basilar invagination.

Type B facetal instability is when the facet of the atlas is dislocated posteriorly in relation to the facet of the axis or when there is retrolisthesis of the facets. We have also identified type C facetal instability, when the facets are in alignment on lateral imaging but there are clinical parameters and other radiological evidences—such as the presence of a short neck, torticollis, basilar invagination, assimilation of the atlas, bifid anterior and posterior arches, platybasia, Klippel–Feil abnormality, Chiari malformation type I and syringomyelia—that suggest the presence of instability. The atlantodental interval may not be increased and the odontoid process may not be displaced posteriorly in type B and C facetal instability. The clinical symptoms are long-standing or chronic in this situation. We recently hypothesized that musculoskeletal and neural malformations are secondary effects of atlantoaxial instability and are probably protective in their function [18]. The atlantoaxial instability in type B and C cases can primarily be identified on direct bone manipulations during surgery. The evidence that atlantoaxial instability is the cause of myelopathic symptoms and also is the primary cause of various musculoskeletal and neural responses is supported by the immediate postoperative recovery of symptoms and reversal of both symptoms and musculoskeletal and neural alterations following surgery that involves atlantoaxial fixation alone without any bone or soft tissue removal.

As our understanding of the region has matured, we have identified that instability of the atlantoaxial joint is relatively frequent and significantly underdiagnosed. Considering that the atlantoaxial joint is the most mobile joint in the body, the possibility of developing instability is most profound. The relatively flat joint surfaces that facilitate extensive movements also make this joint prone to instability. It appears that missing or ignoring the presence of atlantoaxial instability can frequently cause persistence of symptoms or failure of treatment. Atlantoaxial instability can be present even when there is no alteration in the positioning of the odontoid process, no alteration in the atlantodental interval and even no malalignment of the facets. Direct handling of the bones of the atlantoaxial vertebrae during surgery can provide significant evidence of instability. However, to diagnose instability on the basis of direct bone handling necessarily requires extensive surgical experience.

We recently presented our study on degenerative spondylosis of the spine and concluded that vertical instability of the spinal segments that is manifested by slipping or telescoping of the facets in the subaxial spine is the primary nodal point of the genesis of spondylotic spinal changes [15, 19–26]. This is at variance with the disc-centric theory suggesting that disc space reduction due to loss of water content is the initial pathological event. The standing human position and misuse or disuse of the muscles supporting the spinal pillar are the primary causes of spinal instability. Because of the oblique profile of the cervical and dorsal facets and the vertical profile of the lumbar facets, it is difficult to identify instability of the facets by using conventional and even modern imaging techniques. However, malalignment of the atlantoaxial facets is relatively easily identified by imaging, because of their flat and rectangular brick-like structure on lateral imaging. Corroborative evidence such as the presence of osteophytes, ligamental buckling and associated clinical symptoms can suggest facetal instability in the subaxial spine. In the atlantoaxial region the presence of basilar invagination, assimilation of the atlas, C2–C3 fusion, retroodontoid and parafacetal osteophytes, bifid arches of the atlas, platybasia, a short neck, torticollis and several similar features provide evidence of the presence of atlantoaxial instability, even when this is not demonstrated by imaging [18]. Instability of the atlantoaxial and subaxial facet joints can be identified by direct observation of the facets and manual handling of the bones during surgery. An open joint cavity and the presence of excessive and abnormal movements are indications of instability of the joint. Instability of the facet joint can be present even when there is no MRI evidence of spinal changes in the segment. Alert assessment of

the presence of facetal instability during surgery and treatment can lead to a satisfactory and gratifying therapeutic outcome.

Atlantoaxial instability may be the primary nodal point of the genesis of 'degenerative' changes in the region and in the cervical spine. Corroborative evidence such as the presence of retro-odontoid soft or 'firm' tissue that represents osteophytes in the region, the presence of osteophytes around the facets and the odontoid process, and the presence of subtle atlantoaxial dislocation and basilar invagination are indicators of degenerative instability of the atlantoaxial joint [27–30]. In the presence of other evidence, instability of the atlantoaxial joint can be expected even when there is no other corroborative radiological evidence. Direct handling or manipulation of the facets can confirm the presence of instability. Ignoring atlantoaxial instability and avoiding stabilization surgery can lead to disastrous clinical consequences.

Atlantoaxial instability can be frequently associated with single or multilevel cervical spondylotic myelopathy [31, 32]. Cervical spondylosis can be primary and associated with atlantoaxial instability or even secondary to atlantoaxial dislocation. Atlantoaxial instability and prolonged neck spasm and shortening of the neck can lead to secondary spondylotic changes in the cervical spine. Long-standing shortening of the neck can lead to bone fusions and Klippel–Feil syndrome.

Cervical spondylosis is commonly identified in the lower cervical spine. The presence of degenerative changes in atlantoaxial dislocation is generally but erroneously considered extremely rare. We identified that atlantoaxial instability is frequently associated with subaxial facetal instability that leads to so-called degenerative cervical spinal changes. It might even be that multilevel cervical spinal spondylosis is invariably associated with atlantoaxial instability [31, 32]. When the clinical symptoms related to myelopathy are disproportionately greater than is evidenced by subaxial cervical spinal imaging, instability of the atlantoaxial joint can be suspected. Atlantoaxial instability in such cases is usually type B facetal instability. However, even type C facetal instability can be present. Identification of instability by direct bone handling of the atlantoaxial joint appears to be a major criterion for assessment of instability that can otherwise be missed, and an opportunity for rational and comprehensive treatment can be lost.

We recently hypothesized that instability of the spinal segments is the cause of ossification of the posterior longitudinal ligament (OPLL) and related symptoms of myelopathy [33, 34]. Accordingly, we recommended stabilization alone as the rational mode of surgical treatment. We identified that atlantoaxial instability is frequently associated with OPLL. It may be that atlantoaxial instability is the primary event and that subaxial instability and OPLL are secondary spinal events. It may also be that multilevel instability that includes atlantoaxial instability is the nodal pathogenetic factor for OPLL. Treatment by atlantoaxial and subaxial stabilization seems to be a rational mode of therapy in such cases.

Ignoring the presence of atlantoaxial instability in cases with basilar invagination, Chiari malformation, syringomyelia, cervical spondylosis, OPLL and several other common spinal ailments may be a cause of failure of treatment [35, 36]. Apart from facetal malalignment, identification of the instability on the basis of direct bone handling during surgery can be an important diagnostic criterion. The remarkable clinical recovery that can occur in the immediate postoperative period following atlantoaxial stabilization emphasizes the importance of diagnosing the instability.

Diagnosis of atlantoaxial instability by evaluation of the status of facets on imaging and evaluation of stability during surgery has opened up a new dimension in the understanding and treatment of the craniovertebral junction.

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Competing Interests The author declares that he has no competing interests.

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Ossification of the Posterior Longitudinal Ligament: Analysis of the Role of Craniovertebral and Spinal Instability

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Abstract *Background:* This paper reviews an experience of surgically treating ossification of the posterior longitudinal ligament (OPLL) with fixation of the involved spinal segments alone, without resorting to any bony or soft tissue decompression or attempts at direct resection of the OPLL. While in the early part of the experience, stabilization of only the involved subaxial cervical spinal segments was done, in the later part of the experience, atlantoaxial fixation was included in the multisegmental spinal fixation construct. This treatment is based on the understanding that spinal instability that includes atlantoaxial instability forms the nodal point of the pathogenesis and development of OPLL, and maturation of the presenting clinical symptoms.

Materials and Methods: Twenty-nine patients were treated in this series. There were 28 males and one female, and their ages ranged from 28 to 75 years (average 57 years). All patients presented with symptoms of neck pain, and progressive and disabling myelopathy-related quadriparesis. In the early part of the series (from 2012 to 2014), 14 patients underwent multilevel subaxial cervical spinal fixation by a transarticular technique of facetal fixation. After November 2014, atlantoaxial lateral mass fixation was included in the fixation construct in the subsequent 15 patients. Clinical assessments were done using a visual analogue scale (VAS), the Japanese Orthopaedic Association (JOA) scale and Goel's clinical grading scale.

Results: All patients' clinical symptoms improved in the immediate postoperative period, and the improvement was sustained and progressive in 28 patients.

Conclusion: Atlantoaxial and subaxial spinal instability seems to be the nodal pathogenetic factor in OPLL. Only stabilization of spinal segments that includes the atlantoaxial joint can provide a safe, simple and rational form of treatment.

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Keywords Atlantoaxial dislocation · Facetal fixation · Ossified posterior longitudinal ligament · Spinal instability

Introduction

Ossification of the posterior longitudinal ligament (OPLL) is a disabling cervical spinal disease. The complex structural presentation and potential for devastating postsurgical neurological complications make this disease a surgeon's nightmare. A number of possible aetiological factors have been implicated in its development, but none has been seen to be convincing or consistent [1–10]. As the pathogenesis is unclear, the treatment strategy adopted is essentially based on radiological evidence of spinal canal intrusion by the bony mass anterior to the spinal cord, and is focused on restoring spinal canal dimensions that permit an unrestricted and uncompromised traverse of the neural structures.

This paper presents an experience of 29 cases where multiple spinal segment fixation alone was done, without any form of bony or soft tissue decompression or direct resection of the OPLL. In 15 cases in the latter half of the series, the atlantoaxial joint was included in the fixation construct. This treatment strategy is based on the understanding that atlantoaxial and spinal instability forms the basis of the pathogenesis and development of OPLL [11, 12].

Materials and Methods

During the period from June 2012 to April 2016, 29 patients with OPLL were treated with fixation alone as the treatment strategy, aimed at arthrodesis of the spinal segments. This analysis of the subject includes a case experience described in two previous publications [11, 12]. There were 28 males and one female in the series, and their ages ranged from 28 to 75 years (average 57 years). All patients presented with progressive

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Table 1 Table showing the demographics of the patients, types of OPLL, and the number of spinal levels affected

Sex	Number of patients
Male	28
Female	1
Mean age (years)	57 (28–75)
Levels involved	
C1-2	2
C2-3	9
C3-4	20
C4-5	27
C5-6	23
C6-7	7
C7-T1	1
Number of levels fixed	
One-level	2
Two-level	4
Three-level	4
Four-level	6
Five-level	6
Six-level	7
Type of OPLL	
Continuous	14
Mixed	8
Segmental	5
Unclassified	2

quadriparesis as the primary symptom. Neck and hand pain were also prominent symptoms. Table 1 summarizes the clinical and radiological findings at the time of presentation and prior to surgery [13, 14]. Preoperative imaging included dynamic plain radiographs, computed tomography (CT) scanning and magnetic resonance imaging (MRI) in all patients. During the period from June 2012 to August 2014, 14 patients were treated with multisegmental subaxial cervical spinal fixation. After November 2014, in 15 patients, atlantoaxial fixation was additionally included in the fixation construct. Atlantoaxial instability was diagnosed on the basis of our recently described classification based on facetal malalignment in a neutral head position (type A: when the facet of the atlas is dislocated anterior to the facet of the axis; type B: when the facet of the atlas is dislocated posterior to the facet of the axis) or when facetal instability was identified during direct bone handling and manipulation during surgery (type C) [15]. Eleven patients had type B and four patients had type C atlantoaxial facetal instability. With use of the transarticular screw fixation technique described by Roy Camille and Saillant in 1972, subaxial spinal fixation was done [16]. Atlantoaxial fixation was done with use of the technique described by us in 1994 [17–19] (Figs. 1 and 2). For transarticular screw fixation the screws were 14 mm in length and 2.8 mm in diameter [15]. In nine patients, two screws were used for transarticular fixation as this method was considered to be safe and possible. Such 'double insurance' transarticular fixation was seen to add significant stability to the implant [20]. For atlantoaxial fixation the atlas and axis screws were 28 mm in length and 2.8 mm in diameter. A bone graft was harvested from the iliac crest and placed in the atlantoaxial joint cavity and in the appropriately prepared host bone of the midline spinal elements, which included the laminae and spinous processes and the lateral gutter. The patients were mobilized within 24 h of surgery and were advised to wear a hard cervical collar for a period of 3 months. After the 3-month healing period and confirmation of bone fusion, all activities and neck movements were permitted. Clinical assessments were done using a visual analogue scale (VAS), the Japanese Orthopaedic Association (JOA) scale and Goel's clinical grading scale.

Results

All patients showed 'remarkable' immediate postoperative clinical improvement. During the follow-up period, which ranged from 4 to 50 months (average 29 months), the improvement was sustained and progressive in 28 of the 29 patients. Tables 2, 3 and 4 depict the clinical outcome after a minimum follow-up of 3 months. One 45-year-old male who had undergone subaxial spinal fixation in a case of OPLL that extended from C2 to C6 had postoperative improvement, but his neurological condition worsened 3 months after surgery. He then underwent anterior spinal decompression surgery and was subsequently lost to follow-up. The rest of the patients are well, are improved in terms of their clinical symptoms, are independent and active, and have not needed any further surgical treatment.

All patients underwent evaluation using static and dynamic cervical spine radiographs, CT scans and MRI. Static neutral lateral radiographs were used to assess cervical sagittal balance, while anteroposterior radiographs were used to exclude abnormal coronal alignment. The lordotic angle was measured using Cobb's method of measurement [11, 12]. Preoperative assessment suggested that all patients had loss of cervical lordosis, with a lordotic angle ranging from 5° to 15°. After surgery there was a mild decrease in the lordotic angle, with the postoperative angle ranging from 4° to 12°. There was no significant difference between the preoperative and postoperative values. There were no wound infections, implant-related failures or complications. There was restriction of all spinal movements. Although all patients complained of this problem, none were unduly

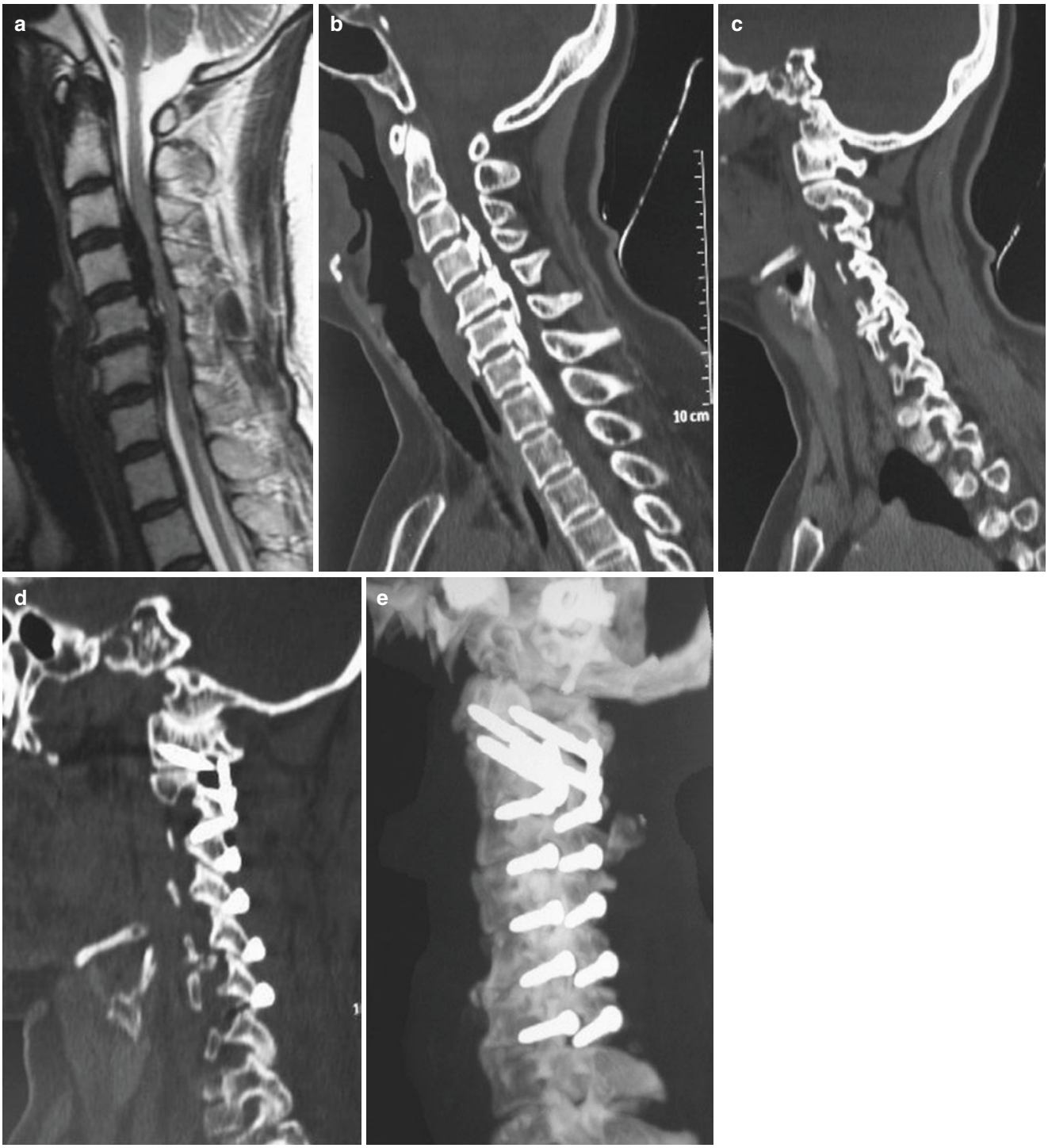

Fig. 1 Images of a 53-year-old female patient. (**a**) A T2-weighted magnetic resonance image (MRI) shows ossification of the posterior longitudinal ligament (OPLL) extending from the C3 to C6 cervical levels. (**b**) A sagittal computed tomography (CT) scan shows the OPLL. (**c**) A sagittal image shows that the facets of the atlas and axis are in alignment. (**d**) A postoperative CT scan shows the facetal implant from C1 to T1. (**e**) A CT scan shows the implant

disturbed, considering the improvement in their limbs and their general ability to perform all activities of daily living. Fusion of the spinal segment was defined as the presence of bone formation across the facet joints, with absence of all kinds of motion between spinous processes and intervertebral bodies on flexion–extension, using CT images. According to these criteria, successful bone fusion was observed in all cases at follow-up assessments.

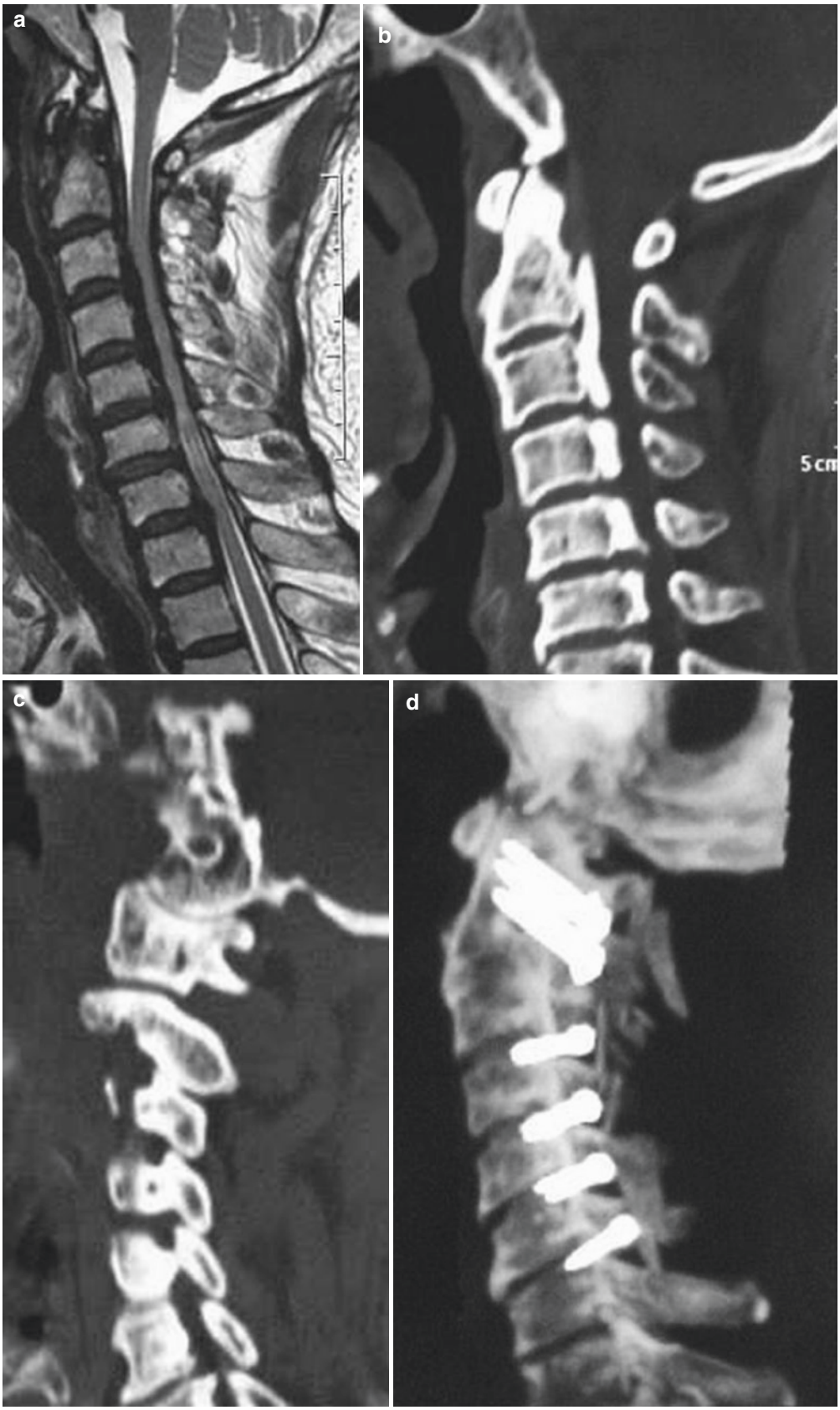

Fig. 2 Images of a 55-year-old male patient. (**a**) A T2-weighted magnetic resonance image (MRI) shows multisegmental ossification of the posterior longitudinal ligament (OPLL). (**b**) A sagittal computed tomography (CT) scan shows the OPLL. (**c**) A sagittal image of the atlantoaxial facets shows marginal type B facetal instability. (**d**) A CT scan shows the implant

Table 2 Distribution as per clinical grading system

Grade	Description	Number of patients (pre-operative)	Number of patients (post-operative)
Grade 1	Independent and normally functioning	2	14
Grade 2	Walks on own but needs support/help to carry out routine household activities	10	10
Grade 3	Walks with minimal support and requires help to carry out household activities	8	2
Grade 4	Walks with heavy support and unable to carry out household activities	7	2
Grade 5	Unable to walk and dependent for all activities	2	1

Table 3 Grading of myelopathy by the Japanese Orthopedic Association Score

Score	Pre-operative (No. of patients)	Post-operative (No. of patients)
<7	11	3
8–12	16	5
>13	2	18
16–17	–	3

Table 4 Visual analog scale (0: no pain, 10: maximum pain)

VAS score	Pre-operative	Post-operative	Post-operative (3 months)
Neck pain	6.2 (3–9)	2.1 (0–3)	0.3 (0–1)

Discussion

OPLL , with its related myelopathy, is a relatively rare clinical event. Although it has been identified throughout the world, the entity has been more frequently reported in Asian countries. OPLL frequently presents in an advanced state, when it has already occupied a significant dimension of the spinal canal. OPLL may be segmental or continuous and can extend over several spinal segments. The additional intrusion of bony elements, which compromise the diameter of the spinal canal and indent into the neural tissues, poses a significant therapeutic challenge. The location of the OPLL anterior to the spinal cord and posterior to the vertebral bodies, and its hard bony consistency, make wide surgical exposure and therapeutic resection difficult and dangerous.

The pathogenesis of OPLL is entirely unclear and has been only speculated about [1, 2, 21]. Dietary, environmental, infective and physical constitution–related factors, apart from a host of other factors, have been incriminated as possible causes [1–10, 22–24]. In general, patients with OPLL are moderately obese, have a relatively thick neck girth and in general have a sedate lifestyle. Although a number of nonsurgical treatment forms have been advocated, the progressive and devastating nature of the clinical symptoms mandates a surgical solution. The surgical treatment is difficult to conceptualize, as the pathogenesis of the disease process is unclear.

The pathology of OPLL involves introduction of additional bony elements into the spinal canal, which traverses between the spinal segments. Considering this issue, the general opinion has been that patients with OPLL have a stable spine or a spine that is more than normally stable [11, 12]. The symptoms of progressive myelopathy and a radiological appearance of severe and long segmental spinal neural compression make the correlation straightforward. Most surgeons dealing with OPLL conceptually favour direct removal of OPLL and consider it the best surgical option, associated with long-term relief from symptoms and a cure for this condition [21, 23, 25–37]. However, the formidable nature of the surgical procedure, the need for extensive bone removal for wide exposure, the high risk of neural damage during exposure and OPLL dissection off the spinal cord, and the significant risk of cerebrospinal fluid (CSF) leakage make the surgical option of direct resection of OPLL less favourable [38]. Moreover, the devastating nature of the neurological complications following a failed operation makes the entity of OPLL a generally feared disease.

Considering the issues related to direct resection of the OPLL, the majority of surgeons prefer an indirect form of decompression. Some surgeons decide on anterior or posterior decompressive surgery on the basis of the presence or absence of kyphosis or lordosis of the cervical spine. Anterior decompression involves multilevel corpectomies and discoidectomies, and posterior decompression involves a wide and long decompressive laminectomy or various forms of spinal canal–expanding laminoplasty. The issue of spinal instability and the need for bone fusion of the treated spinal segment is considered a consequence of wide and multisegmental bone removal and has been associated with immediate or delayed postoperative spinal instability. Anterior stabilization techniques that include multisegmental metal cage implants with or without associated bone grafting have been identified as a satisfactory mode of spinal stabilization after decompression [23, 25, 26, 29, 30, 34, 36]. Posterior stabilization includes a number of wire/screw and metal rod/loop/ring fixation methods [27, 30, 33, 39]. Essentially, spinal stabilization is a treatment to ward off the possible destabilizing effects of decompressive surgery.

More recently some authors have identified that instability of the spinal segments is associated with OPLL. While neural compression is a static factor of cord compromise, instability is the dynamic factor affecting cord function [2, 11, 12]. On the basis of the premise that the entire pathological sequel of OPLL is related to spinal instability, fixation alone without any form of bony or soft tissue decompression or direct resection of OPLL has been identified as an ideal form of surgical treatment [11, 12]. Despite extensive bone formation in the posterior longitudinal ligament, the articular joints are always functional and active. Direct observation of the facets suggests that the joints not only are functional but actually appear to function excessively and pathologically. In the earlier part of the present study (from June 2012 to August 2014), transarticular screw fixation was done on segments in the vicinity of the OPLL after assessment of their unstable character [12]. Clinical improvement without any decompression and with stabilization alone confirmed that instability was the defining feature in the pathogenesis of OPLL. The observations emphasized that it is not neural compression or deformation but repeated microtrauma related to instability that is the cause of the symptoms [11, 12, 40–46]. Neural structures have remarkable elasticity, plasticity and capacity to sustain deformation if it is of a long-standing and slowly progressive nature. Such deformation of the spinal cord can be observed in benign spinal tumours and in cases of syringomyelia where the cord substance may be remarkably reduced in girth but the spinal cord still retains its significant functional ability.

As the concept of spinal instability as the cause of the symptoms and pathogenesis of OPLL gained ground, it was identified that atlantoaxial instability was associated with high cervical OPLL, particularly where the OPLL extended above the level of the C3 vertebra [11, 12]. Although the atlantoaxial instability could not be identified on dynamic imaging by assessment of alteration of the atlantodental interval, atlantoaxial facetal instability was identified [15]. Types B and C atlantoaxial facetal instability, as identified in our patients, have been grouped as central or axial instability. In these two types of atlantoaxial instability the atlantodental interval may not be altered and direct compression of the neural structures by the odontoid process is not a hallmark [15]. As cord compression is not a major or primary issue in these cases, the symptoms are subtle and long-standing. It was speculated earlier that instability of the atlantoaxial joint is the primary issue and that musculoskeletal alterations, torticollis, basilar invagination, Chiari malformation type I and syringomyelia are secondary and probably protective bodily responses [47, 48]. It may be that the bone formation in the posterior longitudinal ligament is protective and a response of the body to multisegmental spinal instability [11, 12]. We had speculated earlier that the presence of a retro-odontoid pseudotumour and ossification and osteophyte formation in the degenerative spine are secondary spinal features and a response to spinal instability [49]. It was also suggested that the presence of a retro-odontoid pseudotumour and cervical subaxial osteophytes as indicators of spinal instability suggests the need for stabilization. Direct removal of a retro-odontoid tumour or osteophytes has been considered a counterproductive operative procedure [43, 44, 49]. The understanding that atlantoaxial instability is associated with high OPLL has led to inclusion of atlantoaxial stabilization in the multilevel spinal fixation construct.

With further maturation of our understanding and with increasing experience in the subject, it was realized that atlantoaxial instability is frequently associated with even mid- and low cervical OPLL. In the subsequent part of our series, all 15 patients underwent multilevel spinal fixation that included atlantoaxial fixation. Although it cannot be concluded from the present study, it may be that atlantoaxial instability is the primary nodal point of the pathogenesis of cervical OPLL. While the results regarding the extent of spinal stability in patients treated for OPLL are still under evaluation, it seems that multilevel fixation aimed at arthrodesis of spinal segments including the atlantoaxial joint appears to be a rational form of surgical treatment that addresses the nodal point of the pathogenesis of OPLL.

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Role of Subaxial Spinal and Atlantoaxial Instability in Multisegmental Cervical Spondylotic Myelopathy

Atul Goel

Abstract *Aim:* In this paper the role of atlantoaxial and multilevel subaxial spinal instability as the primary nodal point of the pathogenesis of degenerative cervical spinal disease–related myelopathy, and the focus of surgical treatment for it, is evaluated.

Materials and Methods: The series analyses the treatment of 73 patients with single or multilevel degenerative cervical spinal disease by fixation of the involved spinal segment(s) alone, aimed at arthrodesis. No bone decompression or disc/osteophyte resection was done. In 23 patients, the atlantoaxial joint was included in the spinal fixation, as atlantoaxial instability was identified by facetal malalignment on imaging or by observations on direct bone manipulation during surgery. There were 70 males and 3 females. The ages of the patients ranged from 35 to 76 years (average 57 years). The transarticular screw method was deployed for subaxial spinal fixation and a lateral mass plate/rod and screw technique was used for atlantoaxial fixation.

Results: During the follow-up period, which ranged from 3 to 42 months (average 27 months), all patients improved in terms of their clinical symptoms. There were no surgery- or implant-related complications.

Conclusion: Atlantoaxial joint instability is frequently associated with subaxial multilevel spinal instability in degenerative spinal disease. Fixation of the spinal segments provides a safe, effective and rational treatment for single or multilevel spinal degeneration.

Keywords Atlantoaxial instability · Subaxial spinal instability · Cervical myelopathy · Degeneration

Introduction

The craniovertebral junction is generally excluded from the ambit of discussion on multisegmental degenerative cervical spondylotic disease. More commonly, degenerative cervical spondylotic disease is identified in the lower cervical region and the incidence of spinal involvement progressively reduces in higher segments, and the discussion generally does not involve the spine above the C2–C3 level. This paper discusses the relationship of subaxial and atlantoaxial region 'instability' in the pathogenesis of multisegmental spinal degenerative disease, particularly in those cases associated with relatively 'severe' myelopathic symptoms.

Materials and Methods

During the period from March 2013 to June 2016, 73 patients with single- or multilevel cervical spinal degeneration were treated with fixation of the involved spinal segments alone. The case material updates and includes the experience analysed in our previous publications on the subject [1, 2]. Atlantoaxial joint fixation was included in the fixation construct after June 2013. Among 40 subsequent cases of treatment for multilevel spinal degeneration, 12 cases included atlantoaxial fixation. The criteria to include the atlantoaxial joint in the fixation construct included the presence of facetal malalignment in a neutral head position (type A and type B facetal instability) and observation of atlantoaxial instability during manual manipulation of bones in the region during surgery (type C facetal instability) in the presence of severe myelopathy symptoms (Goel grade 3–5) [3] (Figs. 1 and 2).

There were 70 males and 3 females. All patients had chronic or long-standing symptoms related to myelopathy with or without radiculopathy. The duration of significant neurological symptoms ranged from 6 to 24 months (average 10 months). The study excluded patients with radicular

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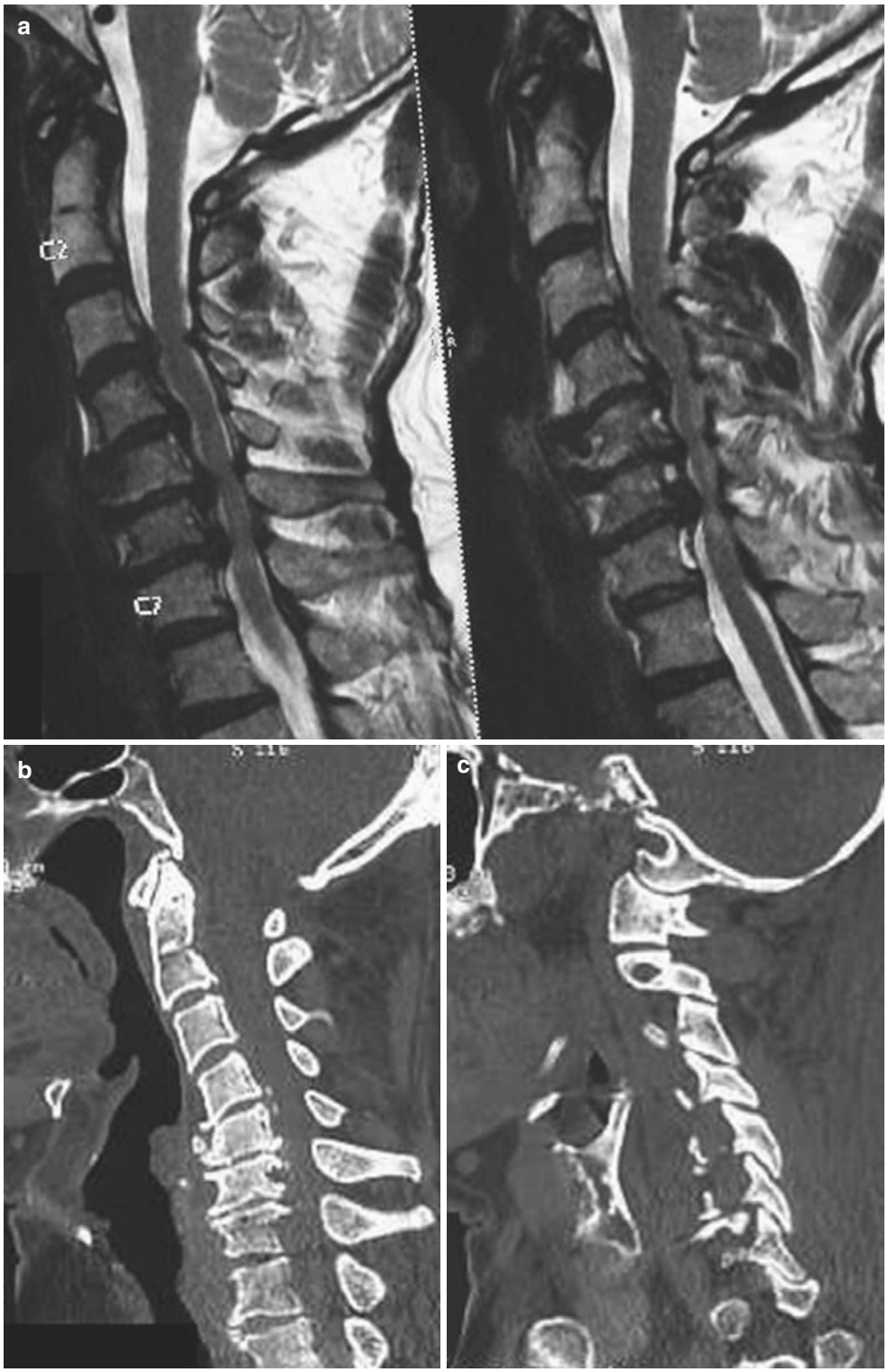

Fig. 1 Images of a 70-year-old male patient. (**a**) A T2-weighted magnetic resonance image (MRI) shows multilevel cervical degenerative changes. (**b**) A sagittal computed tomography (CT) scan shows changes of spinal degeneration. (**c**) A sagittal CT scan shows the facets; the atlantoaxial facets are normally aligned. (**d**) A postoperative CT scan shows transarticular screw fixation. (**e**) A postoperative CT scan shows reduction of kyphosis. (**f**) A sagittal cut through the facets shows the transarticular screws

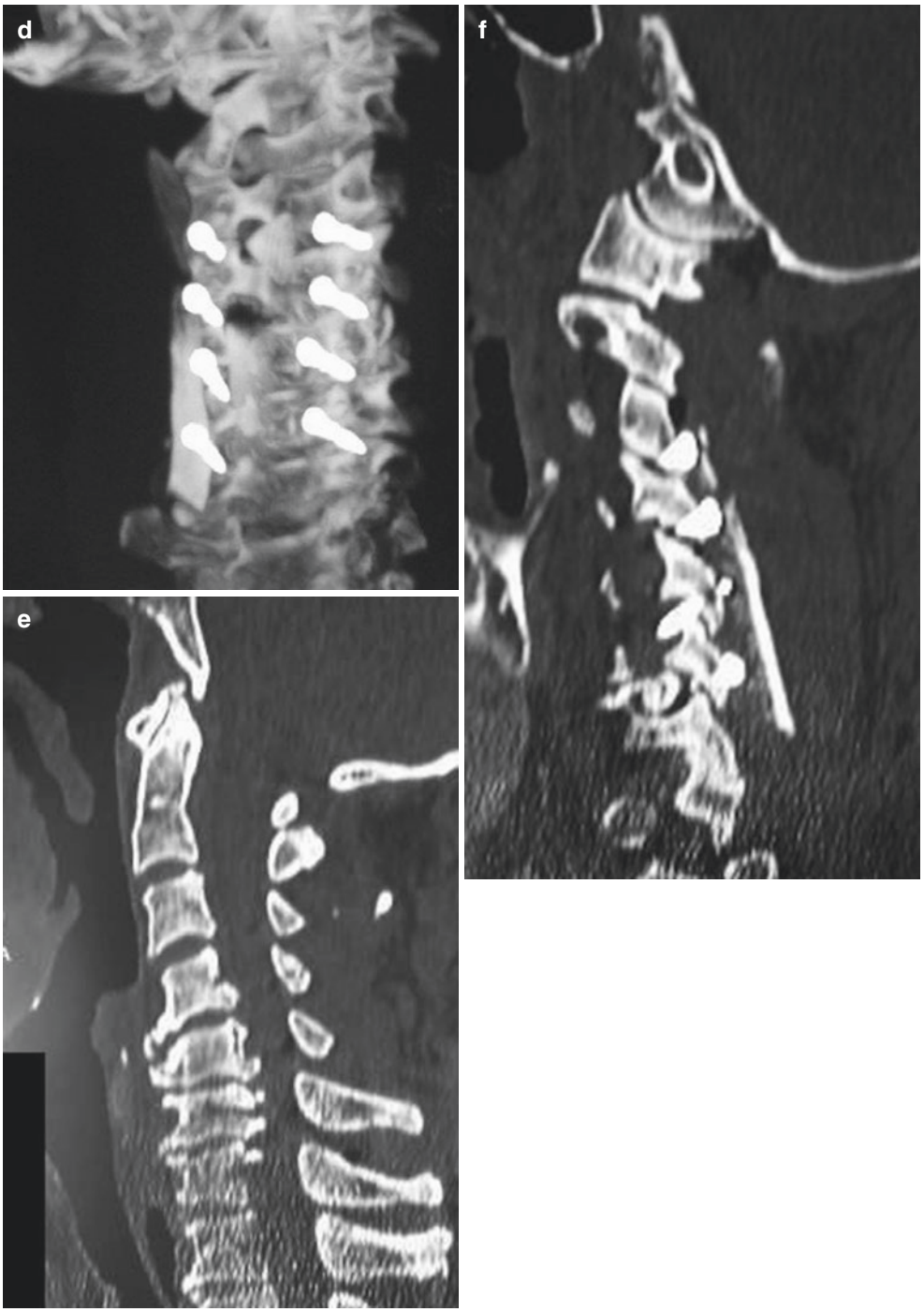

Fig. 1 (continued)

symptoms alone, patients with acute symptoms related to disc herniation, patients with a history of significant trauma to the head or the neck, and patients with bone anomalies that included fusions.

The clinical and radiological features at the time of presentation are detailed in Table 1. Clinical assessments on the basis of a five-point clinical grading scale [4], the Japanese Orthopaedic Association (JOA) scale [5] and a visual analogue scale (VAS) [6] are elaborated in Tables 2, 3, and 4. Preoperative investigations included dynamic plain radiography, computed tomography (CT) scanning and magnetic resonance imaging (MRI). The patients were placed in a prone surgical position with the head under traction. The exposure included C1–C2, and the subaxial spinal exposure

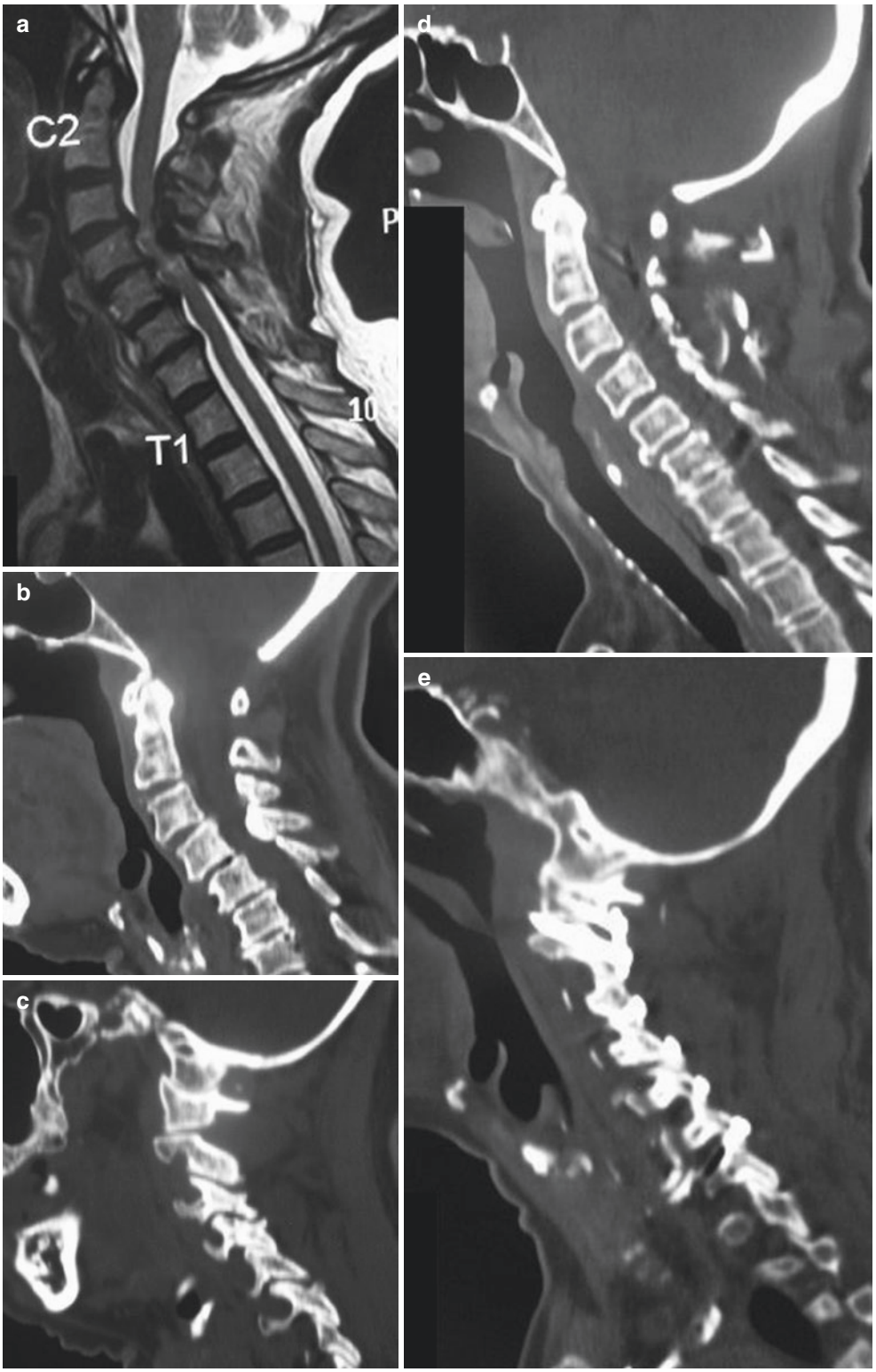

Fig. 2 Images of a 55-year-old female. (**a**) A T2-weighted magnetic resonance image (MRI) shows multilevel cervical spinal degeneration and evidence of cord compression; there is no cord compression at the level of the craniovertebral junction. (**b**) A sagittal computed tomography (CT) scan shows multilevel spinal degeneration. (**c**) A sagittal CT scan passing through the facets; type 2 atlantoaxial facetal instability can be observed. (**d**) A postoperative sagittal CT scan. (**e**) A postoperative CT scan shows atlantoaxial and subaxial facetal fixation

Table 1 Table showing the presenting clinical and radiological features

Sex	
Male	70
Female	3
Mean age	57 (35–76)
Number of levels fixed	
Two-level	5
Three-level	26
Four-level	29
Five-level	9
Six-level	4

depended on the extent of spinal degeneration. Transarticular fixation was done for the subaxial spine, using the technique described by Camille and Saillant [7]. For C1–C2 fixation a lateral mass plate and screw technique was used, as described by us in 1994 [8, 9]. The levels of spinal fixation are elaborated in Table 1. For transarticular screw fixation, screws 14 mm long and 2.8 mm in diameter were used [10]. For atlantoaxial fixation the screws measured 28 mm in length and 2.8 mm in diameter. A bone graft harvested from the iliac crest was placed into the atlantoaxial articular cavity, into the facet joints alongside the screws and in the midline after appropriate preparation of the host bone over the laminae and spinous processes. Postoperatively the traction was removed and the patients were advised to wear a cervical collar for a period of 3 months.

Results

All patients in the series improved in terms of their symptoms in the immediate postoperative phase, and the improvement progressed subsequently. The follow-up period ranged from 3 to 42 months (average 27 months). Tables 2, 3, and 4 depict the preoperative clinical status and the postoperative clinical status 3 months after surgery. During the period of follow-up (range 3–42 months, average 27 months), no patient showed delayed neurological worsening or needed any further surgery on the cervical spine. Bone fusion could be observed in the facets and lamina in all cases where the follow-up was more than 6 months. There were no infections or implant failures.

Discussion

The analysis of degenerative spinal disease has been 'disc-centric' for several decades. The age-related reduction in the water content of the disc and its effects on disc height have been implicated as the genesis point of the entire spectrum of degenerative spinal disease [11–14]. The prominence of 'empty space' seen on plain radiography images allows the clinician to evaluate its pathology without actually visualizing the disc. Osteophyte formation, ligamentous 'hypertrophy', retrolisthesis of the facets and similar features have been discussed more often as associated pathological entities and less frequently as a secondary response to primary disc disease [11–13, 15]. The overall consequence of degenerative spinal disease is a reduction in the dimensions of the spinal canal and neural foramina. With technological improvements in radiological investigations and the advent of computer-based technology, the radiological evidence of cord compression and its effects on the cord substance have become more starkly evident. With imaging clarity showing cord compression, surgical endeavours have been focused on removing compressing factors, 'decompressing' neural

Table 2 Distribution as per clinical grading system

Grade	Description	Number of patients (pre-operative)	Number of patients (post-operative)
Grade 1	Independent and normally functioning	–	29
Grade 2	Walks on own but needs support/help to carry out routine household activities	27	32
Grade 3	Walks with minimal support and requires help to carry out household activities	30	7
Grade 4	Walks with heavy support and unable to carry out household activities	12	3
Grade 5	Unable to walk and dependent for all activities	4	2

Table 3 Table showing the pre-operative and post-operative clinical assessment as per JOA scoring system

JOA score	Pre-operative (No. of patients)	Post-operative (No. of patients)
<7	20	3
8–12	41	9
13–15	12	33
16–17	–	28

Table 4 Visual analog scale (0: no pain, 10: maximum pain)

VAS score	Pre-operative	Post-operative	Post-operative (3 months)
Neck pain	5.9 (4–7)	1.5 (0–3)	0.4 (0–1)

structures, increasing the spinal and neural canal dimensions, and providing a free traverse for the neural structures [12, 16, 17].

Although instability of the spine and spinal segments is sometimes considered in the overall pathogenesis of disease and of clinical symptoms, its *numero uno* position in the entire scenario of degenerative disease has not been extensively evaluated and therapeutically exploited. In general the issue of instability and the need for stabilization and fusion are considered, as decompressive surgery involves significant bone/soft tissue/disc/osteophyte removal, using either an anterior or a posterior approach, and can have a destabilizing effect on the spinal column in the immediate postoperative period or as a delayed consequence [12, 16–18].

Despite the advances in computer-based radiological imaging, the facets and their instability continue to be inadequately visualized because of their lateral location and oblique profile. A recent evaluation of the subject identified that instability of the spinal segments is the primary nodal point of the pathogenesis of degenerative spinal disease [1, 2, 10, 19–24]. Standing human posture and disuse/misuse or injury of the paraspinal muscles lead to 'vertical' spinal instability, which is manifested by telescoping or slipping of the superior facet over the inferior facet. This vertical instability has also been labelled as *retrolisthesis of facets* [22]. Retrolisthesis has been previously identified to be a consequence of disc degeneration and disc space reduction. It appears that subtle instability of the spinal segment(s) may be paramount in the pathogenesis of the entire structural deformation. Such instability is rather easily observed on direct visualization of the joint during surgery, even when preoperative dynamic radiographs do not depict it. Identification of the fact that instability of the spine is the primary event, and that the rest of the so-called pathological features (such as disc space reduction, osteophyte formation and ligamentous pathology) are secondary and possibly protective in function, has the potential to revolutionize the treatment paradigm for the entity of 'spinal degeneration' [1, 2, 10, 19–24]. Stabilization of the affected spinal segment addresses the primary pathology of the disease process [1, 2, 15, 21–27].

In the year 2010, we introduced facetal distraction of the affected spinal segments as a stand-alone form of treatment for patients with single or multilevel disc degeneration who presented with radiculopathy or myelopathy [19, 20, 25–27]. Specially designed 'Goel facet spacers' were impacted into the intra-articular cavity after appropriate distraction of the facets. The technique was aimed at achieving distraction–arthrodesis of the affected spinal segments [25–27]. The observation that distraction of the facets resulted in immediate postoperative reversal of all known pathological features of the degenerative spine gave credence to this hypothesis [25–27]. Distraction of the facets resulted in stretch reversal of buckling of circumferential intervertebral ligaments, an increase in the disc space height and an increase in the spinal and neural canal dimensions. It was identified that with distraction of the facets, there was potential for regression of osteophytes. We recently reported immediate postoperative regression of 'contained' disc herniation, which had displaced the posterior longitudinal ligament, following distraction of the facets [27]. Essentially, the operation involved stabilization of the spinal segment and avoided the need for resection of any part of the bone, soft tissues, osteophyte or disc [21]. The treatment thus changed to dealing with posteriorly located facets than an anteriorly located disc and from surgery that was previously aimed at decompression of neural structures to that directed at spinal fixation.

On further evaluation of the subject, it appears that it is not neural compression or deformation but repeated microinjuries to the spinal cord, as a result of instability, that is the cause of neurological symptoms [21]. It was realized that more than distraction of the spinal segments, it is their stabilization that is most important for restoration of neural function and amelioration of symptoms. Accordingly, we resorted to fixation alone as the treatment for single or multilevel spinal degeneration–related radiculopathy or myelopathy [1, 2, 15, 23, 24]. Transarticular facetal fixation was identified to be a safe, strong and rather straightforward surgical option for fixation. The oblique profile, relatively large size and biomechanical strength of the facets can be used effectively and safely for transarticular screw insertion [10]. The mineral density of the bones of the facets and pedicles is significantly superior to that of any other part of the vertebra, imparting greater strength to the process of screw implantation. Moreover, the fixation is at the fulcrum of all spinal movements. Extension of the levels of fixation is relatively easy and remarkably quick.

Although instability of the subaxial facets is difficult to evaluate radiologically, because of their oblique profile, atlantoaxial facetal instability is relatively easy to decipher because of the brick-over-brick rectangular configuration. Our studies have concluded that the atlantoaxial joint, which is the most mobile joint in the body, is most likely to develop instability [2, 23]. An alternative classification divided atlantoaxial dislocation on the basis of facetal malalignment and identification of instability on direct bone manipulation during surgery [3]. Atlantoaxial instability in the absence of disturbance of the atlantodental interval and direct compression of neural structures by the odontoid process was labelled as *central or axial atlantoaxial dislocation*. In such a form of instability the facet of the atlas is dislocated posterior to the facet of the axis in a neutral head position (type B atlantoaxial facetal dislocation), or the facets of the atlas and axis are in alignment but the dislocation or instability is identified by direct bone handling or manipulation during surgery (type C atlantoaxial facetal instability). As neural compression is not an early or prominent feature, the symptoms are chronic and long-standing in nature. Such forms of disloca-

tion are associated with chronic pathological entities such as group B basilar invagination, Chiari malformation type I, syringomyelia, a short neck, torticollis, bone fusions and several features that seem to be long-standing protective responses to atlantoaxial instability [4, 5, 24, 28–31]. Central or axial atlantoaxial instability is also associated with chronic degenerative spinal changes that manifest as multilevel spondylosis. On the other hand, in type A facetal instability, where in lateral-profile imaging the facet of the atlas is dislocated anterior to the facet of the axis, the symptoms are relatively acute, as there is an abnormal alteration in the atlantodental interval and compression of the spinal cord by the odontoid process. Type A facetal instability is less frequently associated with chronic pathological entities such as degeneration-related spinal disorders. It appears that atlantoaxial instability could be a primary pathology that leads to secondary degenerative changes in the cervical spine, or it may be associated with multilevel spinal instability. Identification of atlantoaxial instability and subaxial multilevel spinal instability and stabilization may provide a rational form of treatment for multilevel spinal degeneration. All of our patients who were treated with such a surgical strategy of fixation alone showed a remarkable recovery from their clinical symptoms in the immediate postoperative period and at follow-up assessments [1, 2, 15, 23, 24].

The exact indication for inclusion of the atlantoaxial joint in the fixation construct will have to be evaluated by further clinical studies. Moreover, larger case studies with multiple treating surgeons will have to collect and compile their observations to validate the concept. The efficacy and safety of the techniques of fixation we have adopted are apparent from our successful results. The technique has resulted in demonstrated improvements in gait, strength, sensations, pain, degree of myelopathy, and bladder and bowel control. We have not observed recurrent disease, pseudoarthrosis or hardware failure in any case. The drawback of this study is that it did not include a comparative cohort of patients who had undergone traditional open anterior or posterior surgery.

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Competing Interests The author declares that he has no competing interests.

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The Craniovertebral Junction in Rheumatoid Arthritis: State of the Art

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Abstract Rheumatoid arthritis (RA) is a chronic inflammatory disorder, characterized by polyarticular inflammation causing progressive joint damage and disability. The mechanisms underlying its pathogenesis involve activation of innate and adaptive immunity, microvascular endothelial cell activation, and inflammatory infiltration of lymphocytes and monocytes into the synovium. Spinal involvement in RA is not typical; when it occurs, the main radiological features are (1) atlantoaxial subluxation (AAS), which is the most typical form of cervical spine involvement; (2) cranial settling—also known as basilar impression, atlantoaxial impaction or superior migration of the odontoid—which is the most severe form of associated spinal instability; and (3) subaxial subluxation. A combination of these alterations may occur. Synovitis is characterized by infiltration of innate and adaptive immune cells; joint destruction is a consequence of activation of synovial fibroblasts, which acquire aggressive, inflammatory, invasive features, associated with increased chondrocyte catabolism and synovial osteoclastogenesis.

Neck pain is the most frequent symptom of spinal involvement in RA; it occurs in 40–80% of patients and is mostly localized at the craniocervical junction. Other symptoms—caused by compression of neural structures such as the greater occipital nerve (at C2), the nucleus of the spinal trigeminal tract and the greater auricular nerve—are occipital neuralgia, facial pain and ear pain, respectively. Irritation of the lesser occipital nerve (at C1) can cause pain in the suboccipital region. Sometimes patients may complain of a sensation of their head falling down with flexion, weakness, reduced endurance, loss of ability, gait alterations, paraesthesias or other symptoms due to cord and medullary compression, and upper or lower motor neuron signs, or both. Surgical management of RA remains a challenging field.

Keywords Craniovertebral junction · Rheumatoid arthritis Cervical spine · Inflammation · Transnasal decompression Transoral decompression · Instrumentation and fusion procedures · Atlantoaxial dislocation · Atlantoaxial instability · Atlantoaxial synovitis · Basilar invagination

Introduction

Rheumatoid arthritis (RA) is a chronic inflammatory disorder, characterized by polyarticular inflammation causing progressive joint damage and disability. The mechanisms underlying its pathogenesis involve activation of innate and adaptive immunity, microvascular endothelial cell activation and inflammatory infiltration of lymphocytes and monocytes into the synovium. The final consequence of these immunological processes is development of synovial hypertrophy with pannus formation, which finally leads to erosion of articular cartilage and subchondral bone [1].

RA is a chronic, symmetrical, erosive disease, mainly involving small joints in the hands and feet. However, cervical spine involvement may also be present in a significant number of patients, sometimes in early disease. Cervical spine involvement, however, is more commonly a finding of long-standing RA, observed in over half of all patients after a mean of 10 years of the disease [2, 3].

The prevalence of cervical spine abnormalities in RA is estimated to be between 17% and 88% [4, 5]. The breadth of this range is related to variability in populations (as observed in several retrospective studies), use of different classifications of the disease and evolving medical therapies for the disease [6]. Almost 1% of the general population in Europe and in the

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USA is affected by RA, and approximately 10% of these patients develop significant cervical spine involvement [7].

Cervical spine involvement in RA is related to the presence of peripheral erosions, the use of corticosteroids and previous joint surgery, which are considered independent risk factors for development of serious cervical spine abnormalities. Beyond inflammatory involvement of cervical spine related to RA, patients can develop age-related degenerative alterations (known as cervical spondylosis), which also affect the general population [8].

Anatomy and Physiopathology

The cervical spine can be separated into two different parts: the upper tract (C1 and C2, with the atlantoaxial, atlanto-odontoid and atlanto-occipital joints) and the lower tract (C3–C7, with uncovertebral and facet joints present at each level). The upper cervical spine is mainly involved in rotational movements of the neck, whereas the lower tract is involved in flexion–extension movements. Like the rest of the spine, the cervical spine has the function of guaranteeing protection of the neural structures contained in the spinal canal and allowing both physiological stability and movement capacity between the vertebrae through the joints connecting them, which are powered by various muscle attachments. The occiput–C1 and C1–C2 articulations lack intervertebral discs, consisting exclusively of synovial joints. Thanks to these peculiar anatomical features, the atlas and axis allow increased mobility of the cervical spine. The atlas, without a vertebral body, supports the head though lateral articulations with the occipital condyles. The superior articular facet of the atlas receives occipital condyles at the base of the skull, whereas the inferior articular facet stands upon the axis; articulation between the atlas and axis is permitted by vertical projections of the odontoid process, which assumes a position between the lateral masses of C1. The joints are stabilized by different ligaments, such as the transverse ligament, the alar ligaments and the accessory atlantoaxial ligaments [9, 10]. The integrity of the transverse ligament prevents anterior subluxation of the atlas, particularly during neck flexion. In RA, this ligament is often compromised because of its involvement in the inflammatory process of the synovial articulation of the dens. Whole rupture of the transverse ligament causes only 4–5 mm anterior subluxation of the atlas if the secondary stabilizers are not damaged. In RA, the stability of the atlantoaxial joint is definitely compromised as a result of impairment of the secondary stabilizers, and it can be further compromised by possible erosions of the odontoid process. Atlantoaxial instability (most frequently anterior) is often due to loss of ligament support caused by development of erosive pannus at the C1–C2 level and bone destruction (Fig. 1). This process leads to damage to the ligamentous complex that usually stabilizes the atlas in the axis, especially damage to the transverse ligament, but also to the articular capsular joint of C1–C2.

The weight of the head and insufficient mobility of the thoracic spine create dynamic forces that worsen the situation, further compromising the ligamentous stabilizers, causing fracture of an impaired dens, or both.

The most frequent clinicopathological presentations of spinal involvement in RA are (1) atlantoaxial subluxation (AAS), which is the most common manifestation of cervical damage; (2) cranial settling—also known as basilar impression, atlantoaxial impaction or superior migration of the odontoid—which is the most severe form of spinal involvement in RA; and (3) subaxial subluxation (SAS). A combination of these alterations may occur [11, 12].

In AAS, the normal <3 mm range of the anterior atlantodental interval (ADI) is extended and that of the posterior atlantodental interval (PADI)—the space between the posterior border of the dens and the anterior aspect of the posterior arch of C1—is decreased, inducing compression of the upper spinal cord [12]. Winfield et al. [13] observed anterior atlantoaxial dislocation (AAD) in 12% of RA patients during a follow-up period of 7 years. In autopsy studies, the rate changed from 11% to 46% [9]. Anterior AAD accounts for 75% of all cases of AAD. According to evidence from

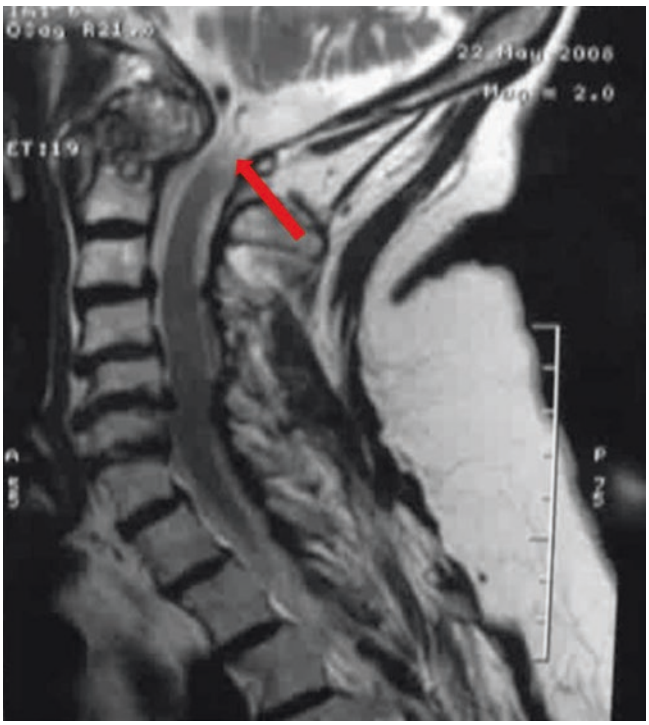

Fig. 1 Magnetic resonance imaging (MRI) T2-weighted sagittal reconstruction of retro-odontoid pannus impinging on the bulbomedullary junction in a 68-year-old woman (*red arrow*)

experimental studies, disruption of the transverse ligament alone can cause slipping of C1 anteriorly over C2. When the transverse ligament is the only one compromised, the maximum displacement is about 5 mm, but it increases to 6.5–10 mm and 7 mm when the alar or atlantoaxial ligaments, respectively, are also compromised, and 12 mm when all three ligaments are damaged [10, 14].

In 20% of cases of AAD, C1 is shifted laterally [15], causing an abnormal head posture. Lateral AAD is the result of unilateral or asymmetrical involvement of the lateral atlantoaxial joint. When no more than 1 mm of subchondral bone on the lateral mass of C1 or the articular process of C2 is lost, C1 shifts laterally by 2.5 mm. When the loss of bone is more than 1 mm, the shift can reach 5 mm and is limited by contact between the lateral mass of C1 and the dens. At the same time, the lateral mass of C1 is in contact with C2, shifting and tilting C1. An anteroposterior open-mouth x-ray permits diagnosis, evidencing involvement of either or both of the C1 and C2 joints with a major than 2 mm displacement of C1 on C2 and tilting of C1 on C2. The degree of compromise of the lateral mass of C2 determines the extent of the tilting [15]. Rotatory dislocation is caused by unilateral C1–C2 joint damage with impairment of the transverse ligament. Dislocation is well demonstrated by an open-mouth projection scan, which is considered the best x-ray projection because it shows lateral displacement of the dens, asymmetry of the C1 lateral masses with respect to the dens and abnormal lateral mass geometry (Fig. 2a). A rarer manifestation is posterior atlantoaxial subluxation, which is usually caused by a fracture of the dens and carries a greater risk of cord injury than AAS (6–7%) [16]. The dens damage caused by the pannus underlies posterior subluxation, subsequent to

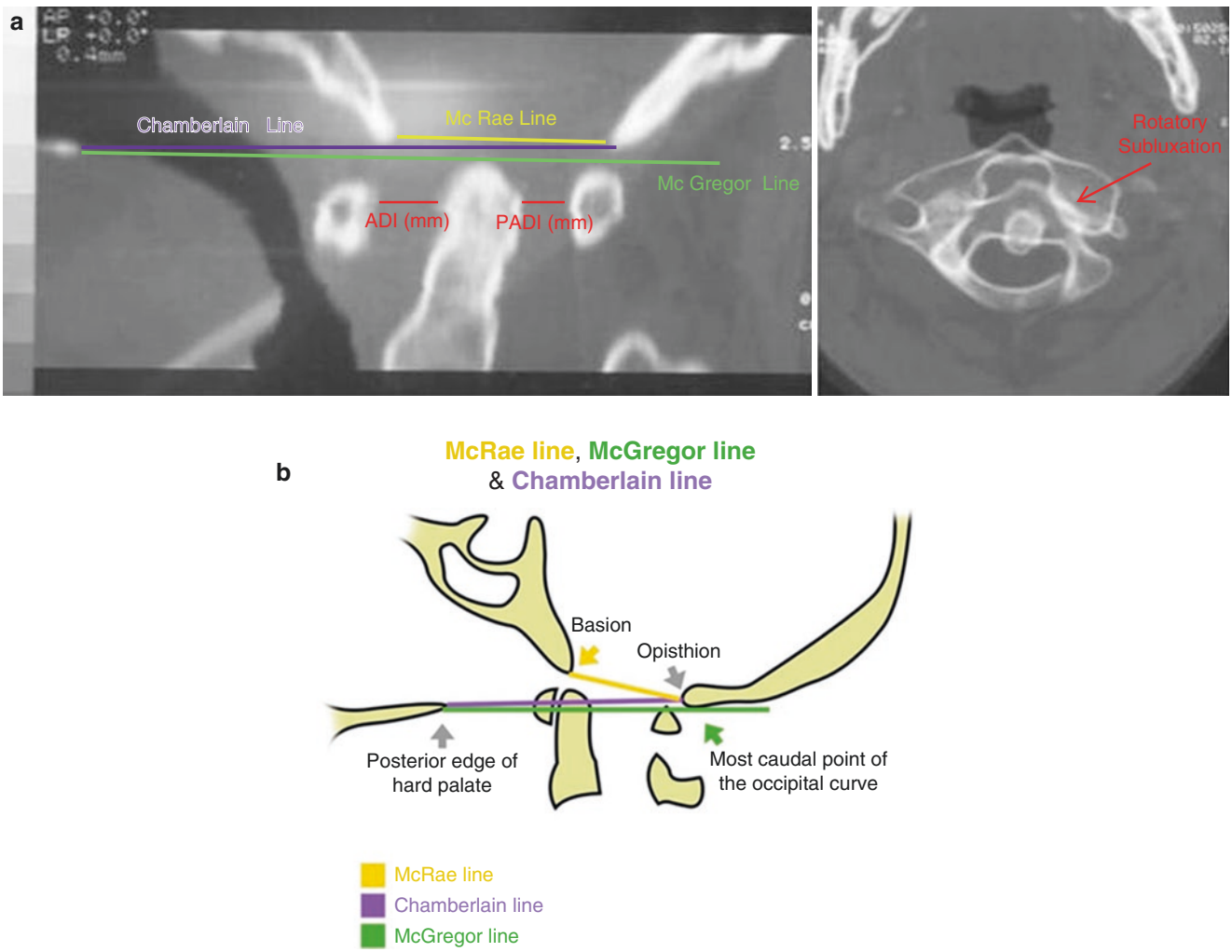

Fig. 2 (**a**) Traditional X-ray measurements investigating the C1–C2 relationship. *ADI* anterior atlantodental interval, *PADI* posterior atlantodental interval (*left*) and rotatory subluxation (*right, red arrow*). (**b**) Radiological criteria for cranial settling. Chamberlain line: findings are considered positive if the apex of the odontoid is 3 mm above a line from the posterior edge of the hard palate to the opisthion (the posterior rim of the foramen magnum). McGregor line: findings are considered positive if the apex of the odontoid is >4.5 mm above a line drawn from the posterior hard palate to the most inferior point on the occipital curve. McRae line (1953): findings are positive if the tip of the odontoid extends above a line drawn from the basion (the anterior rim of the foramen magnum) to the opisthion

posterior slippage of C1. The result is upward movement of the anterior arch of C1 and downward inclination of the posterior arch until it is located in front of the spinous process of C2. The erosion of the occipitus–C1 and C1–C2 joints causes cranial settling, with displacement of the odontoid in the foramen magnum. This can cause brainstem compression and possibly death [17].

Cranial settling is observed in 4–35% of RA patients and is responsible for 20% of all AAD cases [15, 18]. It is caused by erosion of the occipital condyles, the superior articular processes of C2 and the lateral masses of C1. The collapse of the compromised joints is followed by settling of the skull on the upper cervical spine.

Subaxial subluxation can cause several abnormalities in the cervical spine, ranging from spondylolisthesis to cervical kyphosis and/or a "staircase" deformity due to the destruction of the facet joints as well as the uncovertebral joint. Plain X-rays permit us to visualize subaxial cervical spine abnormalities. Spinal cord compression is rarer than with upper cervical spine lesions but it can result in more severe lesions [19].

Pathogenesis

The pathogenesis of RA is heterogeneous and is characterized by activation of innate and adaptive immunity. The presence of autoantibodies is a negative prognostic factor and is related to more severe clinical manifestations, erosive disease and increased mortality [20, 21]. The increased mortality seems to be probably related to the presence of immune complexes between autoantibodies against citrullinated peptides (ACPAs) and citrulline-containing antigens, leading to complement activation [22]. The lung seems to be the main tissue in which the immune response is activated, through formation of ACPAs, which is increased by smoking. Several studies have demonstrated an association between smoking and the presence of shared citrullinated peptides in both lung and synovial tissue biopsies [23]. ACPAs themselves have a pathogenetic role, triggering innate immunity by directly stimulating macrophages (e.g. by binding to toll-like receptors through the bound antigen, by Fc-receptor engagement, or both). Recent studies have suggested that ACPAs could promote activation of osteoclasts through development of immune complex and Fc-receptor involvement or probably by ligating membrane citrullinated vimentin [24], therefore directly causing bone loss. In RA, activation of immune responses leads to development of leucocyte infiltration in synovial membranes, causing synovial hypertrophy, which clinically occurs with joint swelling. The cellular composition of synovitis in RA is heterogeneous, including innate immune cells (e.g. monocytes, dendritic cells, mast cells and innate lymphoid cells) and adaptive immune cells (e.g. T-helper-1 and T-helper-17 cells, B cells, plasmablasts and plasma cells) [24]. Joint destruction is a consequence of activation of synovial fibroblasts, which acquire aggressive inflammatory and invasive features, associated with increased chondrocyte catabolism and synovial osteoclastogenesis [25, 26]. Ultrasound-guided biopsies of small joints and detailed molecular analyses (in particular, transcriptomic analyses) have demonstrated the presence of different subtypes of synovial inflammatory infiltrates—namely, myeloid-dominant, lymphocytic-dominant and fibroid-dominant infiltrates—providing new insights into pathogenetic processes, which could have significant therapeutically implications [27].

The mediators of inflammation that are mainly implicated in progression of RA are chemokines, cytokines, growth factors and metalloproteinases. The immune response is characterized by activation and attraction of inflammatory cells from peripheral blood to the site of inflammation and by subsequent proliferation of synoviocytes. Tumour necrosis factor (TNF), interleukin 6 and probably granulocyte–monocyte colony-stimulating factor play central roles in the development and maintenance of the inflammatory process [28].

Activated fibroblasts—in synergy with inflammatory infiltrates composed of activated T and B cells, monocytes and macrophages—finally act by triggering osteoclast activation through the receptor RANKL (receptor activator of nuclear factor κB ligand, expressed on T and B lymphocytes and fibroblasts), which interacts with its cognate receptor, RANK, expressed on preosteoclasts, macrophages and dendritic cells [29, 30]. Activation of osteoclasts induces the appearance of joint erosions by release of proteolytic enzymes (matrix metalloproteinases). The cartilage matrix is degraded by matrix metalloproteinases and aggrecanases. The proteases can consequently infiltrate and damage articular cartilage, subchondral bone, tendons and ligaments, ultimately leading to instability and subluxation of all of the joints involved, including the cervical spine.

Clinical Manifestations

Cervical spine involvement in RA is often a silent condition. Even in the presence of severe cervical spine damage, many patients may be asymptomatic. Beyond RA-related inflammation in the cervical spine, this site can also be compromised by degenerative age-related disease. Both conditions occur with almost the same symptomatology, characterized by neck pain, myelopathy and radiculopathy. Neck pain is the most frequent symptom reported by patients with spinal involvement in RA; it occurs in 40–80% of patients [31], mostly localized at the craniocervical junction. Occipital headaches are often present as well. Compression of neural

structures—such as the greater occipital nerve (in C2), the nucleus of the spinal trigeminal tract and the greater auricular nerve—can cause occipital neuralgia, facial pain and ear pain, respectively. Irritation of the lesser occipital nerve (in C1) can also cause pain in the suboccipital region. Sometimes patients may complain of a feeling of their head falling down with flexion and with a clunking sensation, weakness, reduced endurance and dexterity, gait alterations, paraesthesias [12, 32] or other symptoms due to cord and medullary compression, and upper or lower motor neuron signs, or both.

Axial pain originates from involvement of various components of the vertebral column and is due to damage of the bone and cartilage in the neck or deformities resulting in misalignment.

Myelopathy describes a constellation of symptoms usually caused by compression of the brainstem or spinal cord; earlier symptoms can worsen coordination of hand movements and cause disturbances in balance, a sensation of heaviness in the lower extremities and gait disorders. Other clinical manifestations may be difficulty in execution of fine motor skills—such as buttoning a shirt or inserting a key into a lock—or a progressive change in handwriting [33].

Neck movements, particularly flexion, can cause the occurrence of electric shock sensations in the torso and extremities (Lhermitte's sign), or either alone. Vertebrobasilar insufficiency or mechanical compression of the cervicomedullary junction can cause symptoms of tinnitus, vertigo, visual alterations, disturbances of equilibrium, diplopia and dysphagia; the most severe manifestations of upper cervical spine involvement, caused by vertebrobasilar insufficiency, are stroke or sudden death, which are rarely reported [12, 34].

A complete history and physical examination can highlight bowel and bladder disturbances, movement disabilities, balance and coordination alterations, and/or difficulties in manual dexterity. Physical examination findings can differ significantly and have been demonstrated to be variable in patients with cord compression. Beside Lhermitte's sign, suggestive signs of myelopathy are Hoffman's sign, an inverted brachioradialis reflex, hyperreflexia, continuous clonus or more than five beats of clonus, a positive Romberg sign and/or a Babinski sign, as well as the occurrence of dysdiadochokinesia, dysmetria or problems with the heel to shin test and/or the tandem gait test. These signs/symptoms can be associated with various levels of motor and/or sensory alterations in the upper and/or lower extremities. A complete neurological examination should be performed, including all of the aforementioned provocative tests. Unfortunately, in patients with RA, severe joint involvement may sometimes prevent a thorough neurological examination from being done, delaying diagnosis and making it more difficult [35].

Diagnosis

Patients with RA, even asymptomatic ones, should undergo an X-ray evaluation in lateral, anteroposterior (AP) and open-mouth odontoid views, and in lateral flexion–extension dynamic projections, to assess cervical spine involvement [12, 36, 37]. Radiographic alterations related to early cervical spine involvement are most commonly represented by odontoid erosions, disc narrowing, and atlantoaxial and subaxial subluxation [38, 39]. X-ray evaluation permits clear identification of bone alignment, quality and deformities, but it has some limitations for identification of bony erosions, the craniovertebral junction (CVJ) and cervicothoracic junctions (because of superimposition of the cranial base structures and of the glenohumeral joints) and soft tissue changes such as pannus and spinal cord compression. In the presence of radiographic alterations in cervical spine involvement and/or in cases of neurological symptoms or cervical pain, computed tomography (CT) scans and/or magnetic resonance imaging (MRI) of the cervical spine are mandatory [11, 40].

CT scanning with multiplanar reconstruction represents the gold-standard method for clear visualization of bone changes (erosions, anatomy, ankylosis and spondylosis) but has limitations for assessment of soft tissues, the spinal cord and nerve roots [11, 41]. MRI evaluation is the most sensitive method for evaluation of cervical spine involvement in RA, especially because it allows better visualization of soft tissue and neural elements. It should be performed in all patients with suspected or confirmed radiographic signs of cervical spine involvement and in patients complaining of neurological symptoms [42, 43] (Fig. 1). Both CT scanning and MRI are fundamental for surgical planning.

As reported above, traditional X-ray evaluation, considering the C1–C2 relationship, includes the ADI; a distance of <3 mm is considered normal. However, patients with RA, even asymptomatic ones, frequently have measurements of 5 mm or even up to 10 mm [39]. In this regard, it has been demonstrated that the PADI is a better predictor of paralysis and recovery. The PADI, in particular, estimates the greater amount of space available for the upper cervical spinal cord [39, 43] (Fig. 2a). The cervical spinal cord occupies 10 mm of the canal diameter; additionally, it needs 1 mm for the dura and 1 mm for the cerebrospinal fluid (CSF) anterior to the cord, and the same posteriorly, for a total of 14 mm. Compression of the cord occurs when the available space is <14 mm. X-ray evaluations should include both PADI and ADI measurements obtained in flexion and extension. Boden et al. [39] observed a high rate of neurological recovery, after fusion and stabilization, in patients with a PADI >14 mm, whereas a PADI <10 mm was related to a worse clinical outcome. However, neither the ADI nor the PADI can be used to

assess cord compression caused by soft tissues, such as pannus development in the retro-odontoid space. Thus, spinal cord compression can be present even in the absence of abnormalities in plain X-ray measurements.

The possibility of rotatory AAS should be considered in the presence of asymmetry or lateral displacement of the atlas on the axis by >2 mm in an open-mouth view or in cases of asymmetrical collapse of the lateral atlas mass [44]. Fracture of the dens may be another sign of lateral displacement. A CT scan is the method of choice for confirmation of the diagnosis. Diagnosis of cranial settling through X-ray evaluation can often be very difficult because osseous structures of the cranial base are superimposed upon the landmarks, particularly in the cervical spine [44]. Diagnosis of cranial settling may be further complicated by the presence of erosions of the dens. Plain X-rays generally allow evaluation of the extent of cranial settling in relation to several parameters based on different anatomical landmarks (Fig. 2). Various parameters are used to assess cranial settling with plain X-rays, including McRae's line, McGregor's line, the Ranawat index, the Redlund-Johnell value and the Clark stations. The measurements most commonly used are the Ranawat index [45] and the Redlund-Johnell value [46]. Lateral scanning allows clear visualization of the anatomical landmarks used for these measurements (Fig. 2b). An altered Ranawat index most likely matches settling at the C1–C2 level, and the Redlund-Johnell value shows occiput–C2 changes. In 1989, Clark et al. [47] described a method involving identification of three equidistant stations of the odontoid process of the axis. Usually the superior third of the odontoid is at the same level as, or close to, the anterior ring of the atlas. With mild cranial settling, the middle third of the odontoid process is level with the anterior ring of the atlas (station II). With severe cranial settling, the inferior third of the odontoid process is level with the anterior ring of the atlas (station III) [48]. In addition, these authors recommended considering as positive the results of screening for basilar impression when at least one of the following three criteria is positive: the Clark stations, the Redlund-Johnell value or the Ranawat index. Use of these criteria in combination increased the sensitivity to 94% and the negative predictive value to 91%. In the presence of a positive result on at least one of the three criteria, a more detailed study with CT scanning or MRI is mandatory. However, MRI has replaced CT scanning as the evaluation of choice in clinical evaluation because of its greater sensitivity in identifying inflammatory alterations in the joints and synovial changes, especially at the early stages of the disease. MRI allows better visualization of soft tissue, neural structures (the spinal cord and nerve roots) and the contents of the epidural space, providing further information for assessment of spinal cord compression [49].

Through triplanar images, MRI permits clear visualization of facet subluxations, joint damage and dens dislocation [50], giving precise information about CVJ relationships.

MRI is also suggested for assessment of the cervicomedullary angle: in two studies, patients with an angle <135° had a diagnosis of cranial settling and myelopathy [51, 52]. In addition, MRI alterations can be useful for prognosis: T1-weighted spinal cord signal changes are associated with a poor clinical status and also a poor final postoperative outcome [52]. MRI is also useful to evaluate pannus regression after surgical treatment.

Since pannus formation is probably related to articular hypermobility, fusion of the affected joint can lead to pannus regression (especially in cases imaged with contrast enhancement) [53]. Dynamic MRI has been used with the patient in flexed or extended positions and in the traditional neutral position. Roca et al. recommend performing functional MRI in a flexed position in patients with RA with suspected cervical subluxation when routine MRI findings in the neutral position are normal [54]. Other authors have suggested performing functional MRI as a preoperative examination [55].

Management

The goals of RA treatment are symptom relief and a reduction in the disease activity rate. Good control of disease activity is considered to be the primary non-surgical treatment of cervical spine involvement in RA. However, prolonged high-dosage use of corticosteroids represents an independent risk factor for cervical subluxation in RA [56–59]. Moreover, an elevated incidence of SAS has also been reported in patients not affected by RA, and it is directly associated with the duration of corticosteroid exposure.

In a clinical series of 67 patients it was demonstrated that treatment with disease-modifying antirheumatic drugs (DMARDs) was correlated with a greatly decreased incidence of cervical spine involvement: reductions of 9% in the rate of atlantoaxial subluxation, 4% in the rate of basilar invagination and 2% in the rate of subaxial subluxation were reported [60]. Combination therapy with different DMARDs might further reduce the rates of cervical spine involvement. No cases of atlantoaxial subluxation or basilar invagination were observed in a study of 195 patients with early RA treated with more DMARDs during a follow-up period of 2 years, and only 3.5% of these patients were reported to have subaxial subluxation [61]. These findings suggest that biological DMARDs might be more useful for preventing de novo cervical spine involvement than for slowing the progression of pre-existing pathology [62].

Conservative treatment is the preferred therapeutic strategy in RA patients with cervical spine involvement but without neurological deficits. When neurological symptoms and signs are present, surgical treatment should be considered, evaluating different factors such as age, the severity of

the disease and the patient's general condition. Properly administered anaesthesia is crucial, and surgical treatment should be performed at centres with expertise in treating CVJ abnormalities. Nevertheless, surgical management of RA is a stimulating field. There clearly has been a reduction in cases of mutilating RA involving the CVJ, thanks to steady improvement in surgical procedures.

A successful surgical procedure requires a great deal of expertise to achieve stable decompression of the CVJ. Therefore, a skilled preoperative evaluation, an appropriate choice of surgical procedure, and use of suitable haemostatic and sealant devices and stabilization instruments are required to achieve the best functional result and avoid surgical complications [63, 64].

Enlarged transoral approaches—despite being associated with greater morbidity—are helpful when severe basilar invagination, cranial extension of the lesion or limited jaw mobility are present.

The introduction of endoscopy in transoral surgery and the endoscopic transnasal approach have strongly improved the outcome of RA patients with rapidly progressive myelopathic symptoms. For those patients with chronic symptoms, CVJ fixation procedures seem to be more suitable both for stabilization and for secondary progressive pannus reabsorption [62, 65–68].

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Chiari

Chiari Malformations

Cristina Mancarella, Roberto Delfini, and Alessandro Landi

Abstract *Background:* Chiari malformations (CM) represent a group of anomalies characterized by descent of the cerebellar tonsils or vermis into the cervical spinal canal. These malformations can be associated with abnormalities such as hydrocephalus, spina bifida, hydromyelia, syringomyelia, curvature of the spine (kyphosis and scoliosis) and tethered cord syndrome. Hereditary syndromes and other disorders that affect growth and bone formation—such as craniosynostosis, Ehlers–Danlos syndromes and Klippel–Feil syndrome—can also be associated with CM.

Methods: The literature concerning treatment is large, and an extensive range of therapeutic protocols have been described. The literature is inclined in favour of surgery; however, there is controversy over when to perform surgery and which procedure is most appropriate. Lately, the indications for stabilization have been under discussion.

Results and Conclusion: In this paper we review the literature and discuss the historical background, anatomical forms, pathophysiology, clinical presentation, relationships with other diseases and diagnostic procedures for these abnormalities.

Keywords Chiari malformation · Craniocervical junction Syringomyelia

Introduction

Chiari malformations (CM) comprises various pathologies that have in common anatomical deformities of the brainstem and cerebellum. In the early 1890s, Professor Hans Chiari (1851–1916), an anatomopathologist at the German University in Prague, Czechoslovakia, described the congenital anomalies that would later be named Chiari malformation types I–IV [1–3]. These anomalies are characterized by downward elongation or displacement of the cerebellar tonsils or the vermis into the cervical spinal canal. Hydrocephalus is sometimes present. Other abnormalities can be associated with CM, such as syringomyelia, spina bifida, hydromyelia, kyphosis, scoliosis and tethered cord syndrome. Moreover, CM may be associated with hereditary syndromes and other disorders that affect growth and bone formation, such as craniosynostosis, Ehlers–Danlos syndromes and Klippel–Feil syndrome.

The literature concerning treatment is large, and many therapeutic protocols have been described. The literature is inclined in favour of surgery. Moreover, there is controversy over when to perform surgery and which procedure is most appropriate. Lately, the indications for stabilization have been under discussion.

In this paper, we review the literature and discuss the historical background, anatomical forms, pathophysiology, clinical presentation, relationships to other diseases and diagnostic procedures for these abnormalities.

History

In 1883, Cleland was the first author to define what later became known as Chiari malformation type II or Arnold–Chiari malformation, with his report on a single infant with myelomeningocele and hindbrain abnormalities [4]. In 1891, Chiari published his first manuscript in which he collated and analysed data from over 40 postmortem inspections of hindbrain malformations [2] and described CM types I, II, and III [2, 5]. In 1894, Arnold described a case of a child with elongation and descent of the inferior part of the cerebellum into the spinal canal and spina bifida [6]. Later, the type IV malformation was described in Chiari's 1895 publication [3], which stated that "other factors must play a role in this condition"—namely, insufficient skull growth, causing

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increased intracranial pressure (ICP)—in contrast to his previous report, in which he postulated that CM was caused by hydrocephalus.

Anatomical Forms of Chiari Malformation

The CM classification divides the abnormalities according to the severity of cerebellar tonsil descent and other neurological anomalies.

Chiari Malformation Type I

This is the least severe type and usually presents in adulthood. Hydrocephalus is infrequent. Syringomyelia is often found. Neuroimaging shows cerebellar tonsil elongation into the upper cervical canal through the foramen magnum. In general, tonsils lying 5 mm or more below the foramen magnum on neuroimaging are considered consistent with CM, though there is no direct correlation between clinical severity and how low the tonsils are lying. Usually, the origin is mesodermal.

Chiari Malformation Type II

This is the most frequent form. It is also known as 'classic' or 'Arnold–Chiari' malformation. Hydrocephalus is present in more than 70% of cases. The cerebellar hemispheres and the inferior vermis extend into the foramen magnum with displacement of the brainstem (the fourth ventricle, lower portion of the pons and medulla) inside the spinal canal and elongation of the aqueduct and fourth ventricle. It is associated with spina bifida and other cerebral, spinal and meningeal abnormalities. Its origin is a neuroectodermal disturbance.

Chiari Malformation Type III

This is a rare form. Hydrocephalus is present in 50% of cases and is of the obstructive type because of an associated Dandy–Walker malformation or aqueductal stenosis. It is characterized by caudal displacement of the medulla and herniation of part of the cerebellum in an occipital, cervical or occipital–cervical meningocele. Sometimes part of the hindbrain is also herniated. It is a disorder of neuroectodermal origin.

Chiari Malformation Type IV

This is the least frequent but most severe form of CM, characterized by an incomplete or undeveloped cerebellum (cerebellar hypoplasia or aplasia) and alterations of the pons with a 'pigeon breast' deformity of the brainstem. These anatomical alterations cause evident dilatation of the fourth ventricle, cisterna magna and basal cisterns, although hydrocephalus is infrequent. It is a neuroectodermal malformation.

Other Forms of Chiari Malformation

CM type 0 is characterized by the presence of syringomyelia without detectable descent of the cerebellar tonsils [7]. CM type 1.5 is characterized by tonsillar herniation and caudal migration of the brainstem and fourth ventricle, but with absence of spina bifida.

Pathophysiology

Many theories have been proposed to explain the causes of CM, but still none of the causes is completely clear. Here, we discuss the four main theories that have been offered in an attempt to explain the different hindbrain pathology seen in CM, although no single error in development appears to produce the anomaly.

- Hans Chiari, in his initial theory, ascribed hindbrain herniation to hydrocephalus. The theory stated that the ICP due to hydrocephalus and the consequent cerebrospinal fluid (CSF) pressure differential in the cranial and spinal compartments may cause cerebellar vermian herniation. However, this model fails to explain several features of CM. Between 10% and 20% of children with myelomeningocele and CM never develop hydrocephalus. Moreover, from prenatal imaging it is known that CM is often present prior to radiographic appearance of hydrocephalus. In addition, the small posterior fossa, low-lying torcular herophili and upward 'herniation' of the vermis are not explained by this theory [3].
- Cleland postulated that a primary dysgenesis of the hindbrain may occur, characterized by central nervous system (CNS) malformations in the posterior fossa. This theory fails to account for the cranial and supratentorial anomalies frequently associated with CM and myelomeningocele [4].
- Another theory postulated that a mesodermal disorder may result in a low-volume posterior fossa and subsequent

overcrowding; this may explain the vermian and brainstem herniation. A similar concept involves an 'induced' small posterior fossa secondary to chronic CSF leakage from an open spinal defect. Again, these theories fail to explain the widespread CNS abnormalities seen in patients with myelomeningocele.

- Traction theorists, including Penfield, postulated that development of CM occurred by the open and tethered spinal cord pulling the brainstem and cerebellum inferiorly, resulting in elongation and herniation of the posterior fossa structures. This theory does not explain many CNS deformities. Another flaw in this theory is the fact that traction forces applied to the lumbar spinal cord are essentially non-existent beyond four spinal levels from the tethered site [8].

McLone and Knepper combined these theories into a 'unified theory' of CM [9]. In this concept, the loss of CSF troughs via both an open neural tube defect and incomplete spinal occlusion causes a subsequent drop in ICP. CSF drains through the central canal and is therefore not retained in the ventricular system. The absence of the ventricular CSF driving force during fetal development results in poor cranial vault expansion, resulting in a small posterior fossa. The unexpectedly narrowed posterior fossa leads to caudal displacement of the brainstem and cerebellum through the foramen magnum. This theory also explains the development of hydrocephalus due to overcrowding of the posterior; with CSF outflow blocked or impaired at the foramina of Luschka and Magendie, progressive ventriculomegaly ensues.

Epidemiology and Clinical Presentation

The prevalence of CM has been estimated to be between 0.1% and 0.5%, but it is possible that higher rates will be identified by increasing use of magnetic resonance imaging (MRI). Most clinical cohort studies show an equal prevalence in both sexes or a slight female predominance. There is no particular ethnic or geographical distribution, and there are no known risk factors other than family history.

The majority of cases of CM are asymptomatic. Most patients with CM type I are diagnosed in late childhood to early adulthood.

Symptoms related to CM vary. The most common symptoms in these patients are headaches and neck pain, frequently associated with dysaesthesia in the C2 dermatome. The pain is exacerbated by coughing or sneezing. Given that CM type II is often associated with myelomeningocele, the first sign is an open neural tube defect. Approximately 33% of patients with CM type II will develop signs and symptoms of brainstem herniation/compression prior to the age of 5 years, and more than one third of patients with such early manifestations will not survive.

The symptoms of CM include alterations in the pattern of breathing (including periods of apnoea, stridor and dysphagia resulting in aspiration); a depressed gag reflex; involuntary, rapid, downward eye movements; and loss of arm strength.

Whatever the age of the child, it is important to evaluate hydrocephalus if it is present. Untreated hydrocephalus or a malfunctioning shunt can worsen CM and make it symptomatic by increasing the ICP with subsequent downward herniation of an already caudally displaced brainstem and vermis.

In infants, other signs and symptoms of CM include paraparesis (more commonly in the upper extremities) or quadriparesis, hypotonia, opisthotonos, nystagmus, a weak cry and developmental delay.

In older children, symptoms are insidious, slowly progressive and far less frequently life threatening. An occipital headache and/or craniocervical pain with signs and symptoms of cervical myelopathy may be present, with upper-extremity weakness and spasticity being the most common findings. Changes in handwriting skills, agility and self-care may be the first outwardly noticeable signs. Ataxia of both the upper extremities and the trunk is also common. Syringomyelia is associated with CM type II (as in CM type I) and should be looked for in a symptomatic child. Suspended disassociated sensory loss, hand atrophy, scoliosis, back pain and lower motor neuron findings should prompt radiological evaluation of the spine, specifically looking for syringomyelia.

Relationships with Other Disease States and Syndromes

Hydrocephalus

Hydrocephalus is noted in approximately 4–18% of patients with CM type I. It is due to obstruction of the aqueduct or the outflow foramina of the fourth ventricle. These patients all require CSF diversion in addition to surgical posterior fossa decompression. Endoscopic fenestration (ventriculostomy) of the floor of the third ventricle establishes an alternative route for CSF toward the subarachnoid space.

Craniosynostosis

The association between craniosynostosis and CM type I was first documented by [10]. In 1% of cases the abnormal skull base that causes a decreased posterior fossa volume and subsequent tonsillar herniation is due to early fusion of the lambdoid suture. Synostosis can be present alone or as part of a syndrome; it has been reported to occur in 72.7% of patients with Crouzon syndrome, 1.9% of those with Apert syndrome, 50% of those with Pfeiffer syndrome and 100% of those with Kleeblattschädel syndrome. Moreover, CM type I can also be associated with Pfeiffer syndrome type II, Jackson–Weiss syndrome, Seckel syndrome, Antley–Bixler syndrome, Shprintzen–Goldberg syndrome and other rare pathologies.

Endocrinopathy

Growth hormone deficiency is associated with CM type I in 5–20% cases. The tonsillar herniation is due to insufficient development of the posterior fossa.

Acromegaly has also been implicated as an endocrine-related disorder causing CM type I. In these cases an excessive amount of growth hormone is believed to thicken the bones of the posterior fossa, resulting in CM type I.

Achondroplasia has been observed in patients with CM type I, who have a small, shallow posterior cranial fossa.

Hyperostosis

When hyperostosis affects the posterior fossa, it can often lead to CM type I. Paget's disease of the skull is one example in which excessive bone turnover leads to thickening and deformation of bones.

Cases of CM type I relating to craniometaphyseal dysplasia are also extremely rare. CM type I secondary to osteopetrosis and erythroid hyperplasia has been documented but is considered to be exceptionally rare.

Bone Mineral Deficiency

Patients with familial vitamin D–resistant rickets have a higher incidence of CM type I. This is probably caused by bone overgrowth and calvarial thickening because of low serum phosphate levels, with subsequent overcrowding of the posterior fossa.

Cutaneous Disorders

Cutaneous disorders are frequently reported to occur in conjunction with CM type I. Neurofibromatosis type I has been related to CM in 8% of cases; this association is possibly due to a mesodermal deficiency. Several other cutaneous disorders have been postulated to be associated with CM type I, such as macrocephaly–cutis marmorata telangiectatica congenita, blue rubber bleb nevus syndrome, phacomatosis pigmentovascularis type II, LEOPARD [lentigines, electrocardiographic conduction defects, ocular hypertelorism, pulmonary stenosis, abnormalities of the genitals, retarded growth and deafness] syndrome, acanthosis nigricans, giant congenital melanocytic nevi and Waardenburg syndrome variants.

Spinal Defects

Scoliosis is often associated with CM. A few disorders—such as spondyloepiphyseal dysplasia, basilar impression, caudal regression syndrome, Klippel–Feil syndrome, atlantoaxial assimilation and odontoid retroflexion—are also associated with CM type I. Little is known about the pathophysiology of these spinal deformities, but it is believed that difficulty in equilibrating the dynamic CSF pulse pressure induced by the Valsalva manoeuvre is responsible for CM type I presentation.

Lipomeningomyelocele has also been shown to be coupled with CM type I in as many as 3–6% of patients.

Space-Occupying Lesions

This category includes both space-occupying lesions (ranging from brain tumours to haematomas) and CSF leaks.

Rare Cases

Rare cases of Beckwith–Wiedemann syndrome, Costello syndrome and Marfan syndrome have been reportedly associated with CM type I. Additionally, associations with Williams–Beuren syndrome have been found, with morphometric analyses suggesting a diminished posterior fossa leading to CM type I. Finally, associations with disorders such as cystic fibrosis, situs inversus, Pierre Robin syndrome, Fabry disease, Ehlers–Danlos syndromes, Kabuki syndrome, CHERI [CM type I with or without a cleft palate, deviant

electrocardiogram or epilepsy, and retarded intelligence with delayed language development] syndrome and cloacal exstrophy have been reported, with no clear pathophysiological mechanism being identified yet.

Preoperative Evaluation

A complete neurological examination is indispensable in the preoperative evaluation. The lower cranial nerves have to be examined because dysfunction of these nerves could possibly necessitate postoperative tracheostomy or gastrostomy. Cardiac and pulmonary function should also be assessed preoperatively.

Thanks to the advent of MRI, which has become the gold standard for diagnosis of syringomyelia and CM, these pathologies are now diagnosed more often. In 1985, Aboulezz et al. used MRI to establish that the tonsil tips, in normal conditions, can extend up to 3 mm below the foramen magnum; in patients affected by CM their extension can exceed 5 mm[11]. Barkovich et al. reported that the limit in normal cases was 5 mm below the foramen magnum; in Chiari cases they described the 'peg-like' aspect of the tonsils and narrowing or complete effacement of the subarachnoid space at the foramen magnum. The cerebellar vermis can occasionally be very low, entering the upper thoracic canal. An important role in our understanding of the pathophysiology of CM type I is played by cine-flow MRI, allowing us to study the dynamics of CSF flow at the posterior fossa and foramen magnum. Moreover, because of these advanced technologies, it has been possible to demonstrate that myelomeningocele is present in all children with CM type II [12].

Another characteristic sign is the ultrasonographic 'banana sign', which is due to the abnormal shape of the cerebellum together with an absent cisterna magna. This sign is well identified on prenatal ultrasonography. Despite the small size of the cerebellum, the posterior fossa appears 'crowded' because of the abnormally small size of the compartment. The tentorium, which is typically hypoplastic, is abnormally low, often placing the torcular herophili just above the foramen magnum—a critical point to keep in mind when a suboccipital craniectomy is being considered or performed.

Approximately two thirds of patients will show a medullary 'kink' dorsal to the upper cervical spinal cord. The presence of a medullary kink has been associated with a more symptomatic clinical course in 75% of patients with this finding below C4. Depending on the presence and severity of this sign, the lower cranial nerves exiting from the medulla can travel in a cephalad direction and the tracts of the spinal cord can double back on themselves for a short distance.

Multiple ventricular anomalies are commonly found in the patient with CM type II. The fourth ventricle, which is typically small and poorly visualized, is frequently displaced into the cervical canal. The aqueduct is similarly small and rarely seen on routine imaging but probably does not contribute significantly to hydrocephalus. The third ventricle is termed the 'shark tooth' deformity because it may appear in a narrow-angled form. The lateral ventricular appearance varies from nearly normal to severely deformed and hydrocephalic. 'Beaking' of the frontal horns is sometimes seen when the frontal horns point inferiorly. This finding is attributed to interdigitations of the cerebral hemispheres in the region.

Surgery

History

In 1932, Van Houweninge Graftdijk was the first person to propose surgical treatment of CM [13]. In a monograph, he described suboccipital decompression and resection of the tonsils. The aim of this procedure was to relieve CSF flow obstruction at the foramen magnum. All of the patients described in this report died as a direct consequence of the surgical procedure or afterward because of postoperative complications. In 1934, Ebenius described a case of basilar impression treated with suboccipital decompression [14]. In 1938, McConnell and Parker reported that five adult patients died after surgical treatment of hydrocephalus. Autopsies showed tonsil descent through the foramen magnum in three of them [15]. In the same year, Aring [16] described the case of a 20-year-old boy who died after surgical treatment for increased ICP and in whom posterior fossa exploration showed tonsil descent through the foramen magnum. In 1940, Gustafson and Oldberg described two patients with an intraoperative diagnosis of CM [17]. At autopsy, one of them was also found to have basilar impression and syringomyelia. With the aim of ensuring order in the nomenclature, Russell suggested in 1949 that the 'Arnold–Chiari' term should be used only for patients affected by spina bifida [18]. Subsequently, Baker [19] described 11 cases as Arnold–Chiari malformations with tonsil descent to the C1 level, and remarked that the normal level of the cerebellar tonsils is when the tips are above a line between the clivus tip and the posterior rim of the foramen magnum on a lateral X-ray. Three cases underwent surgery; the rest of them were described as "mild forms of Arnold–Chiari malformation"

and did not require surgery. The confusion regarding the nomenclature persisted until not long ago; even in 1971 it was suggested that the designation 'Chiari malformation' should be reserved for infants with associated spina bifida and hydrocephalus.

The Midline Posterior Approach

It is possible to use the sitting, ventral or lateral position. A midline incision is performed from the occipital protuberance down to the upper cervical region. The posterior muscles are opened through the midline avascular plane, up to the occipital protuberance and down to the spinous process of C2. The drill and Kerrison rongeurs are used to open the bone. The opening is always limited to the lower part of the occipital bone and the posterior arch of the atlas. Some surgeons perform a Doppler ultrasound study to determine whether to open the dura or not. Occasionally, removal of the bone alone may re-establish normal CSF flow. The dura is then opened in a T- or Y-shaped fashion and retracted with stitches. The tonsils and cisterna magna are now visible to the surgeon. Depending on the extent of the tonsil herniation, the damaged parts may be shrunk with electrocautery. This shrinkage guarantees elimination of the blockage of CSF flow out of the fourth ventricle. To enlarge the dura opening and the space around the tonsils, a patch is necessary. The patch can be made of synthetic material or part of the patient's pericranium (deep scalp tissue from just outside the skull). The dural patch is sealed in a watertight fashion. The suture line is topped with a dural sealant to reduce the risk of CSF leakage [20].

New Prospects

The real nature and origins of CM have still not been entirely elucidated. Lately, new proposals have been suggested for treatment of this malformation. One of the most important and interesting theories was suggested by Goel in 2015 [21]. He proposed a new 'philosophy' in which CM (with or without concomitant basilar invagination) is considered secondary to atlantoaxial instability. He observed that the previously suggested concept that a small posterior cranial fossa volume causes tonsillar herniation no longer seems to be valid. He noted that the cerebellum is more frequently atrophied in such cases; consequently, decompression cannot be a solution to the problem. Goel proposed that CM and syringomyelia are secondary results of subtle and long-standing atlantoaxial instability. The atlantoaxial facet joint is one of the most mobile joints in the body; it is the centre of mobility and also the centre of instability of the atlantoaxial region. His hypothesis was that the tonsils in CM position themselves in a strategic location. He defined them as "nature's airbag", which protects the cord from getting pinched between the bones in the event of instability. Similar beliefs appear to be valid for syringomyelia. The cause of instability in the atlantoaxial region is the joint. Such instability begins in the joint and then manifests in the rest of the constituent bones of the region. This is why the Goel proposed performing C1–C2 posterior instrumented fusion alone, without concomitant posterior fossa decompression. In patients treated only with atlantoaxial fixation, the tonsils migrate back to their original position and the syrinx cavity collapses following the surgical procedure. The Goel believes that screw fixation should not include occipital bone, since the instability is not at the occipitoaxial joint but at the atlantoaxial joint. Fixating a joint that is not unstable damages the biomechanical strength of the construct and limits neck movements. Moreover, opening the atlantoaxial joint and filling bone graft within its margins not only offers stability to the construct but also enlarges the bone surface for bone fusion and fixes the region in itself. This new point of view will require further studies in order to be accepted and validated [21].

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The Role of Arachnoid Veils in Chiari Malformation Associated with Syringomyelia

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Introduction

Chiari malformation type I (CM-I), or hindbrain herniation syndrome, has traditionally been defined as a dislocation of the cerebellar tonsils 5 mm or more below the foramen magnum on sagittal magnetic resonance imaging (MRI) [1, 2]. An association of this anomaly with syringomyelia is observed in 45–68% of patients [3, 4].

CM-I is a disorder of the para-axial mesoderm, which leads to posterior cranial fossa (PCF) hypoplasia and consequent overcrowding of the hindbrain [5, 6]. However, CM-I can also represent a condition secondary to disorders unrelated to PCF hypoplasia, such as hydrocephalus [7], intracranial tumours [8, 9], cerebrospinal fluid (CSF) leaks [10], genetic disorders of connective tissue associated with craniovertebral junction (CVJ) instability and cranial settling [11], tethered cord syndrome [5], craniosynostosis [12], acromegaly [13] and Paget's disease.

All of these possible situations that lead to abnormal CSF dynamics can define a clinical picture characterized by occipital Valsalva-type headaches, lower cranial nerve abnormalities and myelopathy [2, 4, 14].

Posterior fossa decompression has proven to be effective in symptomatic CM-I with syringomyelia, except in the presence of ventral compression, where anterior craniovertebral decompression is mandatory [15].

Several authors who have performed exploration of the fourth ventricle in addition to duraplasty after osseous decompression have noticed intradural membranes described as veils, pouches or webs at the fourth ventricular outlet of the foramen of Magendie, interfering with CSF flow through it [16–22].

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The presence of a fourth ventricular outlet obstruction and consequent craniospinal pressure differentials have been inculpated in the genesis of syringomyelia since the hydrodynamic theory was first devised by Gardner [23]. However, the exact relation between arachnoid veils and syringomyelia is not yet completely understood.

Patients

We studied five consecutive patients harbouring CM-1 malformation without syringomyelia in which intradural exploration showed the presence of arachnoid veils.

Headache was the most relevant symptom in all cases. Two patients had dysphagia, causing aspiration pneumonia in one case, and one patient complained of tinnitus. One patient presented with acute intracranial hypertension and was comatose at admission. In this case the patient, an obese 15-year-old, dramatically improved after surgery and had a good neurological status at 3-year follow-up (Fig. 1).

Discussion

CM-I is a common cause of cervical syringomyelia, which has an incidence of around 8.4 cases per 100,000 people and a mean age of presentation of around 30 years [24]. Nevertheless, the aetiopathogenesis of syringomyelia in the context of CM still represents an enigma. The association of syringomyelia with descent of the cerebellar tonsils, PCF overcrowding and altered CSF dynamics has led to a belief that a CSF obstruction occurs at the foramen magnum or above it [15]. This craniospinal CSF dissociation also depends on obstruction of the fourth ventricular outlet. Anomalies of the median fourth ventricular outlet—the foramen of Magendie—in patients with CM-I and syringomyelia are reported rarely. The arachnoid veils at the

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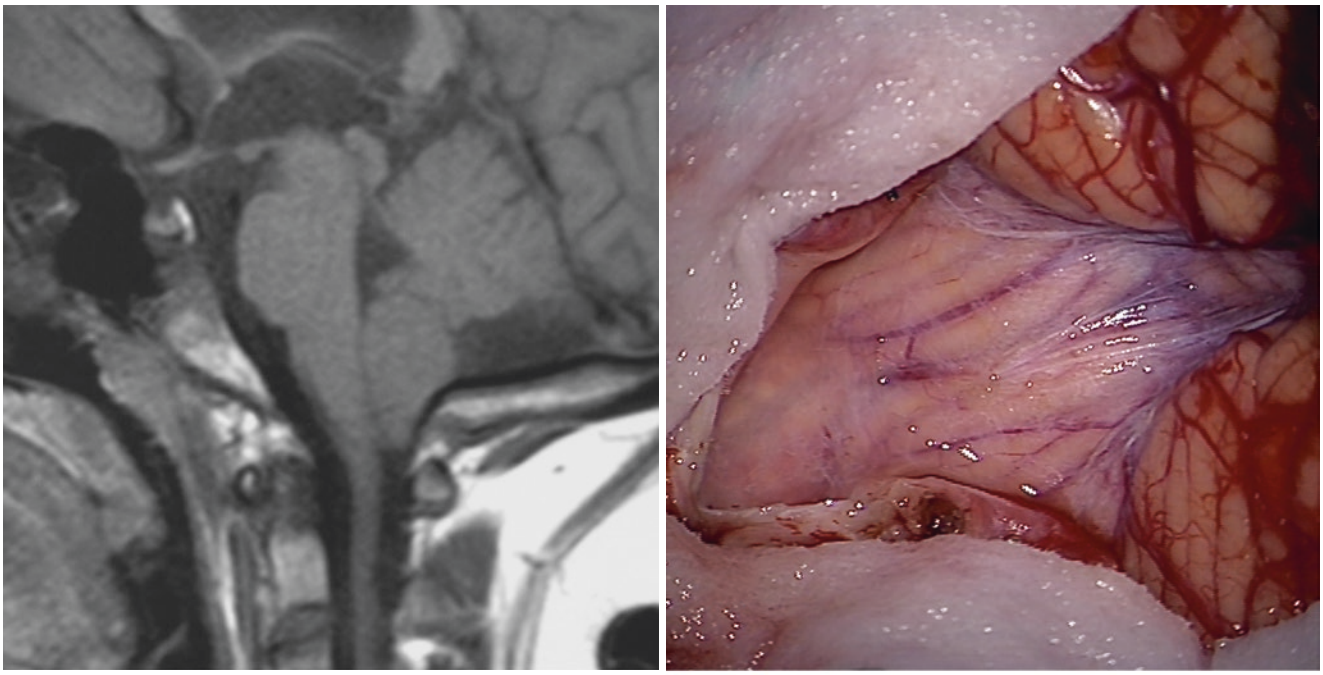

Fig. 1 *Left:* Preoperative magnetic resonance imaging (MRI) scan showing Chiari malformation (CM). *Right:* Intraoperative picture showing arachnoid veils

foramen of Magendie are considered remnants of the rhombic lip [25]—two areas of focal thickening forming along the lateral edges of the roof plate during the fourth to sixth weeks of gestation, which differentiate into the cerebellar hemispheres as they enlarge and approximate each other on the midline [26].

In eight of 30 patients with CM-I who underwent surgical exploration, Saez et al. [20] noticed that the obex appeared 'veiled over' by a thin membrane which, when pierced, allowed an outflow of CSF. Stovner and Rink [21] reported the presence of 'membranes' in patients with CM-I that might have led to CSF flow obstruction at the outflow foramina from the fourth ventricle to the basal cisterns. Tubbs et al. [25] reported that arachnoid veils caused obstruction of the foramen of Magendie in 12.5% of patients they operated on for CM-I and syringomyelia; once the lesion was punctured, the CSF drained freely from this median aperture. In all patients with arachnoid veils and syringomyelia, radiological resolution of syringomyelia was evident on follow-up studies. The authors described these structures as distinct from the arachnoid scarring that might have occurred around the caudally displaced cerebellar tonsils, because the veils in their series were always midline and wall-like. Menezes et al. [15] noticed a veil over the foramen of Magendie in 11% of patients in their CM-I series. In 1988, Bidzinski [17] published a report on 63 cases of PCF decompression in patients with syringomyelia. He found partial occlusion of the foramen of Magendie and CM-I in 16 patients, atresia of the foramen of Magendie and CM-I in nine and atresia of the foramen of Magendie alone in one.

Realistically, arachnoid veils are a frequent condition. In 1987, Rifkinson-Mann et al. [27] supposed that 6% of foramina of Magendie may be partially or completely obstructed.

Probably the presence of arachnoid veils over the foramen of Magendie is not a sufficient condition per se to determine syringomyelia in otherwise normal patients. On the other hand, Barr [16] noted that 20% of the normal population may have congenital imperforate foramina of Luschka, which are frequently bilaterally symmetrical. Tonsillar ectopia–induced compression obstructs the foramina of Luschka, compressing them against the foramen magnum; the simultaneous presence of arachnoid veils could lead to obstruction of the foramen of Magendie and consequently to abnormal CSF dynamics that are prodromal of syringomyelia.

However, this does not explain why, in the so-called Chiari malformation type 0 (without tonsillar ectopia), syringomyelia improves after PCF decompression [28, 29]. Moreover, patients with Dandy–Walker syndrome, with atresia of the fourth ventricular outlets, rarely present with associated syringomyelia [30].

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Atlantoaxial Fixation for Treatment of Chiari Formation and Syringomyelia with No Craniovertebral Bone Anomaly: Report of an Experience with 57 Cases

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Abstract *Aim:* In this paper we evaluate the role of atlantoaxial instability in the pathogenesis of Chiari formation type I and the role of atlantoaxial stabilization for treatment of this condition in cases with no obvious bone malformation in the region of the craniovertebral junction.

Materials, Methods and Results: During the period from January 2010 to July 2016, we identified 57 cases of Chiari formation where there was no bone malformation or evidence of craniovertebral junction instability that could be diagnosed on the basis of an abnormal increase in the atlantodental interval on dynamic imaging. Forty-eight of these patients had syringomyelia. The average duration of follow-up was 42 months. There were 30 males and 27 females in the series. The ages of the patients ranged from 4 to 57 years. The Japanese Orthopaedic Association (JOA), visual analogue scale (VAS) and Goel clinical grading systems were used to assess the patients' clinical status. Atlantoaxial instability was diagnosed on the basis of vertical mobility of the odontoid process on dynamic radiographs, facetal malalignment on imaging or direct bone handling during the surgical procedure. Surgical treatment was achieved using atlantoaxial fixation. Foramen magnum decompression or syrinx manipulation was not done. All patients had immediate postoperative and sustained clinical symptomatic recovery. A reduction in the size of the syrinx was observed in ten patients and regression of tonsillar herniation was observed in 12 of 23 cases in which postoperative magnetic resonance imaging (MRI) was possible.

Conclusion: Atlantoaxial instability is the prime factor in the genesis of Chiari formation even when there is no bone abnormality in the craniovertebral junction.

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Keywords Atlantoaxial instability · Chiari formation · Foramen magnum decompression · Syringomyelia

Introduction

In 2014 we reported our experience with 65 cases of Chiari formation type I treated using atlantoaxial fixation [1]. It was identified that Chiari formation may not be the primary pathology; rather, it may be secondary to atlantoaxial dislocation. Essentially this meant that atlantoaxial stabilization may be the most appropriate treatment for Chiari formation and that foramen magnum decompression is unnecessary. In our earlier publication, there were 46 patients who had basilar invagination and 19 patients who had no bone anomaly at the craniovertebral junction [1]. This paper reports our current experience with 57 cases of Chiari formation with no craniovertebral junction bone anomaly or instability that could be diagnosed on the basis of existing and validated radiological parameters. Apart from our report discussing this pattern of treatment for Chiari formation, there is no other such report in the literature. Considering the potential significance of these observations, our current experience is presented here.

Materials and Methods

During the period from January 2010 to July 2016, 57 cases were identified in which there was Chiari formation in the absence of any bone anomaly in the craniovertebral junction. Five of these patients had previously undergone surgical foramen magnum decompression. This series excluded cases of Chiari formation associated with basilar invagination or bone anomalies and fusions of the region, and cases related to any gross structural brain or spinal cord malformation, hydrocephalus, tumour, infection or any connective tissue disorder.

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Results

There were 30 males and 27 females in this series. The ages of the patients ranged from 4 to 57 years, the average being 34.5 years. Table 1 summarizes the clinical profile of the patients. All patients were evaluated on the basis of the Japanese Orthopaedic Association (JOA), [2] visual analogue scale (VAS) [3] and Goel clinical grading systems [1, 4] both before and after surgery. Pyramidal symptoms were present in 73% of patients. Kinaesthetic sensations were affected in 70.7% of patients and spinothalamic dysfunction was identified in 56% of patients (Table 1). Neck pain was a major presenting symptom in 29.26% of patients. Neuropathic pain in the shoulders and hands was present in 34.1% of patients. The duration of symptoms ranged from 1 month to 21 years (mean 48.48 months). All patients were investigated with dynamic computed tomography (CT) scanning (with the head in flexion, extension and neutral positions) and with magnetic resonance imaging (MRI) both before and after surgery. Vertical atlantoaxial dislocation was identified when the odontoid process moved vertically and rostrally on flexion of the head and returned to a normal position on head extension [5]. Such dislocation was identified in ten patients. Atlantoaxial facetal instability was diagnosed according to our recently described classification [6]. For this evaluation, radiological examinations were done with MRI or CT sagittal images passing through the atlantoaxial facets with the head in a neutral position. Type 1 atlantoaxial facetal instability was diagnosed when the facet of the atlas was dislocated anterior to the facet of the axis. Type 2 atlantoaxial facetal instability was diagnosed when the facet of the atlas was dislocated posterior to the facet of the axis. In type 3 atlantoaxial facetal instability, the facets of the atlas and axis were in alignment and atlantoaxial instability was diagnosed only during the operation by being observed during direct manual manipulation of the bones. Type 2 or 3 atlantoaxial instability was labelled as axial or central atlantoaxial instability, as alteration of the atlantodental interval was not a constant or primary feature (as is observed in type 1 atlantoaxial instability). In 48 patients there was syringomyelia. In nine patients there was external syringomyelia [7], meaning that an excessive amount of cerebrospinal fluid (CSF) was present in the extramedullary space. In 20 cases, both internal and external syringes were present. In 15 cases there was external syringobulbia, suggesting there was an excessive amount of CSF surrounding the brainstem and cerebellum [7].

Table 1 Presenting complaints

Presenting complaints	Number of patients
Pyramidal involvement	40
Kinesthetic involvement	41
Spinothalamic involvement	32
Neck pain	17

All patients underwent atlantoaxial fixation using the techniques described in 1994 and 2004 [8–10]. The aim of surgery in all cases was atlantoaxial stabilization leading to arthrodesis. A bone graft was harvested from the iliac crest, packed into the atlantoaxial joint cavity and placed in the midline, over the (appropriately prepared) host bone of the arch of the atlas and lamina and the spinous process of the axis. Stainless steel plates with monoaxial non-locking-type screws were used in the initial part of the series. After June 2013, the plates and screws used for fixation were made of MRI-compatible titanium material. Foramen magnum decompression was not done in any case.

Clinical improvement was observed in the immediate postoperative phase in all patients. The progress of improvement was sustained at follow-up. The follow-up period ranged from 6 to 78 months, the average being 42 months. All postoperative assessments were performed using clinical and radiological information obtained at least 6 months after surgical treatment. Dynamic plain radiography and CT scanning were used to confirm postoperative arthrodesis. In particular, fusion of the facets and the posterior elements of the atlantoaxial bone were observed. Despite the presence of artefacts related to the use of stainless steel and titanium metal implants, the CT image quality was satisfactory for demonstration of alignment and bone fusion. Fusion was considered to be successful when the implant demonstrated maintenance of fixation on dynamic radiography and bone fusion was observed in the facets. As titanium implants were used only after June 2013, it was impossible to use MRI to evaluate the long-term status of syringomyelia and Chiari formation. However, definite reductions in the dimensions of the syrinx and reductions in tonsillar herniation were observed within a period ranging from 2 days to 12 months after surgery in 12 of 23 cases in which postoperative MRI was available for evaluation (Figs. 1 and 2). No patient's clinical symptoms worsened after initial recovery. Tables 2, 3 and 4 depict the extent of clinical recovery at a minimum follow-up of 6 months. Five patients with preoperative symptoms related to lower cranial nerve weakness had symptomatic improvement following surgery. Although voice volume and quality were not assessed or evaluated during the preoperative examination, at least three patients reported improvements in their voice volume and quality at follow-up. No patient suffered delayed neurological worsening sufficient to warrant foramen magnum decompressive surgery or needed any kind of reoperation. No patient required re-exploration for failure of implant fixation.

Discussion

Chiari formation and syringomyelia are frequently associated with bone anomalies at the craniovertebral junction [11, 12]. The issue up for discussion is whether Chiari formation

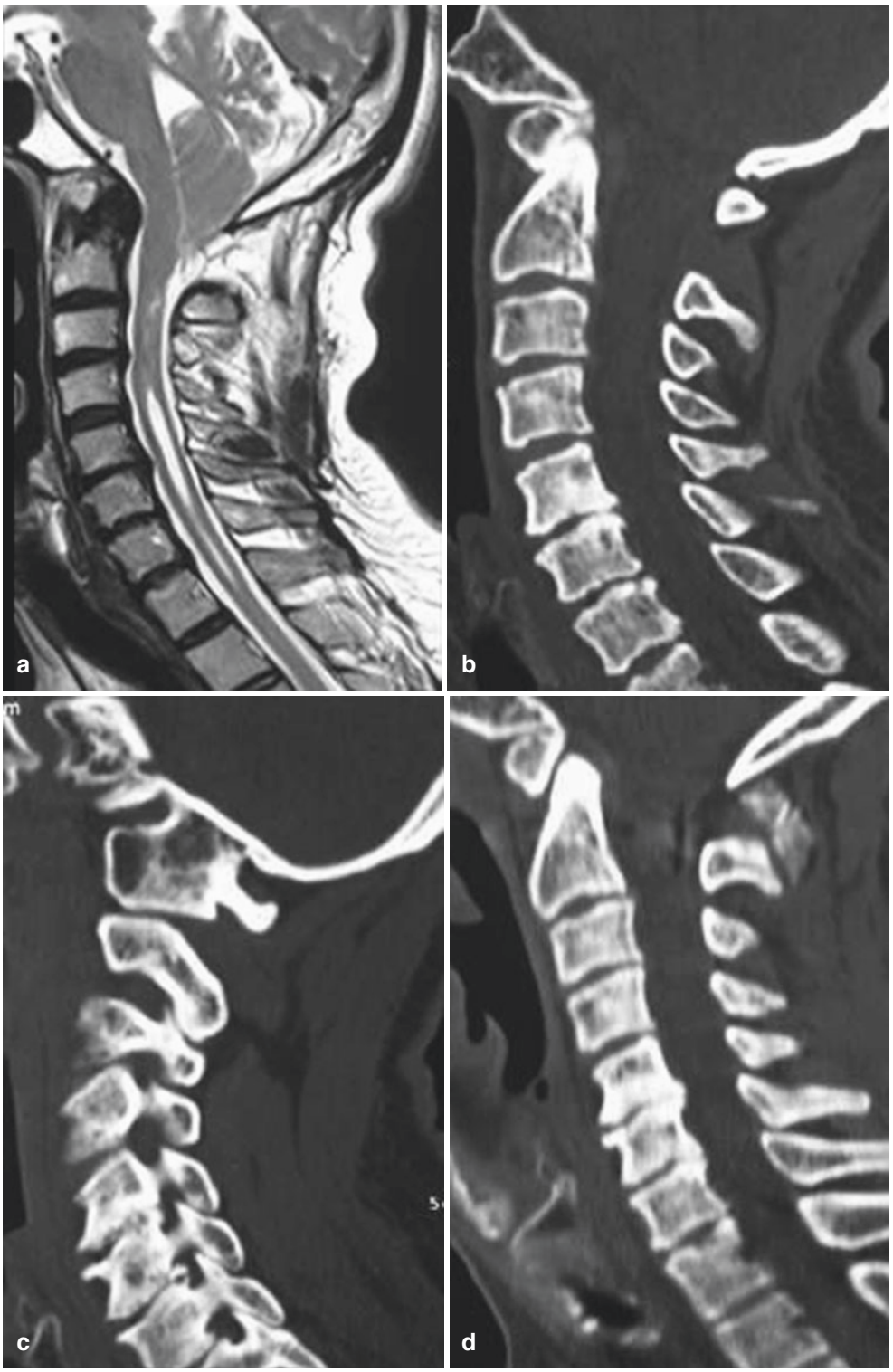


Fig. 1 Images of a 45-year-old male patient. (**a**) T2-weighted magnetic resonance imaging (MRI) showing Chiari formation and syringomyelia. Note the atrophic cerebellum. (**b**) Mid-sagittal computed tomography (CT) scan showing no evidence of any alteration in the atlantodental interval. (**c**) Sagittal CT scan with the cut passing through the facets. There is type 2 atlantoaxial facetal instability. (**d**) Postoperative mid-sagittal CT scan. (**e**) Postoperative CT scan showing the implant and fixation. (**f**) Postoperative MRI showing a reduction in the size of the syrinx and superior regression of the tonsils

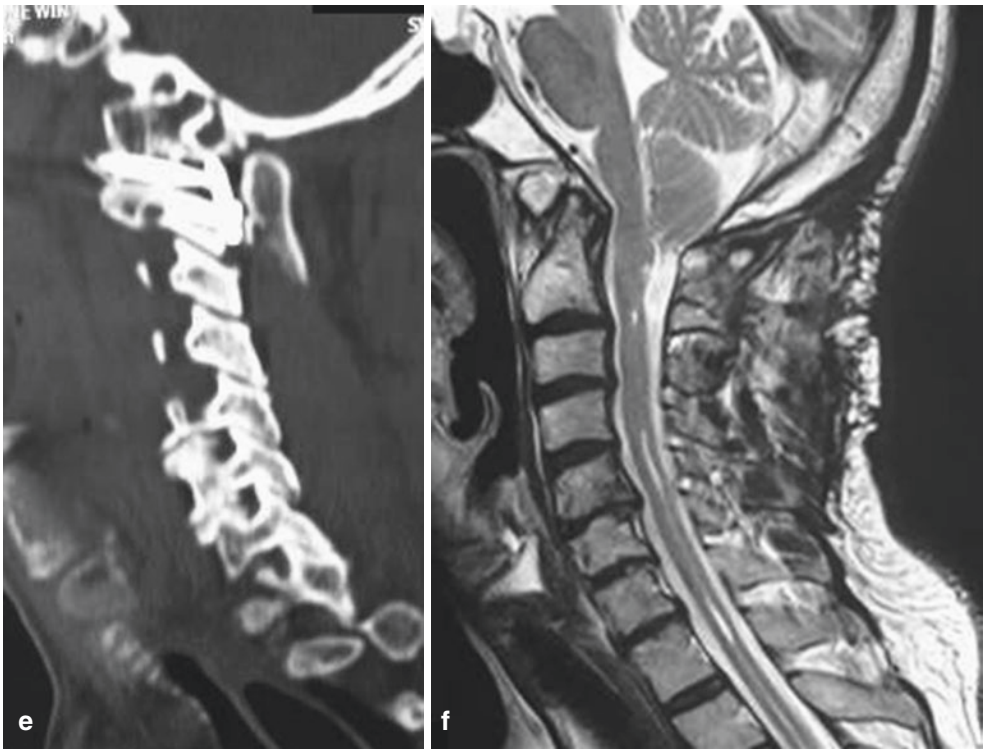


Fig. 1 (continued)

differs in its pathogenesis, and requires different surgical treatment, according to the presence or absence of associated bone anomalies at the craniovertebral junction [13, 14]. We hypothesized that Chiari formation is always secondary to atlantoaxial instability irrespective of whether bone anomalies related to basilar invagination are associated with it [1]. The current report re-emphasizes this concept.

Hans Chiari first identified Chiari formation as a 'pathological' entity in 1887 [15]. Since then there has been considerable speculation regarding its pathogenesis and treatment [16–18]. Foramen magnum decompression has been the most widely accepted method of treatment [8]. When Chiari formation is associated with syringomyelia, tonsillar resection, arachnoid dissection, plugging of the obex, introduction of a shunt into the central spinal canal through the region of the obex, syringoperitoneal/subarachnoid shunts and a host of other treatment forms have been identified and successfully deployed [19–23]. Foramen magnum decompression surgery has ranged from bone decompression to dural decompression, introduction of pericranial grafts and fascia lata grafts for dural reconstruction [19, 20, 22–24], introduction of reverse foramen magnum bone grafts [25] and metal implants, and several such alternatives. In our earlier report, it was commented that there are as many different variations of the surgical procedure for foramen magnum decompression as there are surgeons performing this procedure [1, 23]. Every surgeon is convinced that the methods of foramen magnum decompression used by him or her are optimal and produce the best results.

In 1998 it was identified that when Chiari formation was associated with basilar invagination, the posterior fossa volume was relatively small; accordingly, foramen magnum decompression was the recommended mode of treatment [21]. For the first time in the literature, it was suggested that bone decompression was sufficient and that there was no need to perform dural decompression for Chiari formation, even when there was associated syringomyelia [21]. Some authors have identified that Chiari formation associated with basilar invagination is pathogenetically different from the scenario in which there is no craniovertebral region bone anomaly [26]. Our observations in 2014 suggested that Chiari formation—with or without bone abnormalities at the craniovertebral junction—is secondary to atlantoaxial instability; accordingly, we treated this entity with atlantoaxial stabilization alone [1].

The conventional mode of identification of atlantoaxial instability has been alteration of the atlantodental interval on dynamic imaging. It was identified that atlantoaxial instability can be vertical [5], horizontal–anteroposterior or lateral [27], and central or axial in nature [6]. Identification of such forms of atlantoaxial instability has added a new dimension to our understanding of the subject. On evaluation of the dynamic images, we identified ten cases that had vertical mobile and reducible atlantoaxial dislocation [5]. In all

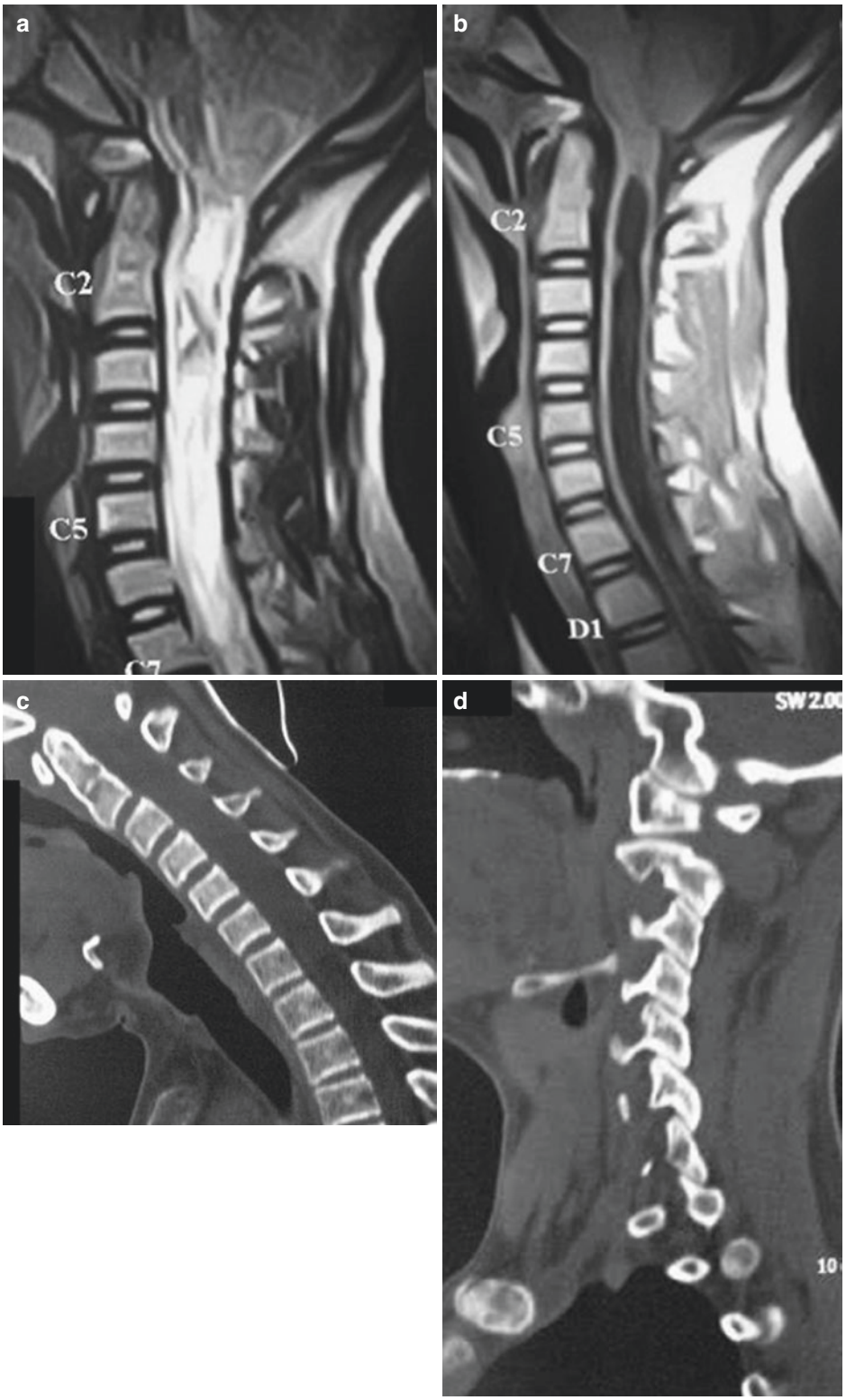
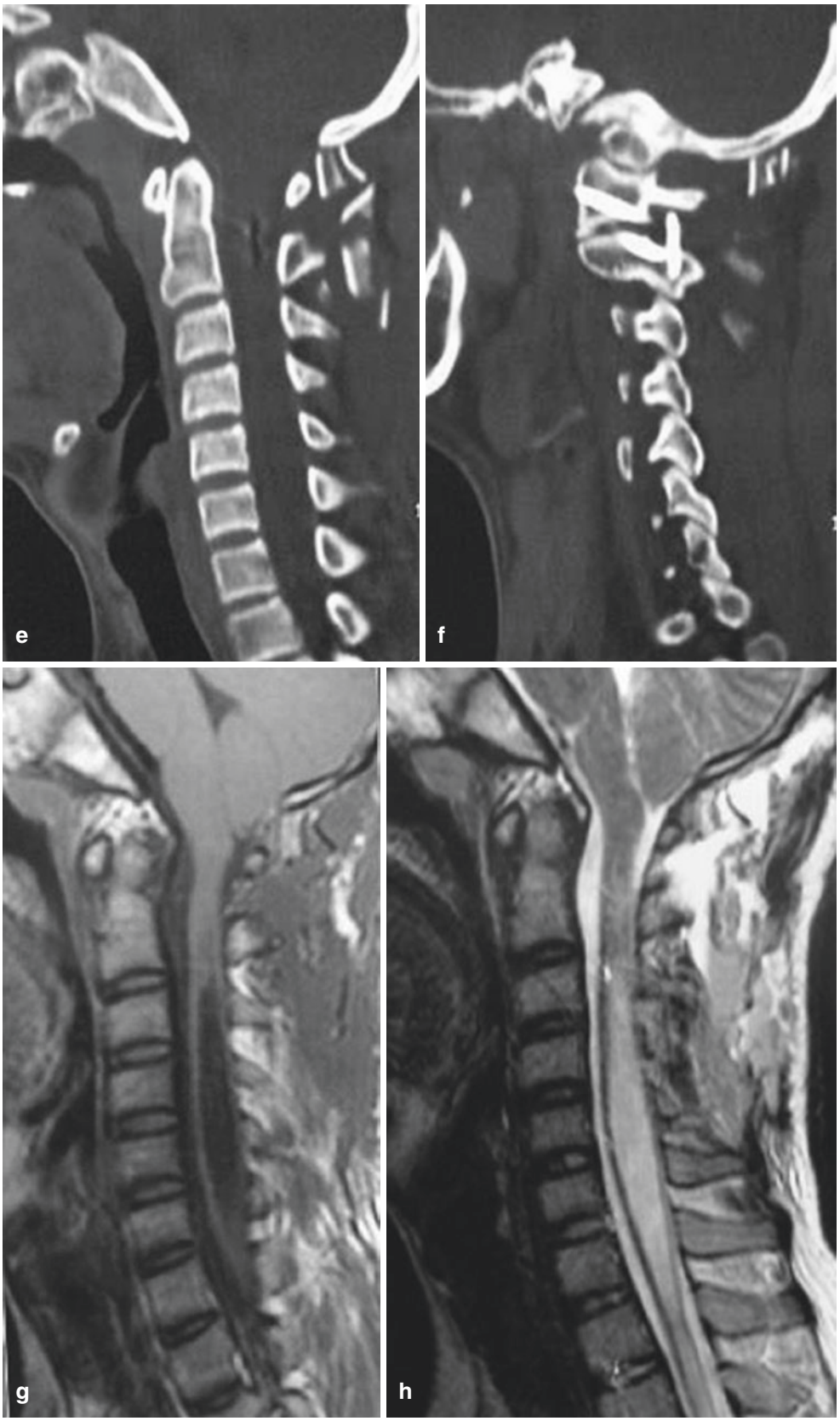


Fig. 2 Images of a 17-year-old male patient. (**a**) T2-weighted magnetic resonance imaging (MRI) showing Chiari formation and syringomyelia. (**b**) T1-weighted MRI showing Chiari formation and the syrinx. (**c**) Mid-sagittal computed tomography (CT) scan with the head in flexion, showing no evidence of any alteration in the atlantodental interval. (**d**) Sagittal CT scan with the cut passing through the facets. There is type 2 atlantoaxial facetal instability. (**e**) Postoperative mid-sagittal CT scan. (**f**) Postoperative CT scan showing the implant and fixation. (**g**) Immediate postoperative (after 18 h of surgery) T1-weighted MRI showing a reduction in the size of the syrinx. (**h**) Immediate postoperative T2-weighted MRI showing a reduction in the size of the syrinx. (**i**) Delayed postoperative T1-weighted MRI (12 months after surgery) showing a reduction in the size of the syrinx and superior regression of the tonsils. (**j**) Delayed postoperative T2-weighted MRI showing a reduction in the size of the syrinx and superior regression of the tonsils



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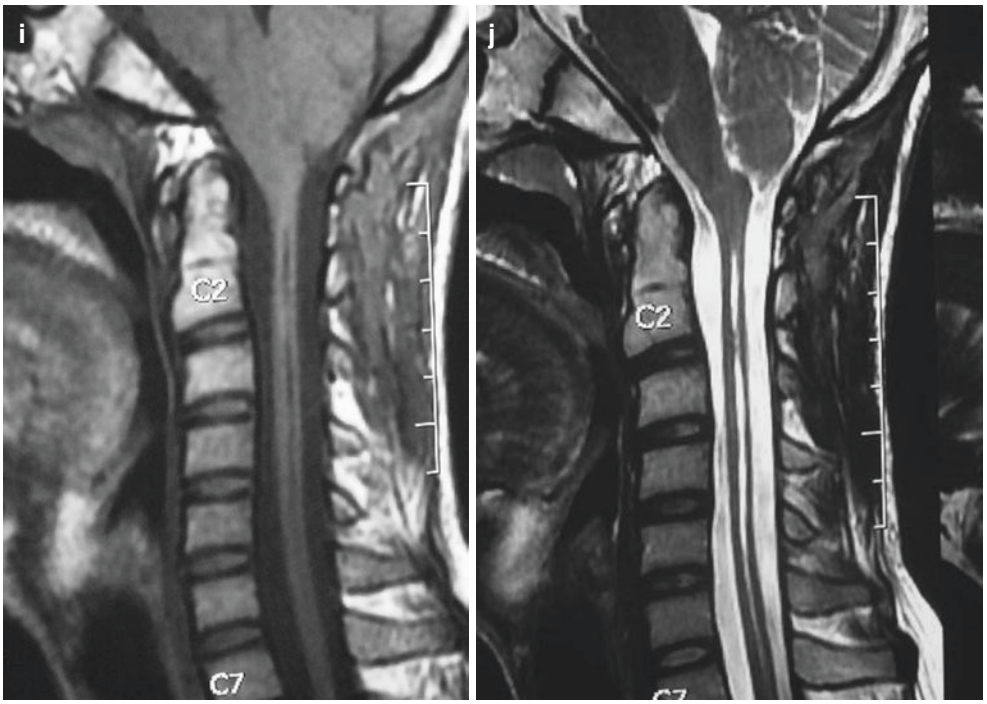

Fig. 2 (continued)

Table 2 Table showing the pre-operative and post-operative clinical assessment as per JOA scoring system

JOA score	Pre-operative (No. of patients)	Post-operative (No. of patients)
<7	9	2
8–12	12	6
13–15	21	19
16–17	15	30

Table 3 Table showing pre-operative and post-operative clinical assessment as per VAS scoring system

VAS score	Pre-operative	Post-operative
Neck pain	3–7	0–1

cases, the vertical dislocation was subtle but definite. Identification of such vertical dislocation seems to be an important additional parameter suggesting that there is instability of the region and incompetence of the atlantoaxial joints. Analysis of facetal malalignment on sagittal imaging with the head in a neutral position provides additional information about the stability of the atlantoaxial joint. Although some millimetres of facetal malalignment can be considered to be within the spectrum of normal physiological limits, the association of this malalignment with other abnormalities in the region indicates the presence of instability. Apart from basilar invagination and Chiari formation, central or axial

Table 4 Distribution as per clinical grading system

Grade	Description	Number of patients (pre-operative)	Number of patients (post-operative)	L = lower cranial nerve deficits
Grade 1	Independent and normally functioning	17	30	
Grade 2	Walks on own but needs support/help to carry out routine household activities	19	22	1
Grade 3	Walks with minimal support and requires help to carry out household activities	12	3	1
Grade 4	Walks with heavy support and unable to carry out household activities	7	2	2
Grade 5	Unable to walk and dependent for all activities	2	–	1

atlantoaxial instability has been associated with chronic or long-standing spinal diseases such as cervical spondylosis, ossified posterior longitudinal ligament and Hirayama disease [28–31].

Earlier it was speculated that a short neck/torticollis, Klippel–Feil abnormalities, assimilation of the atlas, platybasia and several other musculoskeletal abnormalities were unrelated to embryonic dysgenesis but were secondary and naturally protective events in situations of long-standing or chronic atlantoaxial instability [32]. Altantoaxial stabilization can initiate immediate postoperative reversal of musculoskeletal changes. It was therefore decided that the term 'craniovertebral junction alterations' is preferable to the term 'craniovertebral junction anomalies' [1, 4]. As the various issues that are involved became more clarified, it was realised that just as musculoskeletal alterations are external manifestations, Chiari formation and syringomyelia can be internal and neural manifestations of atlantoaxial instability. Chiari formation is a protective 'airbag' that is placed in position in situations with potential or manifest atlantoaxial instability. It provides a natural cushion-like support for neural structures [33]. Along similar lines, it was speculated that syringomyelia is a protective self-neural alteration that ultimately works in the interests of neural structures and of the human body. Accordingly, it was decided that the term 'Chiari formation' is preferable to the term 'Chiari malformation', considering its functional role and the nature of its pathogenesis [34]. Subtle and long-standing atlantoaxial instability and minimal or no direct neural compression by the odontoid process allow both internal and external reparative processes to gradually develop, progress and mature.

In the patients in our series, anteroposterior measurements of the cervical spinal canal dimensions at the mid-C6 vertebral body level suggested that the spinal canal had increased in girth by 10%. Forty-eight patients had syringomyelia. In nine patients there was an external syrinx [7]. In 20 patients there were both internal and external syringes. Measurement of the total spinal cord girth revealed that the neural girth was reduced by 60% although the spinal canal dimension had increased by 10% (Table 5). The extra space thus created was filled with CSF, irrespective of whether it was present inside, outside, or both inside and outside the spinal cord. Evaluation of the posterior fossa content revealed that an excessive amount of CSF was present around the brainstem and cerebellum—an entity identified earlier as external syringobulbia [7]. The fourth ventricle was normal in its shape but larger in its anteroposterior dimensions. These spinal cord and cerebellar features suggested there was no evidence that the posterior fossa contents were tightly or compactly placed. The foramen magnum canal dimension was increased, suggesting that the herniated cerebellar tonsil progressively increased the foramen magnum size to facilitate its positioning.

Table 5 Radiological parameters

Radiological parameter	Present cohort	Normal
Cervical spinal canal dimension at mid-C6 vertebral body level	11–20 mm (mean 15 mm)	11–14 mm (mean 12.78 mm)
Cervical cord girth at mid-C6 vertebral body level	2–7.3 mm (mean 4.4 mm)	6.9–9.4 mm (mean 10.9 mm)

The primary aims of surgery in all cases were to achieve firm stabilization of the atlantoaxial joint and segmental arthrodesis. It was observed that cases in which direct atlantoaxial fixation was performed in the presence of Chiari formation and syringomyelia had marked venous sinusoidal pooling in the lateral mass gutters, making dissection of the region difficult and fraught with the risk of excessive blood loss. The exact cause of venous pooling in the region was unclear, but chronic and long-standing alterations in the pressure dynamics of the region probably resulted in venous congestion. Moreover, the presence of a lax dura, an atrophic cord and an increase in potential extradural space in the region of the craniovertebral junction increased the scope of venous pooling in the region.

Our observations that 100% of patients recovered from their symptoms to a considerable extent following the surgical procedure of atlantoaxial stabilization alone and that these improvements in neurological symptoms were evident in the immediate postoperative period suggest that atlantoaxial instability plays a role in the pathogenesis of Chiari formation and syringomyelia. Moreover, these improvements in neurological symptoms were observed in patients who had previously undergone failed foramen magnum decompression surgery. Direct observation of the joint status was possible during surgery when the lateral mass plate and screw fixation technique was applied [8–10]. Instability of the atlantoaxial joint was invariably identified during surgery. The primary aims of surgery in all cases were to achieve firm stabilization of the atlantoaxial joint and segmental arthrodesis. During the follow-up period, all patients had firm arthrodesis of the region and there were no instances of implant failure or infection. No foramen decompression was done in any case, although such treatment is currently considered to be the gold standard in the treatment of this entity. From this experience, it is clear that foramen magnum decompression surgery may be unnecessary or even counterproductive. Although the follow-up period in our series was relatively short, our positive results indicate that treatment for Chiari formation should be directed toward atlantoaxial stabilization. The fact that no patient's clinical symptoms worsened after initial improvement during the period of

follow-up provides further evidence of the validity of this concept.

This study reiterates our hypothesis that Chiari formation may be nature's own protective mechanism, providing a protective soft cushion, or airbag, to guard the vital spinal cord against potential or manifest compression by the odontoid process and pinching between bones [33]. It appears that Chiari formation is unrelated to reduced posterior cranial fossa volume and is not attributable to a primary or relative increase in cerebellar mass. The presence of syringomyelia, external syringomyelia and external syringobulbia suggest that the neural structures float within an excessive amount of CSF in the spinal canal and posterior fossa. As discussed earlier, the temporary neurological improvement that has been uniformly observed by several authors following foramen magnum decompression could be akin to the effects of deflating a full airbag, and, in the long term, such a form of surgery could be counterproductive. Syringomyelia may be a response of the body that balances the pressures within the neural structures in the interests of the patient [35, 36]. The presence of Chiari formation and syringomyelia suggests chronicity of the process and subtlety of atlantoaxial dislocation. The observations that syringomyelia was reduced and that the tonsils regressed from their herniated position in 52% of the patients in whom MRI was possible—without any form or bone manipulation or dural opening—validate this concept. To consolidate these observations, longer follow-up and multi-institutional experience are mandatory.

Compliance with Ethical Standards No financial support was received for this work.

Competing Interests The authors declare that they have no competing interests.

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The Relationship Between Basilar Invagination and Chiari Malformation Type I: A Narrative Review

Chenlong Liao, Massimiliano Visocchi, Wenchuan Zhang, Shiting Li, Min Yang, Wenxiang Zhong, and Pengfei Liu

Abstract Basilar invagination (BI) and Chiari malformation type I CM-I) are the most common adult craniovertebral junction malformations, and they are frequently associated with each other and present synchronously. The relationship between BI and CM-I has remained incompletely understood, and the choice of surgical strategy has remained controversial. This brief review focuses on the different aspects of BI and CM-I, and further discusses the relationship between these two concomitant pathologies on the basis of the concepts proposed over the last three decades.

Keywords Basilar invagination · Chiari I malformation Craniovertebral junction · Posterior cranial fossa

Introduction

Basilar invagination (BI) and Chiari malformation (CM) type I (CM-I) are the most common adult craniovertebral junction (CVJ) malformations [1]. These two pathologies are frequently associated with each other and present synchronously. According to craniometric studies, it seems that both of these pathologies belong to a spectrum of malformations whose common features include underdevelopment of the occipital bone and consequent neural and cerebrospinal fluid (CSF) flow compression [2]. However, each of these two malformations can present in isolation and be attributed to different aetiological factors and causal mechanisms [3–5]. The relationship between BI and CM-I has remained incompletely understood, thus hindering the development of therapeutic strategies.

With the purpose of better understanding these two malformations, this paper first focuses on different aspects of both BI and CM-I, and then discusses the relationship between these two concomitant pathologies on the basis of the concepts proposed over the last three decades.

Basilar Invagination

Definition and Aetiology

Basilar invagination is defined as a developmental anomaly of the CVJ in which the odontoid process abnormally prolapses upward and backward into the foramen magnum. However, several other terms such as ‘basilar impression’, ‘platybasia’ and ‘cranial settling’ have been interchangeably used in the literature to describe BI, causing terminological confusion over the years. In contrast to primary BI, the term ‘basilar impression’ refers to acquired or secondary BI, which may result from softening of the bone around the skull base. ‘Platybasia’ is an anthropological term describing an abnormally obtuse (>140°) angle between the anterior skull base and the clivus [6]. The term ‘cranial settling’ is typically used when BI is associated with rheumatoid arthritis.

Many pathologies may lead to development of BI. The reported aetiological factors include basioccipital/clivus hypoplasia, occipital condyle hypoplasia, atlas hypoplasia, an incomplete ring of C1 with spreading of the lateral masses, achondroplasia and atlanto-occipital assimilation (occipitalization of the atlas) [4, 5, 7, 8].

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Clinical Presentations

The clinical presentations of BI are related to compression of neural and vascular structures around the CVJ area, as well as obstruction of CSF circulation. The symptoms and signs are diverse because of the multitude of aetiological factors in BI and the wide range of structures involved, including signs of medullary dysfunction such as nystagmus, dysphagia, ataxia, dysmetria and cranial nerve palsy; and signs of myelopathy such as motor dysfunction (weakness, restricted neck movements), sensory dysfunction (neck pain, paraesthesia) and vegetative dysfunction (bowel and bladder disturbance). The clinical presentation varies depending on the underlying or accompanying pathological process [9].

Craniometric Measurement and Diagnosis

A series of craniometric measurements were employed to detect and diagnose BI in the previous literature. However, only a minor part of them are routinely used in clinical work, since most craniometric measurements lack adequate specificity and sensitivity [10]. There are three reference lines widely used in clinical work: the Chamberlain line, the McGregor line and the McRae line [11]. These three lines have been routinely adopted because of their high specificity, sensitivity and reproducibility. They are all viewed on lateral radiographs of the skull (Fig. 1).

The Chamberlain line extends from the posterior portion of the hard palate to the opisthion, which is the midpoint of

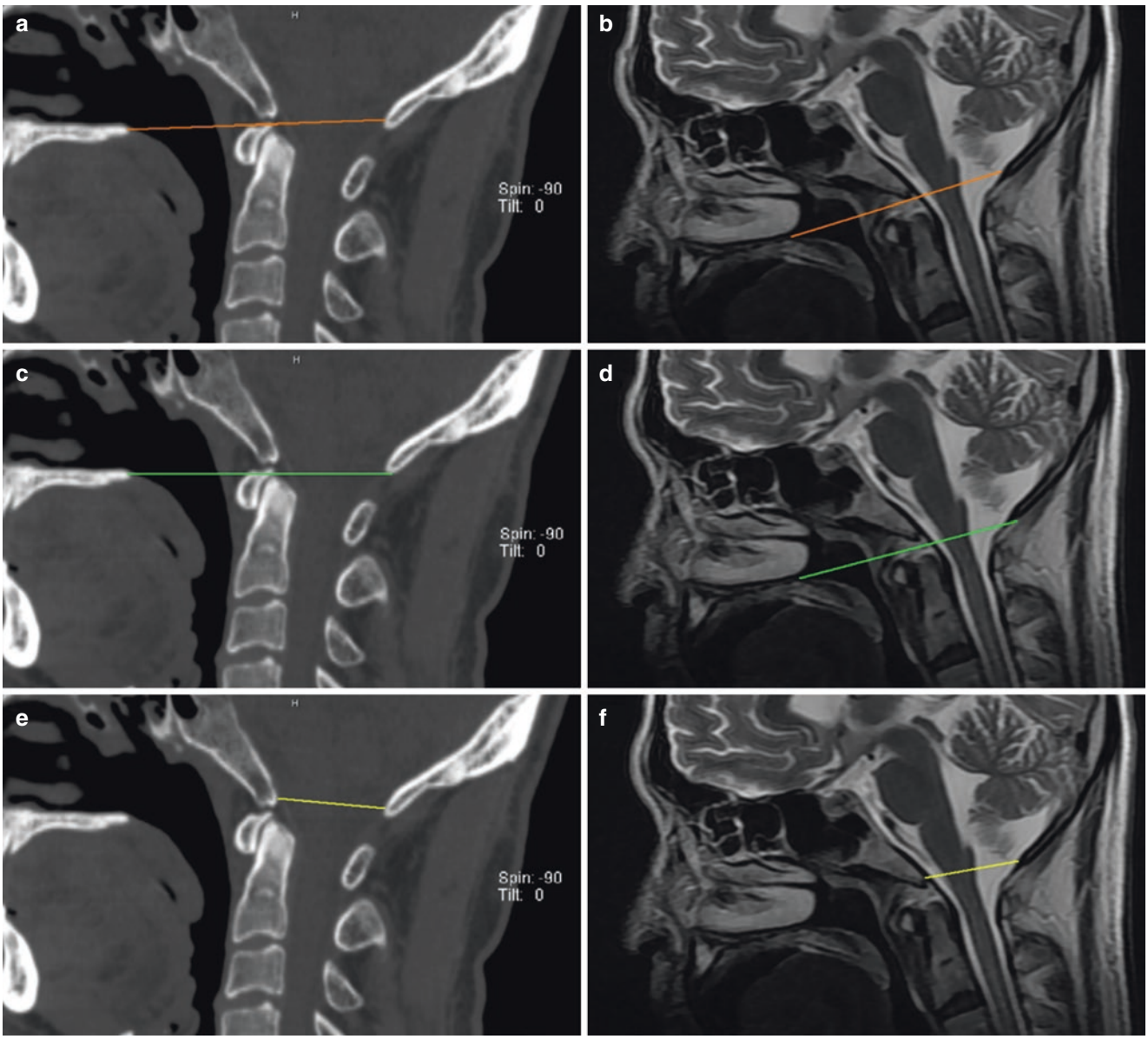

Fig. 1 Demonstration of three reference lines for measurement of basilar invagination (BI). (**a**) The Chamberlain line on a sagittal computed tomography (CT) scan [*orange*]. (**b**) The Chamberlain line on T2 magnetic resonance imaging (MRI) [*orange*]. (**c**) The McGregor line on a sagittal CT scan [*green*]. (**d**) The McGregor line on T2 MRI [*green*]. (**e**) The McRae line on a sagittal CT scan [*yellow*]. (**f**) The McRae line on T2 MRI [*yellow*]

the posterior margin of the foramen magnum (Fig. 1a, b) [6]. The tip of the odontoid process typically lies below or on the Chamberlain line. It is considered normal for the odontoid process to extend no more than 2.5 mm above this line, although the range varies, depending on the literature source [6, 12–14]. Generally, BI is considered present if the extension is greater than 5 mm [10].

Since precise identification of the opisthion on lateral radiographs is always difficult, McGregor proposed a modification of the Chamberlain line—the McGregor line—which extends from the posterior margin of the hard palate to the lowest point of the occipital squamosal surface (Fig. 1c, d) [12]. This line is about 2 mm inferior to the Chamberlain line, and thus it is considered abnormal when the tip of the odontoid process extends more than 7 mm above the line [12, 14].

The McRae line extends from the anterior (basion) to the posterior (opisthion) rim of the foramen magnum and is basically the anteroposterior length of the foramen magnum (Fig. 1d, e) [15]. The tip of the odontoid process should normally lie below this line. Furthermore, this line can also be used to define narrowing of the foramen magnum when the anteroposterior length is less than 19 mm in the sagittal plane [15].

While computed tomography (CT) is ideal for evaluation of osseous anatomy, magnetic resonance imaging (MRI) provides better assessment of soft tissue [8, 10]. In addition to conventional imaging—which includes plain radiography, CT and MRI—the development of dynamic imaging expands the ability to detect associated CVJ instability, functional stenosis of the spinal canal and cord compression [16–19].

Treatment Strategy

Surgical treatment for BI should be considered when neurological disturbance is present or foreseen. A large proportion of patients with BI present with neurological deficits resulting from compression of the cervicomedullary junction or the upper cervical cord [20–22]. The compression can be caused by a prolapsing dens or CVJ instability. Therefore, the definitive treatment for BI includes decompression and stabilization. Otherwise, in some patients without neurological symptoms where BI is an incidental finding, the necessity of surgical treatment should be carefully evaluated, since BI may present as a progressive deterioration and can result in neurological impairment and even sudden death if left untreated [23].

Thus, preoperative evaluation is of great importance. Aside from history taking and physical examination, nutritional status, dental hygiene and pulmonary status should be assessed [24]. Multimodal preoperative imaging of the CVJ, including CT and MRI (and even dynamic imaging), is an integral part of surgical planning [8, 10, 25]. In addition to searching for signs and sites of compression, preoperative assessment mainly focuses on the reducibility of BI, which is the critical factor in selection of the most suitable surgical approach.

A trial of axial cervical traction is usually performed in patients with BI to assess the degree to which the odontoid process might be reduced. It is now generally accepted that a posterior surgical approach alone, including decompression and fusion, can be adapted if the BI is reducible, while anterior decompression combined with posterior stabilization should be performed if the BI is not reducible [4, 23, 25–27].

Chiari Malformation Type I

Definition and Aetiology

Chiari malformation type I is characterized by downward herniation of the cerebellar tonsils and is defined as displacement of the cerebellar tonsils by more than 5 mm below the foramen magnum [28]. However, this is merely a radiographic definition, and the distances reported have varied from 3 mm to 5 mm, depending on the source [29, 30]. It has thus been suggested that the radiographic definition is limited to a terminological criterion and is not necessarily associated with clinical symptoms [31]. Therefore, cerebellar tonsil herniation of less than 5 mm or less than 3 mm does not exclude the diagnosis of CM-I.

In essence, CM-I is a disorder of the para-axial mesoderm, which is characterized by underdevelopment of the posterior cranial fossa (PCF) and overcrowding of the normally developed hindbrain [1, 32]. However, it can be associated with other miscellaneous conditions such as craniosynostosis [33], CSF leakage [34], Paget's disease [35] and intracranial mass lesions [36, 37]. Furthermore, five distinct causal mechanisms of cerebellar tonsil herniation—(1) cranial constriction, (2) cranial settling, (3) spinal cord tethering, (4) intracranial hypertension and (5) intraspinal hypotension—which have been reported to have diagnosis and therapeutic implications, were identified in a previous study [3]. Therefore, CM-I is a disorder of multiple aetiological factors, including genetic predisposition, congenital anomalies and acquisition through trauma or illness, and it should be better defined generically.

Clinical Presentations

Only a small proportion of patients with CM-I are symptomatic [38]. The clinical presentation may be attributed to the original disorders as well as to the secondary pathological changes such as syringomyelia, scoliosis and BI. Taking syringomyelia as an example, it has been reported that

patients with syringomyelia present at a slightly younger age and receive earlier diagnoses than patients without syringomyelia [31]. According to a large-scale patient-reported CM-I symptom study, pain is the most common symptom, and headache is the most frequently implicated type of pain [39]. Headache is also an indication for surgical treatment of CM-I in the absence of syringomyelia [38]. Other frequent clinical presentations of CM-I consist of a wide range of non-specific symptoms such as dizziness, sleeping disorder, neck pain, exhaustion and weakness [40].

Radiological Measurement and Diagnosis

According to the definition of CM-I, it is diagnosed radiologically as herniation of the cerebellar tonsils below the plane of the foramen magnum. However, the precise degree of herniation is not well established. It was suggested that in doubtful cases, cardiac-gated cine MRI is valuable in demonstrating a CSF flow obstruction as an indicator of clinically relevant herniation [41–43]. Furthermore, as the feature of CM-I, the ectopic tonsil position may result from a wide range of anatomical anomalies—such as craniosynostosis and an underdeveloped PCF—around the CVJ, leading to overcrowding of the PCF [44]. Therefore, analysis of the PCF is critical for better understanding of the pathogenesis and development of CM-I. Investigation of the PCF includes morphological and volumetric analyses, performed with the application of MRI.

Morphological Analysis of the Posterior Cranial Fossa

Measurements are performed on midline sagittal images, and four parameters are used to characterized the morphology of the PCF: (1) the length of the supraocciput, measured from the centre of the internal occipital protuberance to the opisthion; (2) the length of the clivus, measured from the tip of the dorsum sellae to the basion; (3) the slope of the tentorium, measured by calculation of the angle formed by the tentorium and a line drawn between the internal occipital protuberance and the opisthion; and (4) the extent of the cerebellar herniation, measured from the tips of the tonsils to a line drawn between the basion and the opisthion (Fig. 2) [31].

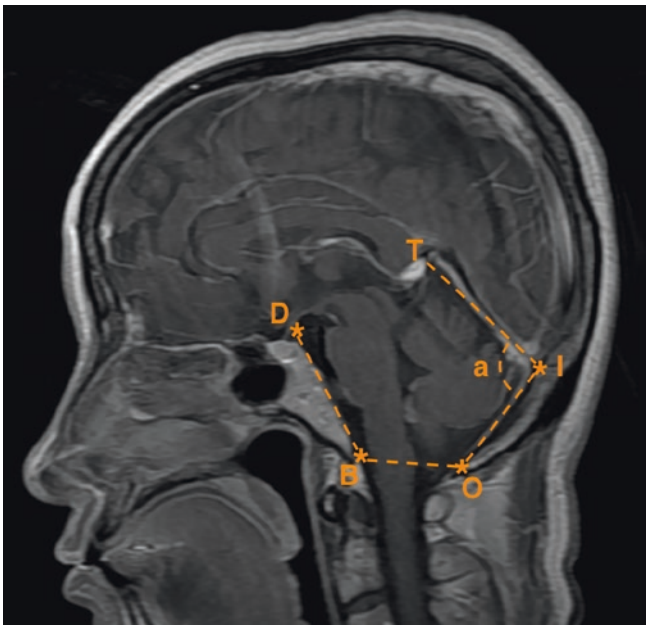

Fig. 2 Measurement of posterior cranial fossa (PCF) morphology on T1 sagittal magnetic resonance imaging (MRI). The length of the supraocciput (IO) is measured by the distance from the centre of the internal occipital protuberance (I) to the opisthion (O). The length of the clivus (DB) is measured by the distance from the top of the dorsum sellae (D) to the basion (B). The slope of the tentorium (T) is calculated from the angle (a) formed by the supraocciput and tentorium. The level of the foramen magnum is defined by a line drawn from the basion (B) to the opisthion (O)

Volumetric Analysis of the Posterior Cranial Fossa

The posterior cranial fossa is defined as a space bounded by a series of osseous anatomical structures, including the clivus, occipital bone, tentorium and bilateral petrous ridges. Volumetric measurement is performed by the Cavalieri method. A clear grid with regularly spaced dots (4 mm apart) is placed on each equally spaced consecutive axial image of the posterior fossa. The section thickness is recorded as T, while the number of points that fall on the posterior fossa for each slice is recorded as P_i. Then the distances between the dots on the grid are correlated with the actual distance on the MRI by comparison with the centimetre scale on the image. The area (A_p) between points on the grid is calculated by squaring the actual anatomical distance between each set of two adjacent dots. The volume of the PCF (V) is calculated with the use of the following equation: $V = A_p * \sum P_i * T$.

Apart from a reduced volume of the PCF, additional anomalies may involve the joint and discs of the CVJ. Klippel–Feil syndrome, atlanto-occipital assimilation and BI are features to look for in preoperative imaging, since these may indicate CVJ instability [24, 42, 45, 46]. Thus, routine employment of preoperative functional X-rays of the cervical spine in flexion and extension is recommended.

Treatment Strategy

The primary goals of surgery for CM-I include removing the compression from the brainstem and re-establishing CSF circulation. In the patient with syringomyelia, the goal is to prevent any additional neurological deficit and to decrease the size of the syrinx. However, there is no general consensus on the indication for the surgery. In a survey of American Society of Pediatric Neurosurgeons, Rocque et al. proposed that the presence of a syrinx seems to be a reasonable justification for operation [47]; other researchers have suggested that the decision regarding surgery should depend on the likelihood that a fixed neurological deficit as a consequence of the syrinx is more probable than spontaneous syrinx resolution, which has been observed, and the range of time necessary for a CM-I-related syrinx to resolve is yet to be explored [38, 48]. Another opinion from the perspective of symptomatology, held by Klekamp, is that asymptomatic patients have not been considered for surgery whether there was any sign of a syrinx or not, since he stated that he had not encountered an asymptomatic patient with a progressive syrinx [49].

With regard to the surgical strategy, although foramen magnum decompression is widely accepted as the treatment of choice for CM-I, controversies remain as to how this operation should be performed [50, 51]. The ongoing debate focuses on the risk and benefit of posterior fossa decompression alone versus posterior fossa decompression with duraplasty. According to a meta-analysis performed by Durham et al., posterior fossa decompression alone was associated with a significantly higher rate of reoperation (12.6% versus 2.1%) but a lower rate of CSF-related complications (1.8% versus 18.5%) than posterior fossa decompression with duraplasty [52]. In another meta-analysis, no convincing conclusion that one method was superior to the other could be drawn [53]. Complication rates are lower with procedures that leave the dura intact. However, this would be counterbalanced by lower rates of syrinx reduction and higher rates of symptom recurrence. Therefore, it appears that the surgical strategy should be tailored for individual patients according to preoperative evaluations using a series of radiographic tools and intraoperative assessments, such as intraoperative ultrasound [54–56].

On the other hand, another completely different surgical strategy has been proposed on the basis that the pathogenesis of CM-I is primary associated with atlantoaxial instability. In his series of studies, Goel suggested that CM-I may be a secondary phenomenon and a natural neural alteration in the face of atlantoaxial instability, and that surgical treatment should aim to restore atlantoaxial stabilization [57–59]. This conception was drawn from the observation of frequent concomitant presentation of BI and CM-I and the speculation that BI and CM-I are a continuum of the same pathological phenomenon originating from atlantoaxial instability, which needs to be investigated further.

The Relationship Between Basilar Invagination and Chiari Malformation Type I

In contrast to scenarios in which each of these two clinical entities presents in isolation, when BI is associated with CM-I (which frequently occurs in clinical observation), a variety of aspects—including the clinical presentation, natural progress, treatment strategy and prognosis—become very complicated and different.

Basilar Invagination with or Without Chiari Malformation Type I

The clinical course and symptoms of BI have been reported to be different when it is associated with CM-I. In an early study of 190 patients treated surgically for BI, the symptom onset of patients without CM-I was relatively acute, while the duration of symptoms of those with CM-I was long lasting. The most common presentations in the former population included weakness (100%), torticollis (69%), neck pain (59%), restricted neck movements (59%), posterior column dysfunction (39%), a low hairline (48%), a short neck (41%), bowel and bladder disturbance (28%) and paraesthesia (25%), while the most frequent presentations in the latter population included weakness (94%), paraesthesia (79%),

disturbances of the posterior column and spinothalamic tract (56%), a short neck (50%) and ataxia (47%). Accordingly, in that study, Goel et al. presented a classification system for BI, which was divided into two subgroups on the basis of absence of CM (group I) and presence of CM (group II) [26]. In essence, group I included patients with invagination of the odontoid process into the foramen magnum, as well as potential compression of the brainstem. The tip of the odontoid process was distanced from the anterior arch of the atlas or the inferior aspect of the clivus, suggesting the presence of instability of the CVJ. However, the angle of the clivus and the volume of the PCF were not affected. On the other hand, in group II a reduced PCF volume could be noted from the superior migration of the assembly of the odontoid process, the clivus and the anterior arch of the atlas, thus leading to the presence of CM-I. This classification provided a comprehensive understanding of the pathology and pathogenesis of the anomaly and would be helpful in selection of surgical treatment, as well as in prediction of the outcome. On the premise that CM-I in the presence of BI results from reduced PCF volume, it could be deduced that while anterior transoral surgery should be performed in group I patients, posterior foramen magnum decompression should be additionally performed in group II patients. In 2014, Visocchi et al. first reported that clinical and radiological resolution of CVJ compression after transoral correction of BI was evident for up to 2 years postoperation, at which time the child had a relapse of some of the presenting symptoms and follow-up CT and MRI scans showed quite complete regrowth of the odontoid process, partial clival regeneration and recurrence of preoperative CM. Also, in this case the correlation of BI and CM was further confirmed [60].

Chiari Malformation Type I with or Without Basilar Invagination

Different incidence rates of BI in patients with CM-I have been reported, ranging from 12% to 35% [24, 31, 61]. Although the symptoms of CM-I commonly include occipital pain and gait ataxia, several differences in clinical presentation are noted between patients who have CM-I with invagination and those who have CM-I without invagination. Patients with invagination tend to be more affected by caudal cranial nerve deficits and gait disturbances owing to ventral compression, while patients without invagination are more likely to suffer from sensory disorders or neuropathic pain, which may be related to syringomyelia. In comparison with radiological findings in patients without invagination, segmentation anomalies such as assimilation of the atlas or Klippel–Feil syndrome are more common in patients with invagination [61].

The combination with BI makes treatment of CM-I more complicated. Although complication rates have reportedly been significantly higher in patients who have CM-I with BI than in those who have CM-I without BI [61], the issue of whether the combination with BI is associated with a worse long-term outcome of surgical treatment of CM-I remain controversial [61–63]. The clinical outcome may depend on the treatment strategy adopted, as well as individual differences, and thus there is a series of subsequent questions that remain to be resolved. As stated above, symptomatic patients with CM-I require surgical decompression of the foramen magnum. However, whether additional operations such as ventral transoral decompression, traction and realignment should be incorporated for treatment of concurrent BI remains controversial, since the presence of ventral compression or instability should be carefully evaluated before or during the operation.

The sign of instability is sometimes difficult to detect on preoperational radiographs or CT scans because the range of motion may be restricted by biomechanical limitations. When tabling the paradoxical discussion on low rates of symptomatic improvement and high rates of recurrence with conservative surgery, or the other way around with aggressive surgery, Goel proposed a series of surgeries aiming at atlantoaxial stabilization by listing the negative long-term outcomes of foramen magnum decompression [64, 65] and claiming that CM-I was caused by atlantoaxial dislocation regardless of the presence or absence of BI [57]. However, in studies with large populations, the coincidence of atlantoaxial instability with CM-I is less frequent [31, 45, 66]. Furthermore, defining instability in complex patients can be challenging. Although intraoperative findings may provide clues regarding instability, no objective criterion is available so far for diagnosing instability during surgery. As demonstrated in Goel's clinical grading system for atlantoaxial dislocation, no gross physical or radiological abnormality may be present in cases of type III dislocation. It is illogical and even dangerous to perform a stabilization operation alone when instability is not clinically presented.

With regard to compression, the site and degree of compression should be assessed thoroughly for preoperational planning. When CM-I is associated with BI, aside from dorsal compression of the brainstem by herniated cerebellar tonsils, ventral compression by the odontoid peg may also be present. Is there any causal relationship between ventral and dorsal compression, thus making more decompression possible with less surgery? In a comparative study of 323 patients undergoing 350 operations, Klekamp demonstrated good short- and long-term results in patients who had CM-I with or without additional BI, through application of a treatment algorithm. He recommended that patients who had CM-I without BI and those who had BI without ventral compression could be managed by foramen magnum decompression alone, while most

patients with ventral compression could be treated by posterior decompression, realignment and stabilization, reserving anterior decompression for patients with profound, symptomatic brainstem compression [61].

With further development of the research on the pathological theory and surgical strategies, the relationship between BI and CM-I—and the potential compression and instability involved in these two concomitant pathologies—should become better understood and managed.

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Competing Interests The authors declare that they have no competing interests.

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Bony Decompression for Chiari Malformation Type I: Long-Term Follow-Up

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Abstract *Background:* Several surgical techniques are used for the management of Chiari malformation type I (CM-I). Bony posterior fossa decompression is considered a good option in children, though with a higher risk of requiring reoperation. However, there is not enough evidence from the series in the literature, which are often limited by inadequate follow-up. The goal of this study was to assess the effectiveness of suboccipital craniectomy alone in children after long-term follow-up.

Methods: Forty-two children (25 female and 17 male; mean age 6.7 years), operated on with bony decompression alone, were retrospectively reviewed. All patients underwent suboccipital craniectomy. Thirty-eight children required C1 laminectomy, and 21 also underwent dural delamination on the basis of intraoperative ultrasound investigations. The outcome was assessed using the traditional measurement and the Chicago Chiari Outcome Scale (CCOS). The mean follow-up period was 11.3 years (range 5–15 years).

Results: Headache was the most frequent preoperative symptom (81%), followed by neck pain (40%), vertigo (40%), ataxia (26%), and upper and lower extremity paraesthesia (26%). Syringomyelia was present in 19 patients (45%). Resolution and significant improvement of preoperative clinical symptoms were observed in 36.5% and 21.5% of cases, respectively. Three children required adjunctive surgery for symptom recurrence (7%). The tonsil position and syringomyelia were normalized or improved in 50% and 79% of cases, respectively. No complications occurred. According to the CCOS scores, 69.5% of children had an excellent outcome, 28.5% had a functional outcome and 2% had an impaired outcome.

Conclusion: Bony decompression alone is an effective, safe and long-lasting treatment for children with CM-I. A certain risk of symptom recurrence requiring new surgery exists, but it is widely counterbalanced by the low risk of complications. Careful patient selection is crucial for a good outcome. Prospective and randomized studies are needed for further validation.

Keywords Chiari I malformation · Syringomyelia · Posterior fossa decompression · Duraplasty · Intraoperative ultrasound

Introduction

Chiari malformation type I (CM-I) is a heterogeneous condition encompassing a wide spectrum of clinical and radiological presentations. Several surgical techniques are currently used for its correction, ranging from suboccipital craniectomy/C1 laminectomy alone to transoral or transnasal ventral decompression followed by posterior stabilization, and including endoscopic posterior decompression, dural delamination, duraplasty, arachnoid dissection with/without tonsil coagulation, and minimally invasive tonsillectomy [1–9]. When one is dealing with CM-I patients, the first limitation is the lack of standardized surgical management. In fact, no guidelines are available in the literature, where no evidence of a 'best' treatment can be found. Similarly, no standardized methods for outcome evaluation have been identified so far [10]; a further limitation, partially related to this, is represented by the relatively short follow-up reported in many series. In this paper we present the late outcome of a paediatric series after long follow-up to contribute information on the management of selected cases with bony decompression alone.

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Materials and Methods

All consecutive paediatric patients (aged 0–16 years) surgically treated at our institution with suboccipital craniectomy (with/without C1 laminectomy) in the decade of 2001–2011 were considered. This time period was established to ensure a minimum of 5-year follow-up. The patient-tailored selection criteria in use at our institution can be summarized as follows: (1) asymptomatic patients without relevant syringomyelia (<2 mm thickness): clinical and radiological follow-up; (2) symptomatic patients with tonsillar herniation above C2 or asymptomatic patients with 2–5 mm syringomyelia: bony decompression of the posterior cranial fossa; and (3) symptomatic patients (especially those with signs of brainstem dysfunction) with tonsils below C2 and/or >5 mm syringomyelia: bony decompression plus duraplasty (and subpial tonsil coagulation, if needed). Patients with psychomotor delay or behavioural problems but without clear signs/symptoms of CM-I were not considered for surgery but were enrolled for follow-up.

All patients underwent a preoperative workup consisting of physical and neurological examination, craniospinal magnetic resonance imaging (MRI), polysomnography and neuropsychological tests. The same examinations were utilized for the postoperative follow-up with the following timing: physical and neurological examinations every 6 months for the first 2 years, then yearly; neuropsychological tests yearly up to the scholar age and working age, according to the patients' characteristics; craniospinal MRI yearly for the first 3 years, then every 2 or 3 years; and polysomnography 1 year after surgery in negative cases and yearly until normalization in positive cases. Decompression of the posterior fossa was achieved by suboccipital craniectomy with/without C1 laminectomy and/or delamination of the external layer of the dura mater. The epidural fibrous ring at the occipitocervical junction was resected. The extension of the craniectomy, as well as the laminectomy, were tailored on the basis of the patient's age and characteristics. In any case, the re-expansion of the cisterna magna and pulsations of the tonsils were assessed by intraoperative ultrasound (IUS), and the extent of the craniectomy was adapted accordingly. The posterior arch of C1 was routinely removed unless the posterior compression at that level was not relevant. Beyond the traditional treatment outcome measurement ('resolution', 'improvement', 'stability' or 'worsening'), the Chicago Chiari Outcome Scale (CCOS) was used for outcome assessment [11]. According to the CCOS, the outcome is graded as 'excellent' with a score of 16 points, 'functional' with a score of 12–15 points, 'impaired' with a score of 8–11 points and 'incapacitated' with a score of 4–7 points.

Results

Patient Characteristics

Among 164 paediatric patients admitted for CM-I during the relevant period, only 42 children were eligible for the study. Indeed, more than one third of all cases were asymptomatic, while the others needed duraplasty at least. Moreover, in six cases where a suboccipital craniectomy was planned, IUS did not show good expansion of the cerebellomedullary cistern, so duraplasty was performed (and they were not included in the present series).

The cohort was composed of 17 boys and 25 girls (male to female ratio 0.68), with a mean age of 6.7 years (range 15 months–16 years) at the time of surgery. The mean follow-up period for the entire cohort was 11.3 years, ranging from 5 to 15 years. In five cases (12%), the diagnosis was incidental (asymptomatic patients), while 15 children (35.5%) presented with specific signs—such as macrocrania and psychomotor delay—other than a suboccipital headache. Finally, 22 patients (52.5%) showed CM-I/syringomyelia–specific symptoms and signs, such as a suboccipital headache, neck pain, vertigo, ataxia, upper and/or lower limb paraesthesia and scoliosis (see Table 1). Tonsillar caudal herniation was detected in all cases: a 5-mm ectopia in eight cases (19%), a 6- to 10-mm ectopia in 18 cases (43%) and a ≥11 mm ectopia in 16 cases (38%). Syringomyelia was present in 19 patients (45%): cervical (eight cases, 43%), dorsal (three cases, 15.5%), cervicodorsal (five cases, 26%) and holocord (three cases, 15.5%) (see Table 2). Neuropsychological tests did not show significant alterations, with normal full-scale IQ (FSIQ), verbal IQ (VIQ) and performance IQ (PIQ) scores in 88% of cases (37 patients). In five children, some working memory, planning and attention disorders were detected, which clearly correlated with sleep disorders. Polysomnography was normal in 35 cases (83.5%) but revealed sleep-disordered breathing in seven cases (16.5%).

Surgical Outcome

All patients underwent suboccipital craniectomy. C1 laminectomy was carried out in 38 cases (90%), while dural delamination was performed in 21 cases (50%). At the end of the surgical procedure, IUS showed a satisfactory fluid film between the dura and the tonsils, and good tonsil pulsation in all cases. Complete resolution of the clinical picture at late follow-up was achieved in 76.5% of cases (32 patients) and an improvement in nine cases (21.5%); only in one case was

Table 1 Clinical symptoms at onset and late outcomes

Clinical symptom or sign	Cases at diagnosis [*n* (%)]	Late outcomes [*n* (%)]		
		Resolution	Improvement	Stability
Suboccipital 'cough' headache	34 (81)	30 (88)	2 (6)	2 (6)
Neck pain	17 (40)	14 (82)	2 (12)	1 (6)
Vertigo	16 (38)	14 (87.5)	1 (6.25)	1 (6.25)
Ataxia	11 (26)	10 (91)	–	1 (9)
Paraesthesia	11 (26)	9 (82)	1 (9)	1 (9)
Sleep-disordered breathing	7 (16.5)	5 (71.5)	2 (28.5)	–
Progressive scoliosis	5 (12)	4 (80)	–	1 (20)
Psychomotor delay	5 (12)	4 (80)	–	1 (20)
Macrocrania	4 (9.5)	–	–	4[a]

[a]This sign was not considered in the results because it was not correlated with Chiari malformation type I

Table 2 Radiological findings and outcomes

Finding	Cases at diagnosis [*n* (%)]	Late outcomes [*n* (%)]		
		Resolution	Improvement	Stability
Tonsil herniation				
5 mm	8 (19)	5 (12)	1 (2.5)	2 (5)
6–10 mm	18 (43)	6 (14)	3 (7)	9 (21.5)
≥11 mm	16 (38)	2 (5)	4 (9.5)	10 (23.5)
Total	42 (100)	13 (31)	8 (19)	21 (50)
Syrinx				
Cervical	8 (43)	4 (21)	3 (15)	1 (5)
Dorsal	3 (15.5)	–	2 (11)	1 (5)
Cervicodorsal	5 (26)	1 (5)	2 (11)	2 (11)
Holocord	3 (15.5)	1 (5)	2 (11)	–
Total	19 (45)	6 (31)	9 (48)	4 (21)

there no improvement (2%). The evolution of the clinical symptoms is reported in detail in Table 1. These results were obtained after three patients (7%) experienced symptom recurrence ranging from 1 to 7 years after the first operation (all of them had experienced initial clinical improvement). All of them underwent second-look surgery: in the first patient (the youngest in the series), a newly formed suboccipital bone was found and was removed again, with clinical resolution; in the second patient, a thick fibrous scar was found, and clinical normalization was achieved after its excision; the third case required two operations (the first one for fibrous scar removal followed by a second one for tonsil coagulation and duraplasty), resulting in only transient symptom improvement. No surgical complications occurred except for postoperative neck pain in one third of cases, requiring extra analgesic drug therapy. On the basis of CCOS scores and considering the single reoperation as a 'transient complication' and the double reoperation as a 'permanent complication, well controlled', 29 patients (69.5%) had an excellent outcome, 12 (28.5%) had a functional outcome and one (2%) had an impaired outcome.

The position of the cerebellar tonsils remained stable or improved in 69% of cases (29 patients), while normalization of their position was achieved in the remaining 13 cases (31%). The tonsil ascent was mainly observed in the late phases of follow-up, often seven or more years after surgery (Fig. 1). Syringomyelia disappeared or showed a significant reduction in 31% (six patients) and in 48% of cases (nine patients), respectively; it remained unchanged in the remaining four cases (21%). The radiological results are summarized in Table 2. Polysomnography improved in all cases, with normalization in five of seven patients. A sleep-correlated improvement in the neuropsychological deficits was detected in all five affected cases, with normalization in four of them.

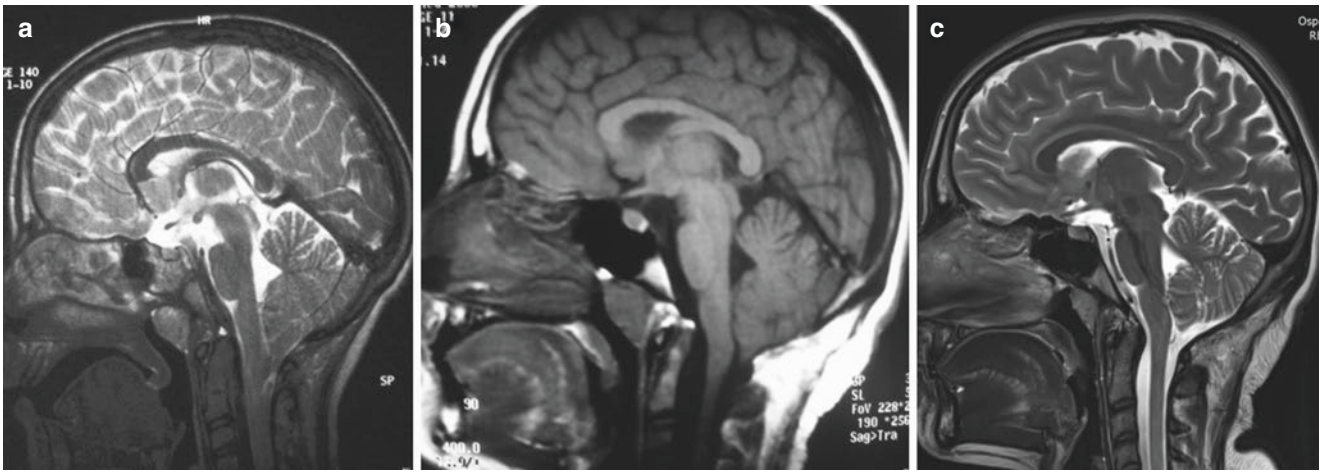

Fig. 1 Magnetic resonance imaging of a boy operated on for Chiari malformation type I at the age of 6 years. (**a**) Preoperative image showing a stenosis of the foramen magnum with 8 mm caudal descent of the cerebellar tonsils. (**b**) Postoperative image after 3-year follow-up, demonstrating ascent of the cerebellar tonsils but not yet normalization. (**c**) Normalization of the radiological picture after 6.5-year follow-up

Discussion

According to a recent analysis of a large US nationwide healthcare network, the numbers of surgical procedures performed for CM-I increased by 51% in the paediatric population and by 28% in the adult population in the last 14 years [12]. Despite this large number of surgical procedures, the 'old' dilemma regarding decompression of the posterior fossa—posterior fossa bony decompression alone (PFD) versus posterior fossa bony decompression plus duraplasty (PFDD)—has not been resolved yet. Although the matter is still controversial, PFDD is currently suggested in adults—despite the higher rate of complications and the longer operation time—on the basis of the significantly better outcome and the lower recurrence (and reoperation) rates [13–16]. As far as children are concerned, the current trend seems to be in favour of PFD, though there is no level I or IIa evidence for this [17]. Some authors, however, advocate the use of duraplasty for the same reasons as those given for adults [18, 19], whereas many authors propose to reserve duraplasty for the small number of children who experience recurrence after bony decompression, which shows a significantly lower rate of complications (especially cerebrospinal fluid [CSF] leakage) [20–25] and virtually no risk of surgical death [26]. One meta-analysis (which is still the only one available in the literature for children), published by Durham and Fjeld-Olenec in 2008, considered seven papers describing 582 paediatric patients treated by suboccipital craniectomy alone (266 cases) or with duraplasty (316 cases), and found that the outcome was statistically similar with the two techniques in term of clinical improvement and syrinx reduction, while PFDD ensured a significantly lower rate of recurrence (2.1% versus 12.6%) [27]. As expected, the data were significantly in favour of PFD when CSF leakage was considered (1.8% versus 18.5%), with no substantial differences regarding the other complications. The authors noted that the follow-up (ranging from 2 to 120 months) was 'inconsistently reported across studies'.

The goal of our study was to report a series with a sufficient length of follow-up to present definitive results. After 11.3 years of follow-up, 76.5% of our cases showed clinical resolution and 21.5% of them showed a considerable clinical improvement, with an excellent outcome in nearly 70% of cases and a functional outcome in 28% of cases (according to their CCOS scores). Only one patient did not show any improvement and had an impaired outcome despite two reoperations (the second one involved subpial tonsillectomy). On these grounds, PFD can be definitely considered a valid option for surgical management of CM-I in children. A certain risk of recurrence exists (7% in the present series) but can be successfully managed by second-look surgery. Moreover, permanent and/or significant complications are virtually absent. Indeed, the risk of reoperation is counterbalanced by the low rate of complications, so the final balance between complications and reoperations in PFDD and in PFD seems to be more favourable in PFD [28, 29]. To achieve this outcome, it is necessary for the patients to be carefully selected, and PFD is used in less severe cases [22, 30]. On these bases, the less satisfactorily radiological results do not represent a limitation since they are frequently reported in the literature and do not preclude a good clinical outcome [20, 31, 32].

Such good outcomes of PFD in children result from two main factors. The first one is progressive enlargement of the posterior fossa in children, which is due to physiological growth of the skull in the paediatric age group. In contrast to the scenario in adults, this growth may make PFDD unneces-

sary simply because the bone opening supports the spontaneous bone enlargement in providing space for the neural structures of the posterior fossa. For these reasons, the extent of bony decompression is crucial and must be tailored according to the patient's characteristics and age. Indeed, in infants and young children the suboccipital craniectomy should not be too large (because of the risk of a pseudomeningocele resulting from cerebellar pulsations) and should not be too small (because of the risk of bone re-formation, as happened in one patient in our series) [6]. In patients with thicker, more slowly growing bone, such as older children and adolescents, the extent of bone removal should be estimated on the basis of the severity of the clinical and radiological picture. IUS currently is the most widely used tool to assess the correct extension of craniectomy/laminectomy because it allows the neurosurgeon to directly visualize the re-expansion and pulsation of the cisterna magna. In our experience, IUS was effective in achieving this goal: six patients (not included in the present series) were actually managed with PFDD because PFD did not provide good intraoperative results; moreover, among the three patients with recurrence, only one needed PFDD, while the other two patients were managed with newly formed bone and scar removal. Successful experiences with IUS have been reported by other authors [33, 34], even though it is less effective in cases of tonsil herniation caudal to C1 [35]. A criticism of this strategy has been raised by the use of intraoperative MRI in CM-I surgery. Bond et al. recently reported their experience involving 15 adults who underwent pre-incision prone intraoperative MRI: at that time, 93% of them (14 of 15 cases) showed a substantial improvement in the CSF flow dorsal to the cerebellar tonsils [36]. Therefore, one could speculate that the prone position is sufficient to improve CSF dynamics, so bony decompression does not add significant advantages to the operation. However, this experience needs to be replicated in children, where the physiologically 'crowded' posterior fossa (in comparison with that in adults) would not respond so well to prone positioning alone.

The second crucial factor is careful patient selection. The main limitation of our study, which is also the main merit of our management policy, is represented by a selection bias due to the adopted criteria (see 'Materials and Methods'). This is the reason why we did not present a comparative cohort: since the patients treated with PFD and those treated with PFDD have different clinical and radiological characteristics, it is not possible to compare them. At the same time, however, this policy allowed us to select the most suitable candidates for PFD and to obtain good and stable long-term results. These considerations apply also to the literature [23, 25, 37, 38] and are further supported by the very recent experience reported by Pomeraniec and co-workers in their series of 95 children with CM-I [39]. After careful patient selection, 70 children were managed conservatively and had a good outcome without clinical or radiological worsening. Only 25 children underwent surgery, with a significant clinical improvement (75%) and radiological improvement (87.5%), regardless of the surgical technique used (bony decompression with a dural split versus duraplasty).

Taking into account the limitations inherent in a retrospective analysis of a single cohort from a single institution, this analysis of long-term results allows us to conclude that in children, PFD should be considered as a first-choice option in patients with a less severe clinical picture (no signs of brainstem dysfunction, tonsils above C2, thin syringomyelia), who are often the majority among paediatric patients. PFDD, with or without intradural manipulation, should be performed in more severe cases or in rare cases of failure of the previous strategy. The good final outcome of our series, according to the CCOS scores, shows that overall the risk of recurrence after PFD is broadly balanced by the very low risk of complications. For correct outcome assessment, a sufficiently long follow-up period (at least 7–10 years) is mandatory. Further prospective and randomized studies are required to validate these observations.

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Competing Interests The authors declare that they have no competing interests.

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Surgical Treatment in Symptomatic Chiari Malformation Type I: A Series of 25 Adult Patients Treated with Cerebellar Tonsil Shrinkage

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Abstract *Background:* The variety of symptoms and radiological findings in patients with Chiari malformation type I makes both the indication for surgery and the technical modality controversial. We report our 5-year experience, describing our technique and critically evaluating the clinical results.

Methods: Between 2012 and 2016, 25 patients (15 female and 10 male; mean age 39.2 years) underwent posterior fossa decompression for Chiari malformation type I. Their clinical complaints included headache, nuchalgia, upper limb weakness or numbness, instability, dizziness and diplopia. Syringomyelia was present in 12 patients (48%). Suboccipital craniectomy was completed in all cases with C1 laminectomy and shrinkage of the cerebellar tonsils by bipolar coagulation; duraplasty was performed with a suturable dura substitute.

Results: Gratifying results were observed in our series. Symptoms and signs were resolved in 52% of patients, and 20% of patients had an improvement in their preoperative deficits. The symptoms of six patients (24%) were essentially unchanged, and one patient (4%) deteriorated despite undergoing surgery. Generally, patients with syringomyelia on magnetic resonance imaging (MRI) showed less symptomatic improvement after surgery. The syrinx disappeared in seven of 12 patients, and complications occurred in three patients (12%).

Conclusion: Cerebellar tonsil reduction and restoration of cerebrospinal fluid (CSF) circulation provided clinical improvement and a stable reduction in the syrinx size in the vast majority of treated patients, with a low rate of complications.

Keywords Chiari malformation type I · Posterior fossa decompression · Cerebellar tonsil shrinkage · Syringomyelia Duraplasty

Introduction

Chiari malformation type I (CM-I) is defined by an ectopic position of the cerebellar tonsils under the foramen magnum [1]. The prevalence of the disease, based on an anatomical definition, is almost 1% in the general population [2] (ranging between 0.56% and 0.77% in the reported literature) [3, 4], and the rate of incidentally discovered malformations has markedly risen with extensive use of magnetic resonance imaging (MRI) [5]. However, only a small proportion of these patients will show signs or symptoms during their lives.

CM-I usually presents after the second or third decade of life [6]. In 30–70% of patients, the malformation is associated with syringomyelia [7, 8].

Evolutionary observations have led to the conclusion that CM-I is a consequence of asynchronism between brain growth and braincase growth [9]. Advances in brain MRI have clearly demonstrated a decreased posterior fossa volume as the common characteristic of CM-I: this is mainly due to basioccipital hypoplasia and sometimes platybasia causing posterior fossa 'overcrowding' and consequently tonsil herniation through the foramen magnum [10–12].

The indication for surgery is usually driven by the presence of a syrinx. Although some patients have been shown to have spontaneous syrinx resolution, the decision to proceed

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with surgery depends on the fact that a neurological deficit caused by the syrinx is more probable than spontaneous syrinx resolution [13]. When syringomyelia is absent, a continuous headache is the main complaint of patients and is exacerbated by Valsalva-like manoeuvres [14].

A variety of technical modalities to perform decompression have been described in the literature [15]; one such modality is suboccipital craniectomy with C1 laminectomy. If insufficient decompression is achieved, surgery can go further with enlargement of the dura. Some authors have advocated incision of only the external layer of the dura (preserving the internal layer); another option is to perform duraplasty, opening both layers of the dura with a vertical [16] or Y-shaped incision [17, 18] and suturing it to the periosteum [19–21], the fascia lata [22] or an artificial dura [18]. Dural opening may be accompanied by subarachnoid space exploration to visualize the foramen of Magendie [23, 24]. Some authors consider bilateral resection of tonsils necessary to achieve optimal decompression of the occipitocervical junction [25–27].

We describe a single-institution, 5-year experience in an adult population: all patients were treated by duraplasty along with bipolar coagulation to shrink cerebellar descendent tonsils, the goal being opening of the obex to restore cerebrospinal fluid (CSF) circulation.

Patients and Methods

We performed a retrospective review of patients with CM-I treated in the Division of Neurosurgery at the ARNAS Civico Hospital in Palermo, analysing medical and surgical records, and radiological imaging. We collected data on the presentation, management, outcome and follow-up evaluations of these patients.

Twenty-five patients with CM-I underwent surgical treatment at this institution between January 2012 and December 2016. The mean age of the patients was 39.2 years (range 19–66 years). There were 15 female patients (60%) and 10 male patients (40%).

Clinical Presentation

All patients were symptomatic; the duration of their initial symptoms prior to definitive diagnosis ranged between 6 months and 4 years (mean 18.2 months).

Pain was the most frequent symptom, occurring in 68% of cases (Table 1). In six patients the headache was limited to the nuchal region, while it was diffuse in the other patients. The occipital headache was associated with radicular pain to the upper limbs in one patient. Ten patients (40%) complained of weakness mostly in one or both arms, and eight (32%) complained of loss of sensation, mostly in the hands. Other common symptoms were instability (in 16% of patients), dizziness and vertigo (in 8%) and diplopia (in 4%).

Table 1 Presenting symptoms in 25 cases of Chiari malformation type I

Symptom	Cases [*n* (%)]
Pain	17 (68)
Headache	10
Neck	6
Upper limb	1
Weakness	10 (40)
Upper limb	9
Lower limb	1
Sensory disturbance	8 (32)
Upper limb	7
Trunk	1
Instability	4 (16)
Dizziness and vertigo	2 (8)
Diplopia	1 (4)

The neurological examination showed motor deficits: hyposthenia, atrophy or reflex changes in 13 patients (52%) and gait instability in four patients (16%). Eight patients had objective sensory disturbances, and in four patients (16%) we found dissociated sensory loss (loss of pain and temperature sensations with preserved touch and joint position sense). One patient had unilateral paresis of the sixth cranial nerve. None of the patients exhibited lower cranial nerve dysfunction.

Radiological Evaluation

All patients had preoperative head and cervical MRI including sagittal and axial spin-echo T1 and T2 sequences to confirm in all cases that the cerebellar tonsils descended into the cervical canal and to search for associated syringomyelia. CSF velocity/flow studies (cine-MRI) were not performed in all cases, so we do not discuss them in this paper. The downward migration of the cerebellar tonsils was measured on MRI and ranged from 5 to 21 mm (only two patients had severe migration >15 mm).

Syringomyelia was present in 12 patients (48%): cervical syringes were found in eight patients and cervicothoracic syringes in four patients. Scoliosis was observed in one patient. None of the patients showed associated spinal dysraphism or basilar invagination.

Surgical Treatment

Patients were operated on in a prone position with the head fixed in a Mayfield® three-point headrest (Integra LifeSciences Corporation, Cincinnati, OH, USA). A midline skin incision extending from the external occipital protuberance down to the level of C2–C3 was performed. A large enough suboccipital craniectomy and C1 laminectomy were carried out in all cases. Under microscopic visualization, a Y-shaped incision was made in the dura mater. Arachnoid dissection allowed exposure of the herniated cerebellar tonsils: the pial surface was cauterized by bipolar coagulation (on a low setting) to achieve tonsil shrinkage. The procedure was completed with exploration of the obex to restore CSF circulation. In all cases the dura was patched with lyophilized bovine pericardium (Lyoplant®, B. Braun Aesculap, Tuttlingen, Germany) (Fig. 1). The graft was closed with a watertight suture and fibrin glue was added around the dural closure to ensure sealing. The anaesthesia team performed Valsalva manoeuvres to test the integrity of the closure.

According to the available recordings, the mean operating time was 120 min (range 80–160 min); the longer surgical procedures were performed in those patients with a short neck or hard tonsils.

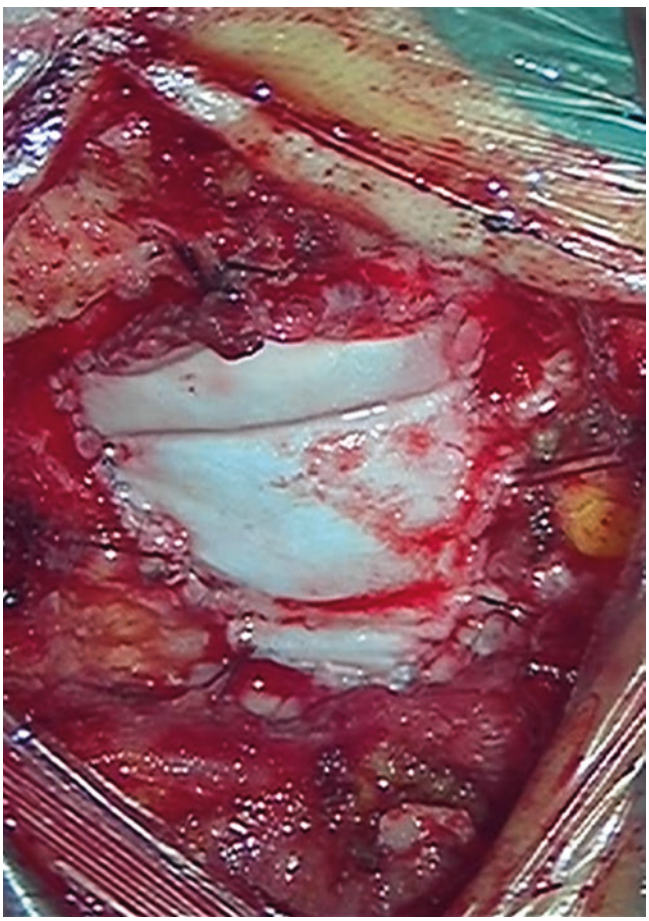

Fig. 1 Intraoperative view of duraplasty with a sutured patch

Postoperative Assessment

All patients underwent early computed tomography (CT) scanning on postoperative day 1. The clinical evaluation consisted of a neurological examination at discharge and at the first outpatient visit after almost 1 month. Patients with syringomyelia had MRI of both the brain and the whole spinal cord at the first outpatient visit, while patients with isolated cerebellar herniation had craniocervical MRI at the second visit 3 months after surgery.

In cases where serious clinical manifestations persisted we performed further MRI at 6-month follow-up. Thereafter, patients were re-evaluated 12 months after surgery and by a telephone questionnaire at 2 years or more, according to follow-up.

The follow-up period ranged from 3 to 60 months (mean 33 months).

Results

There were no major intraoperative complications or deaths, and gratifying results were achieved in our series. One patient's surgery was rescheduled because of the onset of severe bradycardia at the induction of general anaesthesia, but there were no neurological sequelae.

The changes in each symptom were graded at the last follow-up visit, as follows: complete remission, improvement, stabilization or worsening; in the event of multiple symptoms and inconsistent outcomes, we considered the worst outcome among the different symptoms. None of the patients experienced new-onset symptoms after surgery. Evaluation of the entire series showed that symptoms improved or were cured after surgery in 72% of patients. Table 2 shows the overall outcomes in our series. In 52% of patients, complete remission was achieved, with symptoms resolved. Twenty per cent of patients had an improvement of their preoperative deficits. Six patients (24%) were classified as stable, as some of their preoperative deficits improved while others were stabilized. Finally, one patient (4%) deteriorated despite undergoing surgery.

More specific analysis showed that those patients whose complaint was pain responded very well to surgery, with complete resolution in 14 of 17 cases (82.3%). Headaches or

Table 2 Long-term overall outcome in 25 cases of Chiari malformation type I

Outcome	Cases [*n* (%)]
Complete remission	13 (52)
Improvement	5 (20)
Stabilization	6 (24)
Worsening	1 (4)

nuchalgia improved within the first 10–15 days after surgery in ten patients. On the other hand, preoperative motor deficits were cured (with a return to normal strength) in only four of ten patients (40%). Moreover, sensory deficits responded poorly to surgery: four of eight patients (50%) with such deficits had no change, while one patient experienced deficit progression. In general, dizziness and vertigo also improved, except in one patient with significant cervical spondylosis, which remained stable. With regard to outcomes in cases of cranial nerve deficits, the single patient with abducens nerve dysfunction had complete resolution of this sign (Table 3).

Generally, after surgery, patients who had initially presented with syringomyelia on MRI showed less symptomatic improvement than patients without a syrinx (52.6% versus 93.3%) (Fig. 2).

Table 3 Long-term symptomatic outcome in 25 cases of Chiari malformation type I

Symptom	Cases [*n* (%)]
Pain	17
Resolved	14 (82.3)
Improved	1 (5.9)
Unchanged	2 (11.8)
Worsened	0
Weakness	10
Resolved	4 (40)
Improved	4 (40)
Unchanged	2 (20)
Worsened	0
Sensory disturbance	8
Resolved	2 (25)
Improved	1 (12.5)
Unchanged	4 (50)
Worsened	1 (12.5)
Instability	4
Resolved	3 (75)
Improved	1 (25)
Unchanged	0
Worsened	0
Dizziness and vertigo	2
Resolved	1 (50)
Improved	0
Unchanged	1 (50)
Worsened	0
Diplopia	1
Resolved	1 (100)
Improved	0
Unchanged	0
Worsened	0

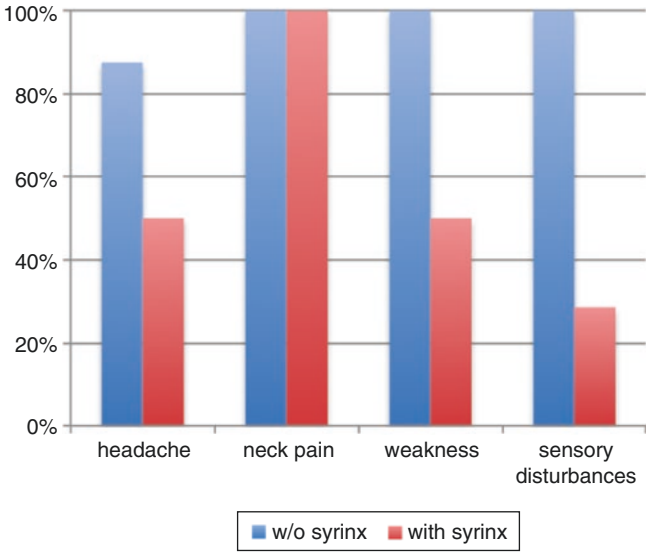

Fig. 2 Comparison of general improvements in symptoms in patients with and without syringomyelia

The syringes disappeared in seven of 12 cases (Fig. 3). In three patients this was evident on postoperative MRI obtained between 1 and 3 months after surgery. In four other cases, syringomyelia was considerably reduced but still present at the last follow-up. Conversely, in one patient the syrinx had not decreased at the 6-month follow-up MRI examination. In no patient was an increased syrinx demonstrated postoperatively.

Complications occurred in three patients (12%): one had a superficial wound infection; one had a CSF leak, which was treated conservatively; and one had a pseudomeningocele, which was treated initially with temporary lumbar drainage and later required a second surgical procedure for revision of the duraplasty.

Discussion

It is of particular importance to identify candidates who are suitable for surgery on the basis of clinical indications: a patient with no clear symptoms who is incidentally diagnosed with CM-I without syringomyelia should not be considered for surgery [28] (given that 15–30% of patients with adult Chiari malformation are asymptomatic) [29]. Asymptomatic patients may be followed up and operated upon if and when they become symptomatic; even patients who have been slightly symptomatic (i.e. experiencing a light headache evoked only by Valsalva-like manoeuvres)

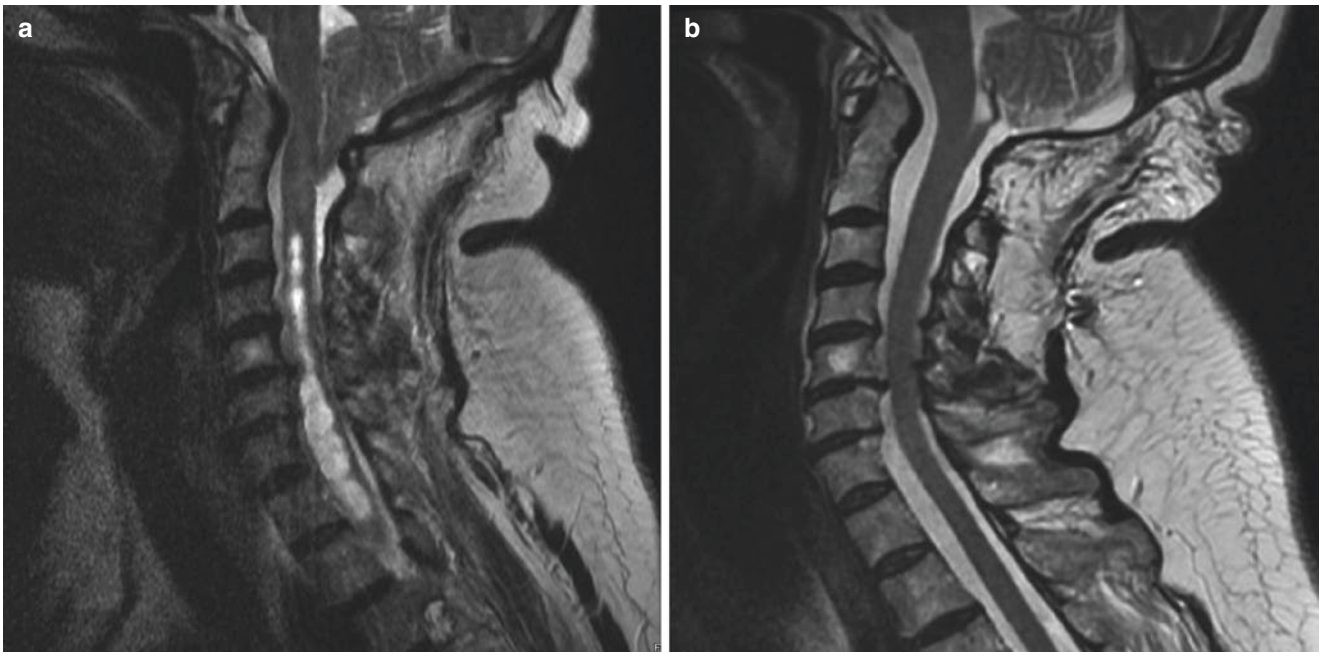

Fig. 3 Magnetic resonance imaging (MRI) in a 33-year-old male patient treated with posterior fossa decompression combined with cerebellar tonsil shrinkage by bipolar coagulation. (**a**) Preoperative T2-weighted sagittal image: Chiari malformation type I with a cervicothoracic syrinx. (**b**) Postoperative T2-weighted sagittal image obtained 3 months after surgery, illustrating a restored cisterna magna and disappearance of the syrinx

and stable for years may be considered for observation, with surgery being indicated in the event of deterioration. Symptomatic patients—meaning those who have an invalidating headache independently of the presence of syringomyelia—should be considered for surgery.

Surgical treatment of CM-I is aimed at expanding the volume of the posterior fossa; several surgical techniques have been used with good outcomes [18, 25].

Generally, postoperative symptomatic improvement has been reported in 61.5–93% of patients in different studies [30–33].

Certainly, the clinical response is the main priority for surgical decision making [34]; the radiological results may not be as clear as the clinical improvement. Though the compression has been relieved by the surgical procedure (enlarging subarachnoid spaces and improving CSF circulation with clinical benefit), the persistence of arachnoid scarring may prevent the nervous structures from regaining their normal position [35]. Moreover, there is no statistically significant correlation between the reduction in the syrinx size and the degree of clinical improvement [32]. Nevertheless, there can be no doubt that a permanent postoperative reduction in the syrinx size is an indicator of sufficient decompression.

One of the largest controversies in the surgical management of CM-I is whether a bony decompression is sufficient to treat the symptoms or whether duraplasty is necessary [36]. As was emphasized in a recent review [37], there is no level I or IIa evidence favouring one approach or the other.

Regarding this vexata quaestio, our experience completely supports duraplasty, in accord with the literature, where lower rates of clinical response and higher rates of reoperation have been reported with dura-sparing procedures [38]. Furthermore, in experienced hands, the rate of CSF-related complications (reported as the main disadvantage of duraplasty) may be considerably outweighed by the clinical improvement provided [39].

In our department, the next step is subarachnoid exploration to open the fourth ventricle and restore CSF flow. This manoeuvre is performed after tonsil shrinkage by bipolar coagulation in order to avoid excessive retraction on the floor of the fourth ventricle. Many authors have advocated arachnoid dissection and eventually cerebellar tonsil resection to provide an opening for the fourth ventricular outlet [40]; this procedure allows CSF circulation to be restored. It is our opinion that intradural exploration is needed, as long as CSF-related complications can be kept low; this is based on the observation that 6–10% of patients have an intradural finding that precludes free flow of CSF out of the fourth ventricle [41].

The microsurgical procedure ends with dural closure, as we firmly believe in the importance of dural suturing to avoid CSF-related complications, such as pseudomeningocele and fistula [15]. There is a plethora of materials available [42] and there is still debate about the optimal dural graft [43, 44], which should provide a watertight suture without inducing arachnoid scarring or stimulating an inflammatory response. Use of autologous tissue requires steps for harvest (extension

of the surgical incision or a second one, with an increased risk of morbidity) but ideally avoids the possibility of reaction to the graft material. Since the commercialization of synthetic materials, progress has been made toward the realization of a resistant, inert, suturable dural substitute. It is our policy to suture lyophilized bovine pericardium (Lyoplant®, B. Braun) in a watertight fashion and test the integrity of the closure with a Valsalva manoeuvre while the patient is in the Trendelenburg position. This, in our experience, minimizes the risk of postoperative CSF leakage and its consequences.

Conclusion

Osteodural decompression of the posterior fossa with cerebellar tonsil shrinkage by cautious bipolar coagulation is an operation with a high rate of success in properly selected patients. The vast majority of treated patients experience clinical improvement and a stable reduction in the size of the syrinx. In this paper, we have presented the technique with a critical analysis of the results in terms of clinical outcome and complications.

Compliance with Ethical Standards No financial support was received for this work.

Competing Interests The author declare that they have no competing interests.

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Treatment of Holocord Syringomyelia–Chiari Complex by Posterior Fossa Decompression and a Syringosubarachnoid Shunt in a Single-Stage Single Approach

Giovanni Raffa, Stefano Maria Priola, Rosaria Viola Abbritti, Antonino Scibilia, Lucia Merlo, and Antonino Germanò

Abstract *Background:* Posterior fossa decompression with expansive duraplasty is the first-line surgical approach for the treatment of symptomatic syringomyelia associated with Chiari malformation. Despite good decompression, the clinical failure rate is reported to be up to 26%. A syringosubarachnoid (S-S) shunt may be used as a secondary option.

Methods: In this paper we describe a single-institution experience of three cases of holocord syringomyelia–Chiari complex treated with foramen magnum decompression, expansive duraplasty and an S-S shunt carried out in a single-stage single approach. Following a standard suboccipital craniectomy, patients were submitted to syrinx fenestration and simultaneous insertion of an S-S shunt through a 1-mm posterior midline myelotomy at the C2 level prior to expansive dural reconstruction.

Results: Postoperative imaging showed immediate reduction of the holocord cavities. Preoperative neurological deficits rapidly improved significantly and were stabilized at follow-up.

Conclusion: In our experience the positioning of the shunt catheter at a high level of the spinal cord (C2) did not add a significant risk of morbidity and obviated the need for a second operation and/or a separate incision in cases of clinical failure. This technique avoided the risk associated with a second surgery and its morbidity, and allowed prompt clinical recovery.

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Keywords Chiari malformation · Holocord syringomyelia Single-stage approach · Syringosubarachnoid shunt

Introduction

Since the first description of syringomyelia–Chiari complex [1], several surgical procedures have been proposed. So far, however, none of these has been able to completely resolve the disease and be proposed as a universally recognized standard [2–11]. In the last decade, surgical strategies have been focused on splitting the outer leaf of the dura mater without duraplasty [12], widening or decompression of the foramen magnum by dural microincisions alone [13] or by excision of the external dura with delamination and widening of the internal layer through longitudinal incisions [14] and, finally, posterior fossa reconstruction with duraplasty [15].

Nevertheless, a syringosubarachnoid (S-S) shunt typically placed at the largest level of the syrinx has also been suggested in patients with a large-sized cavity and central cord syndrome [3, 4, 16, 17].

Despite these numerous treatments aimed at direct and/or indirect resolution of the proposed different pathophysiological mechanisms of this complex disease, in some cases it is still possible to have an unsatisfactory clinical and/or radiological postoperative outcome after posterior fossa decompression (PFD). The current literature reports a clinical failure rate of up to 26% [18] and a radiological failure rate of up to 55% in these cases [19–21]. Therefore, treatment of syringomyelia still requires new surgical strategies to improve the prognosis in these subsets of patients.

PFD and S-S shunting are usually performed separately in different types of patients [3, 4, 16, 17, 22], eventually combined in the same patient but usually separated into two different surgical sessions [4, 9, 17, 23] or performed in a two-stage approach [16, 24].

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The aim of this study is to describe our experience with three cases of holocord syringomyelia associated with Chiari malformation submitted to standard PFD with expansive duraplasty and, simultaneously, placement of a C2 S-S shunt in a single-stage single approach.

Material and Methods

Patient Population

Between September 2010 and October 2011, three patients (two male and one female), ranging in age from 18 to 62 years (mean 46.7 years), were diagnosed with holocord syringomyelia–Chiari complex by magnetic resonance imaging (MRI). We collected and reviewed information on all nosological, preoperative and postoperative clinical symptoms and neurological signs, neuroradiological findings, and length of follow-up for each patient, as reported in Table 1. All patients signed informed consent for surgery and for scientific use of their clinical data.

The patients experienced progressive symptoms over a period ranging from 3 weeks to 2 years before diagnosis, which included a common history of dysesthesia in the lower extremities and extending to the upper ones, unsteady gait, and impairment of fine hand movements. The neurological examination on admission revealed spastic paraparesis with increased reflexes and weakness of the forearm muscles and grasp, with hypesthesia in all cases, increased muscle tone in the upper extremities in two patients and hand deformity (*main en griffe*) in one.

We semiquantitatively assessed the severity of cervical myelopathy by using the modified Japanese Orthopaedic Association (mJOA) scale [25]. The mean preoperative mJOA score was 9.4 (range 9–10).

MRI of the spinal cord showed caudal dislocation of the cerebellar tonsils into the cervical canal below the foramen magnum, ranging from 10 to 17.2 mm (mean 14.1 mm) (Table 1), associated with a large holocord syringomyelia with thinned spinal cord tissue and an obliterated spinal subarachnoid space extending from C2 to T11 level (Fig. 1a). The preoperative mean diameter of the cavity, calculated at the largest level on the axial T2 sequence, was 12.2 mm (range 8.4–18.1 mm) (Fig. 1b; Table 1). The syrinx/canal index proposed by Hida and Iwasaki [3] exceeded 90% in all cases (range 91.3–97.5%) (Table 2).

Table 1 Summary of nosological, neuroradiological and clinical data

Case no.	Patient age (years); sex	Time to diagnosis (months)	Syrinx location	Cerebellar tonsil caudal dislocation (mm)	Main diameter of syrinx (mm)		Symptoms and signs[a]		Follow-up duration (months)
					Preoperative	Postoperative	Preoperative	Postoperative	
#1	18; male	12	C2–T11	17.2	18.1	6.2	*mJOA score 10* I: 2 (right hand *en griffe*) II: 2 III: 1/1/1 (>right side) IV: 3	*mJOA score 16* I: 3 (right hand *en griffe*) II: 4 III: 2/2/2 IV: 3	60
#2	62; male	24	C2–T7	10	8.4	2.8	*mJOA score 9* I: 2 II: 2 III: 1/0/1 IV: 3 Dysphagia and odynophagia	*mJOA score 16* I: 4 II: 3 III: 2/2/2 IV: 3	58
#3	60; female	15	C2–T5	15	10	2.5	*mJOA score 9* I: 2 II: 2 III: 1/1/0 IV: 3	*mJOA score 17* I: 4 II: 4 III: 2/2/2 IV: 3	48

[a]Symptoms and signs according to the modified Japanese Orthopaedic Association (mJOA) scale

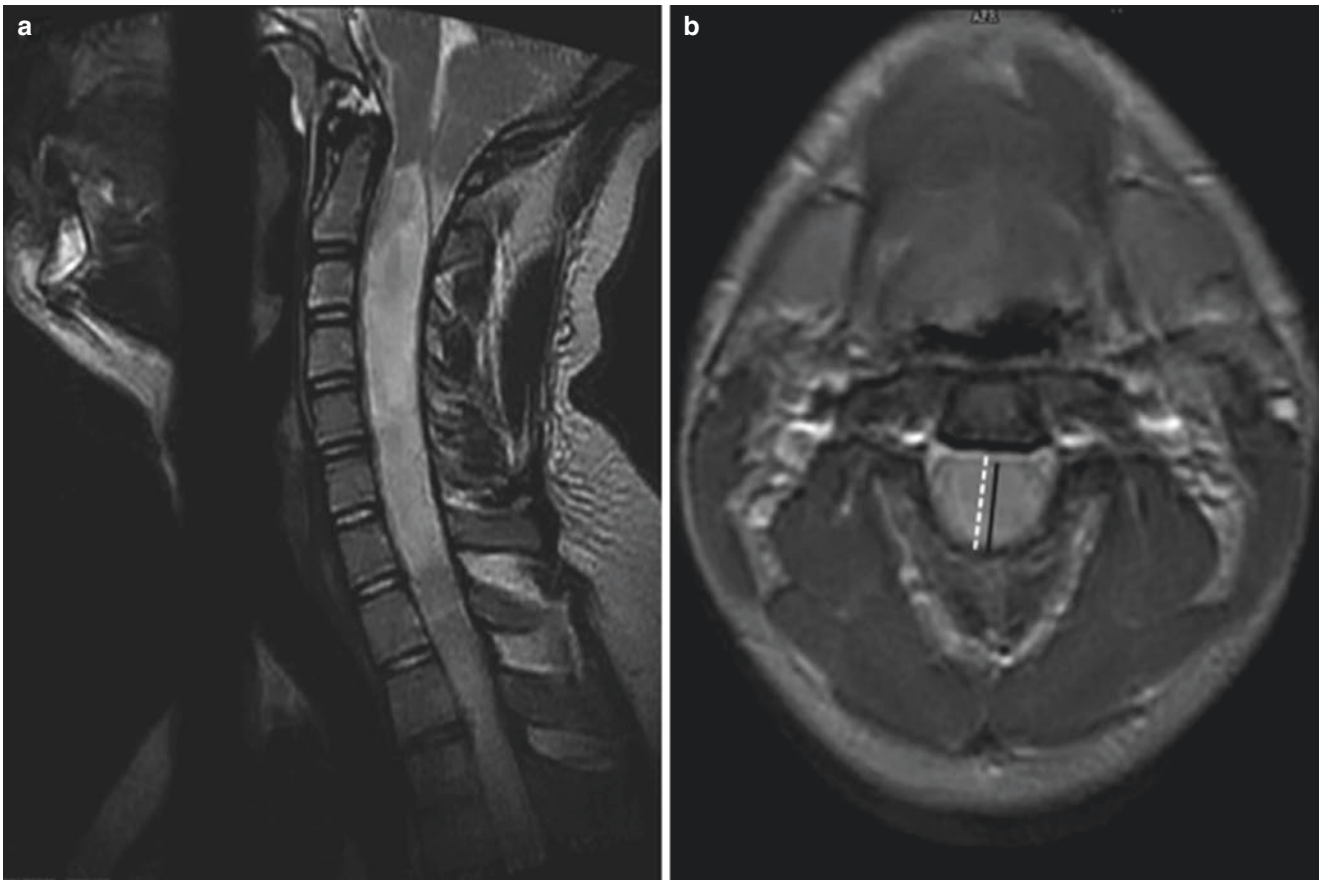

Fig. 1 Preoperative magnetic resonance imaging (MRI) of case #1. (**a**) Sagittal T2-weighted study demonstrating Chiari malformation type I and a large holocord syringomyelia extending from C2 to T11. (**b**) Axial T2-weighted image; the syrinx/canal index at the C2 level exceeds 90%. (The *dashed line* indicates the maximum diameter of the spinal canal; the *solid line* shows the diameter of the syrinx cavity)

Table 2 Pre- and postoperative syrinx/canal indexes [9]

	Syrinx/canal index (%)	
Case no.	Preoperative	Postoperative
#1	97.5	31
#2	91.3	59.7
#3	92.5	42.3

Operative Procedure

Patients were submitted to standard PFD. The operation was carried out in the prone position, with a midline skin incision extending from the external occipital protuberance to the spinous process of C3. A suboccipital craniectomy (3 × 3 cm) with opening of the foramen magnum, removal of the posterior arch of C1 and laminectomy of C2 was performed. The dura mater at the cervico-occipital junction was exposed and an operating microscope was brought in. The dura mater was carefully opened in a Y shape and the arachnoid membrane was incised, showing migration of the cerebellar tonsils into the spinal canal.

At this point, a 1-mm-long posterior C2 myelotomy was performed on the midline, targeted to the syrinx, in a relatively avascular area (Fig. 2a). With use of microscissors, the cavity was fenestrated, obtaining immediate outflow of cerebrospinal fluid (CSF) under pressure. Thereafter, an antibiotic-impregnated silicone catheter with multiple fenestrations at its end [26] (Codman® Bactiseal® EVD Catheter Set; Codman & Shurtleff, Inc., Raynham, MA, USA) was inserted (Fig. 2b). The catheter was placed in a cephalic direction to maintain the physiological CSF outflow, with the caudal part positioned in the subarachnoid space at the C3 level. The distal lateral catheter's outlets were left outside the cavity to facilitate free CSF subarachnoid drainage. The catheter was left in place without any securing [27] to avoid iatrogenic spinal cord fixation. Finally, an expansive reconstructive posterior fossa duraplasty was performed with a dural substitute graft (Gore Preclude® PDX; WL Gore & Associates, Inc., Flagstaff, AZ, USA).

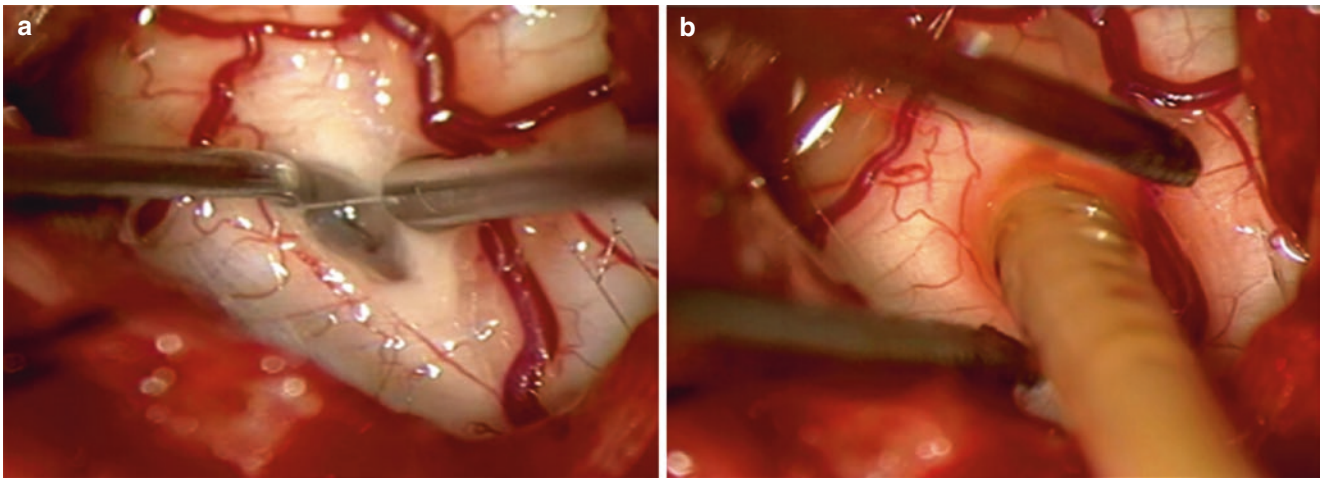

Fig. 2 Intraoperative pictures of case #1. (**a**) Posterior 1-mm midline myelotomy performed in a relatively avascular area at the C2 level following posterior fossa decompression. (**b**) Insertion of a syringosubarachnoid shunt into the syrinx cavity through the myelotomy

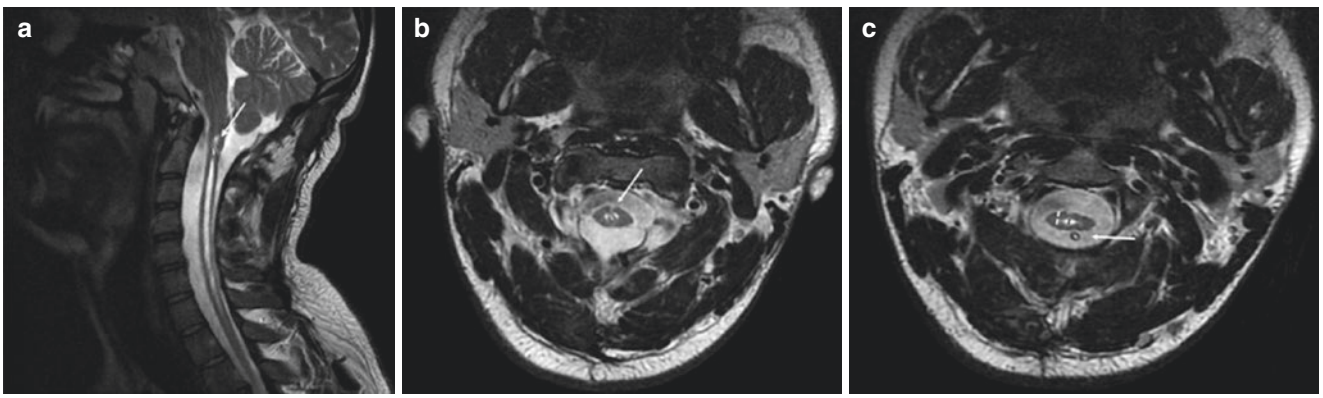

Fig. 3 Postoperative magnetic resonance imaging (MRI) of case #1. (**a**) Sagittal T2-weighted study demonstrating the catheter inside the syrinx (*arrow*) and significant reduction of the holocord cavity. (**b**, **c**) Axial T2-weighted images showing the catheter within the cavity (*arrow*), exiting from the spinal cord at the C2 level and lying freely in the subarachnoid space, and confirming the reduction of the syrinx/canal index (*dashed line*)

Results

Clinical Course and Outcome

The patients had a fast postoperative recovery. Improvement of the preoperative leg weakness and hypesthesia was immediately evident 1 day after surgery. At a mean follow-up of 55 months after surgery (range 48–60 months), all patients had achieved satisfactory recovery of their neurological symptoms. At follow-up the mean mJOA score was 16.4 (range 16–17). In detail, motor weakness had improved by two points in two cases and by one point in the remaining patient, whose right hand was still *en griffe*. Sensitive deficits showed a three-point improvement in the mJOA score in one case and four points in the other two. No bladder dysfunction was present preoperatively or postoperatively in any of the patients. In case #2, the patient experienced dysphagia and odynophagia, but they rapidly resolved in the postoperative period. The patients remained neurologically stable at the follow-up evaluations up to 60 months postoperatively (Table 1).

MRI performed in the postoperative period demonstrated optimal PFD together with a significant reduction in the holocord cavity (Fig. 3a). Axial T2 MRI sequences showed that the maximum diameter of the syrinx cavity at the C2 level was reduced by 75–66.6% (mean 69.1%) to 3.8 mm (range 2.5–6.2 mm) (Fig. 3b, c). Therefore, the syrinx/canal index was reduced to 31% (range 31–59.7%) (Table 2). No surgical complications or damage of the spinal cord at the myelotomy entry point were observed.

Discussion

In this paper we report a retrospective analysis of a single-centre experience of three patients affected by holocord syringomyelia and Chiari malformation treated at our insti-

tution with combined PFD, expansive duraplasty and simultaneous insertion of a C2 S-S shunt. All surgical procedures were carried out in a single-stage single approach. This strategy was performed with the aim of combining the two most widely used surgeries for management of this disease complex. The combination of these techniques allowed effective and less invasive treatment with prompt clinical and radiological recovery.

In recent years we have observed a growing body of literature focused on analysis of the pathophysiological mechanisms of syringomyelia–Chiari complex, suggesting different surgical procedures [1]. PFD is one of the most widely used surgical procedures for treatment of this disease complex. Nevertheless, there is still a significant proportion of patients who fail to improve, either clinically or radiologically, after PFD.

Over the years, several surgical strategies have been proposed to improve the clinical and neuroradiological results of PFD: removal [12] or widening by transverse microincisions [13] of the outer layer of the dura mater; opening of both dural layers, leaving the arachnoid intact [14]; and posterior fossa reconstruction with expansive dural grafting [15].

Although none of these approaches is free of complications [12, 28], PFD with its variants remains the most widely performed surgical approach for syringomyelia–Chiari complex. Therefore, the positioning of an S-S shunt is still considered a secondary procedure that should be performed in the event of failure of the former procedure [29].

There is evidence that in some cases, patients with small syringes could experience progression despite craniovertebral decompression and could require a second surgery for the syrinx shunt [16]. Conversely, in some cases the syrinx resolves after shunting alone, even if PFD is not performed and the CSF flow obstruction at the level of the foramen magnum has not been removed [10]. Subsequently, to ensure postoperative relief of symptoms, PFD and S-S shunting are often both necessary to achieve a satisfactory outcome in many cases.

In the literature, various types of shunts have been described, including S-S [3, 4, 7, 9], syringoperitoneal [5], syringopleural [11] and thecoperitoneal [10] shunting. These procedures are associated with a good postoperative outcome, at least in the early postoperative period. Nevertheless, there are some reports of delayed complications and disadvantages, such as shunt malfunction, slippage and cord injury [4, 30]. Moreover, it has been reported that many patients treated using shunt procedures alone have required a subsequent revision or PFD [29].

In the light of such evidence, PFD and S-S shunts should be considered two complementary procedures, acting on two different pathophysiological mechanisms: compression at the craniovertebral junction and alteration of the normal CSF circulation. Actually, they are usually performed separately in different subset of patients [3, 4, 16, 17, 22]. In many cases, they are combined but done during two separate surgical sessions [4, 9, 17, 23]. In a few cases, they are performed during the same operation [8] or in a two-stage approach [16, 24]. The shunt is usually inserted at the largest level of the syrinx [3, 4, 9, 16, 17, 23, 24, 30, 31], usually at the lower cervical or midthoracic level. The choice of the catheter insertion point usually corresponds to the largest portion of the syrinx, where its insertion is considered to ensure better decompression. This has been reported even for multilocular syringes [9]. Nevertheless, in the literature there is also a lack of consensus regarding myelotomy in terms of its exact location (dorsal root entry [3, 4] versus midline entry [16]) and its length (1 cm [8] versus 2 mm [17]), and the direction of the shunt (craniocaudal [17] versus caudocranial [31]).

Considering the conflicting literature reports, it is actually difficult to prove the usefulness of any therapeutic procedure in decreasing intramedullary tension. Currently there are no objective criteria and no standard radiological parameters to suggest which technique will better resolve a syrinx when it is associated with Chiari malformation. The clinical and radiological recurrence rate of syringes following PFD with duraplasty alone in patients with syringomyelia–Chiari complex has been reported to be up to 26% [18], with a long-term radiological failure rate ranging between 6% [18] and 55% [19–21].

In the light of this evidence, there is still a need for definition of an effective strategy for treatment of syringomyelia–Chiari complex, aimed at improving the postoperative outcome. In the literature, there are only a few reports describing the combination of PFD with an S-S shunt, or insertion of an S-S shunt following failure of initial PFD [8, 9, 16, 22, 24]. In those studies it was concluded that the combination of PFD and an S-S shunt was superior to PFD or shunt placement alone: resolution of the syrinx was achieved in a higher percentage of patients, without recurrences [8, 9, 16, 22, 24]. In the present study, we documented that simultaneous PFD and S-S shunt insertion could be performed in a single-stage approach and was associated with satisfactory short- and long-term postoperative outcomes. Moreover, the strength of this strategy is that it could avoid the need for subsequent surgeries after treatment failure in cases in which PFD or an S-S shunt alone is not sufficient to provide satisfactory relief from symptoms.

Conclusion

In this paper we have presented a different surgical strategy to treat holocord syringomyelia–Chiari complex in a single-stage procedure. The positioning of the shunt catheter at a high level of the spinal cord (C2) did not add a significant

risk of morbidity. This technique, which links two already standardized procedures in a single-stage single approach, avoided the risk associated with a second surgery and its morbidity, and allowed prompt clinical recovery.

We recognize that a retrospective single-institution experience does not allow us to draw definitive conclusions. However, our report supports the existing evidence that the combination of both techniques performed in the same surgical approach is safe, is feasible and contributes to achieving faster recovery times without adding a significant risk of morbidity related to both the site of catheter insertion and a second surgery, in comparison with a standard two-stage approach.

Further studies are required to evaluate the long-term efficacy of this strategy.

Compliance with Ethical Standards No financial support was received for this work.

Competing Interests The authors declare that they have no competing interests.

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Surgical Treatment of Chiari Malformation in Adults: Comparison of Surgical Techniques Described in the Literature and Our Experience

L. Lavorato, A. Spallone, and M. Visocchi

Keywords Chiari · Malformation · Surgery · Techniques

Introduction

Chiari malformations (CMs) constitute a group of different clinicopathological entities with varying aetiology, pathophysiology and clinical features. They represent varying degrees of herniation through the foramen magnum. Professor Hans Chiari (1851–1916) devised a four-tier classification system for these entities [1, 2]. His initial description was based on the findings of these malformations in 40 autopsies he performed in Prague [1, 2].

CM type I (CM-I), initially described in 1891, constitutes a syndrome where the cerebellar tonsils descend through the foramen magnum for at least 3–5 mm (Fig. 1). It may be occasionally associated with an elongated fourth ventricle. Frequently, it is a disorder of mesodermal origin, but neuroectodermal and acquired forms have also been reported [3].

CM type II (CM-II), described in 1896, consists of descent of the cerebellar vermis, the fourth ventricle and the lower brainstem, and almost always is seen in conjunction with a myelomeningocele. It is the most common form of CM. Hydrocephalus always coexists with CM-II, and it may also be associated with spina bifida and other abnormalities. The syndrome has a neuroectodermal origin [3]. CM-II remains the leading cause of death in patients with a treated myelomeningocele [4, 5].

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CM type III (CM-III) is based on the description of a single case, in which there was a large dermal sac in the occipital region, containing herniated cerebellum. CM-III is characterized by displacement of the medulla and herniation of part of the cerebellum into a meningocele. Sometimes part of the hindbrain is also herniated. Hydrocephalus is present in 50% of these cases and is always of obstructive aetiology, with an aqueductal stenosis or an associated Dandy–Walker malformation. CM-III is a neuroectodermal malformation [3].

CM type IV (CM-IV) represents a pathological entity of cerebellar hypoplasia. It is the least frequent form of CM and is characterized by hypoplasia or aplasia of the cerebellar hemispheres and morphological alterations of the pons. Hydrocephalus is quite rare among patients with CM-IV. It is a disorder of neuroectodermal origin [3].

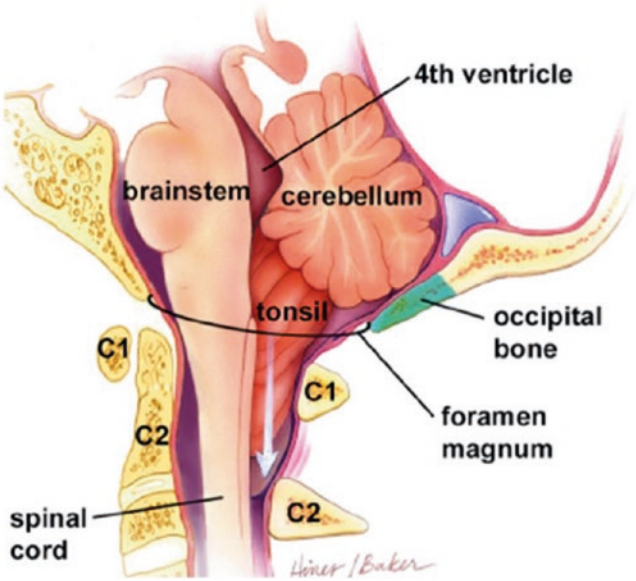

Fig. 1 Chiari malformation (CM)

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Many researchers have described another form, CM type 0, which is characterized by an alteration in cerebrospinal fluid (CSF) hydrodynamics at the anatomical level of the foramen magnum. It is a pathological entity characterized by some degree of posterior tilt of the pons and the medulla, with displacement of the medulla oblongata, a low tip of the obex and a normal position of the cerebellar tonsils. Patients with this condition also demonstrate syringomyelia, either without tonsil herniation or with only mild tonsil herniation [6, 7]. Mottolese et al., like many other neurosurgeons, have expressed doubts regarding the existence of this type of CM [8].

Caudal displacement of the brainstem with cerebellar tonsil ectopia in the absence of spina bifida has been considered a separate form of Chiari malformation and is called CM type 1.5 [9].

The pathogenesis of CM has been extensively studied by several researchers, who have developed various explanatory theories. Examples of these theories are developmental arrest due to myeloschisis [10–13]; the overgrowth theory, which suggests enlargement of the neural plate prior to neurulation, thus preventing fusion of the neural folds [14]; the neuroschisis theory [15, 16]; the neuroectodermal–mesodermal spatial dyssynchrony theory [11]; and the traction theory [17]. In regard to the pathogenetic mechanisms implicated in the development of CM syringomyelia, which frequently coexists with CM-I, Gardner, in 1965, developed the hydrodynamic theory, which postulates that lack of perforation of the rhombencephalic roof and persistence of a patent communication between the fourth ventricle and the central canal of the spinal cord could lead to syrinx development [18]. Oldfield and colleagues reported another theory regarding the creation of a syringomyelic cavity, based on cine magnetic resonance imaging (MRI) and intraoperative ultrasonographic findings [19]. According to this theory, CM-associated anomalies could induce a piston-like motion, thus affecting the cerebellar tonsils and producing a systolic wave in the CSF flow, which acts on the spinal cord and induces CSF leakage through the interstitial and perivascular spaces [19]. Nishikawa et al. studied posterior fossa morphology in 30 sporadic cases of CM-I anomalies and 50 control cases, and they reported underdevelopment of the occipital bone along with compression of the cerebellum and the brainstem, resulting in caudal herniation through the foramen magnum [20]. In addition, several other authors have reported development of acquired CM-I, secondary to prolapse of the cerebellar tonsils through the foramen magnum in patients with previously inserted ventriculoperitoneal or lumbar–peritoneal shunts [21, 22]. Occipital hypoplasia, overgrowth of posterior fossa anatomical elements, decreased osseous posterior fossa volume, underdevelopment of the occipital bone, and the presence of posterior fossa vascular malformations, hydrocephalus, posterior fossa masses, lumbar–peritoneal shunts, and craniofacial and posterior cranial base malformations may contribute to development of CM.

The aim of our study was to review the literature regarding surgical treatment of CM, with evaluation of different surgical approaches, their modifications and our experience with our techniques.

Materials and Methods

A total of 20 patients underwent corrective surgery for CM from 1990 to 2015. All patients underwent craniectomy and laminectomy (C1) and dura opening without arachnoid opening. All patients were noted to have undergone a thorough clinical examination and MRI prior to surgery. All patients received an explanation of the advantages and possible complications of the procedure before giving their written consent for the procedure.

Of the 20 patients, 11 had CM-I and the other nine had CM-II (Fig. 2). The average age of the patients was 31 years (range 27–35 years). All of the patients presented with a headache, and their other symptoms included neck pain, hemiparesis, nystagmus, dysmetria and other cerebellar signs. In two cases, CM was associated with a spinal deformity and in four cases it was associated with syringomyelia.

Surgery was performed in all patients in the prone position with a head elevation of 20–30°. The head was held in a Mayfield three-pin frame. The patient's neck was flexed at the craniocervical junction and extended at the

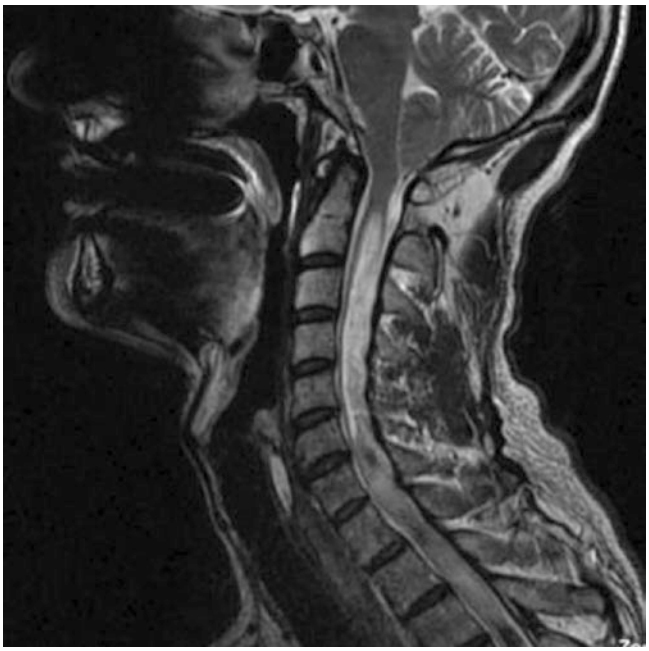

Fig. 2 Preoperative T2-weighted magnetic resonance imaging (MRI)

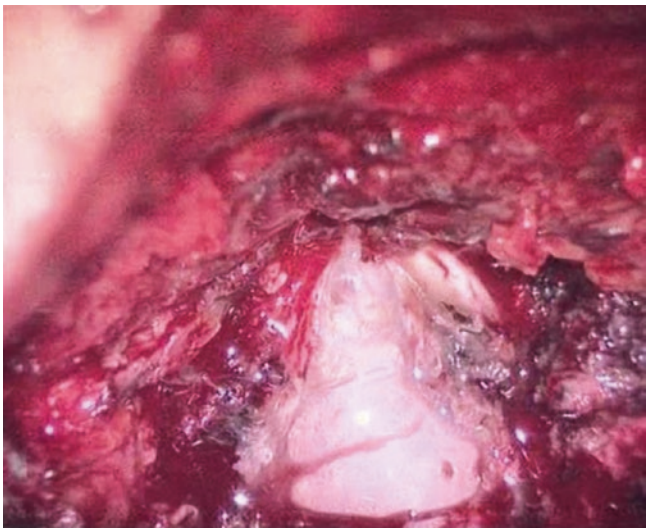

Fig. 3 Intraoperative view: the dura mater has been opened and the arachnoid has been conserved

cervicothoracic junction for easy and safe access to the foramen magnum. Surgery was performed via a midline incision, which extended from 2 cm to 3 cm above the inion to the second cervical vertebra. The inion, the midline of the occiput down to the foramen magnum, the posterior arch of C1 and the lamina of C2 were exposed. Suboccipital craniectomy was carried out from the inferior nuchal line to the posterolateral rim of the foramen magnum, and C1 laminectomy alone was done to expose the dura. The dura was opened without opening the arachnoid (Fig. 3).

After were no placed dural patch. Patients were closely followed up over the years to detect any recurrence of symptoms at an early stage. In all patients, MRI was performed after surgery (Fig. 4).

Results

The data of the 20 patients who underwent corrective surgery for CM were retrieved and analysed.

A CSF leak was noted in five patients (25%). The leak either resolved spontaneously and not needed a shunt procedure.

Neck and shoulder pain, headache and paraesthesia were the common presenting complaints in all patients. These symptoms improved in all patients after the surgery.

During follow-up, we observed an early beneficial outcome and stabilization of the disease in 18 patients (90%). Two (10%) showed late deterioration. The two patients diagnosed with syringomyelia both had a sustained improvement. One patient died from clinical problems not related to CM.

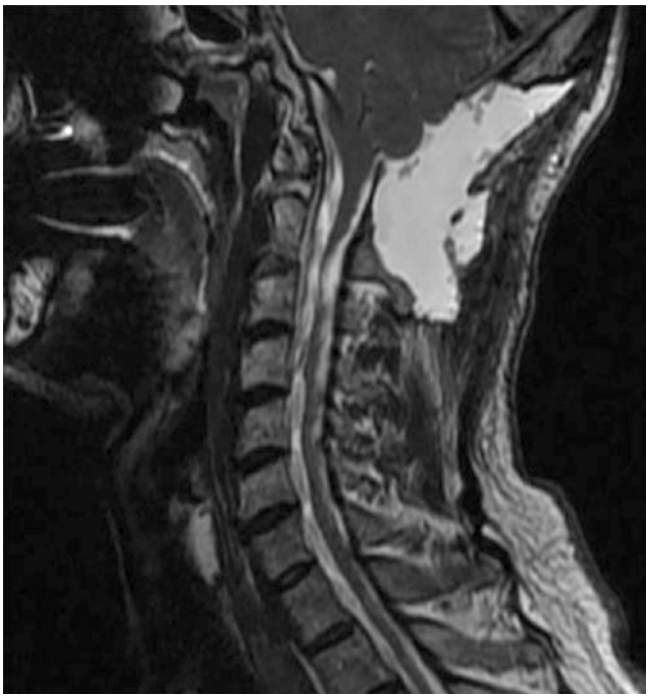

Fig. 4 Postoperative T2-weighted magnetic resonance imaging (MRI): the intrafascial cerebrospinal fluid (CSF) leakage has resolved spontaneously

Discussion of the Surgical Techniques Described in the Literature

There is evidence that underdevelopment of the mesodermal occipital bone is responsible for the small volume of the posterior fossa found in patients with CM, resulting in abnormal flow of CSF at the foramen magnum [11, 12]. The logical target is to extrapolate enlargement of the dimensions of the posterior fossa and re-establishment of CSF flow at the craniocervical junction to improvement of symptoms. Badie et al. [9] demonstrated that CM-I patients with a decreased posterior fossa volume responded better to surgical decompression than those with a normal posterior fossa volume [9]. The outcome is difficult to predict and is variable despite perceived adequate decompression. Improvement or resolution of symptoms has been reported in large series in the literature [10, 14].

The optimal surgical treatment for symptomatic CM remains controversial. The target of surgery is to restore normal CSF dynamics at the craniocervical junction. However, to achieve this surgical target, many different operative techniques have been recommended. Establishment of good CSF flow from the fourth ventricle to the cervical subarachnoid space is the aim of hindbrain decompression. Williams described the traditional technique of creating an artificially enlarged cisterna magna to provide a reservoir for spinal pressure surges [13, 15]. His technique included suturing the

dural edges to the divided suboccipital muscles with wide arachnoid dissection [15]. A modification of this technique includes duraplasty to avoid CSF leakage and pseudomeningocele formation, with dura splitting (leaving the inner layer of the dura and arachnoid intact) or bone-only decompression to avoid CSF exposure and subsequent arachnoid adhesion formation. The extent of decompression necessary to achieve normal CSF dynamics varies between patients and is not dependent only on the level of tonsillar descent, the presence of a syrinx and the resulting symptomatology. Even when good CSF flow is thought to have been achieved by surgery, with the extent of decompression tailored to the individual patient's circumstances, a significant proportion of patients will have no improvement in their symptoms. Bone-only decompression provides a minimally invasive method of restoring CSF circulation. Patients undergoing bone-only decompression, many of whom have previously suffered from headaches, have experienced resolution or improvement of their symptoms, whereas poor outcomes have been seen in patients with dysaesthetic pain or ataxia. This suggests that patients presenting with more than a simple classical Chiari-type headache should undergo a formal dural-opening procedure. In addition, the rate of recurrence of symptoms is significantly higher after bony decompression than after dural opening. Predicting which patients' symptomatology will consistently improve with hindbrain decompression remains an elusive goal. McGirt et al. described the use of cine phase contrast MRI of CSF flow at the foramen magnum as a predictor of which patients are likely to respond to hindbrain decompression, and showed that complete obstruction of CSF flow demonstrated preoperatively was an independent predictor of long-term symptom resolution, regardless of the degree of tonsillar ectopia [17]. In addition, patients with syringomyelia or generalized headaches were at increased risk of symptom recurrence. There was a complication rate of up to 42% in patients where the dura was opened and a 10% complication rate in those where the dura was left intact [18–20]. Indeed, postoperative pyogenic meningitis occurred only in the group treated with dural opening without duraplasty. Several deaths have been reported after posterior fossa decompression for CM, with a reported mortality rate of 3% [19, 21]. Many complications of hindbrain decompression are related to CSF exposure to blood, muscle and cellular debris, resulting in adhesive arachnoiditis and causing impedance of CSF flow. Although bone-only decompression may avoid these complications, the outcome in relation to symptom improvement is poor, with 25% of patients showing no change in their headaches and 60% showing no change in arm dysaesthesia. Some authors advocate a dura-splitting approach to avoid CSF exposure but provide better decompression than removing only bone [22]. Williams noted that signs related to a syringomyelic cavity, such as muscle wasting and sensory loss, seldom improve following hindbrain decompression. The longer the clinical history, the worse the prognosis. Some authors suggest that patients with symptoms lasting longer than 2 years have a worse prognosis [23]. This is particularly true of syringomyelia, where irreversible neurological damage may persist after surgery even when resolution of the syrinx is seen. This may explain the poor outcome of patients with dysaesthetic pain or weakness. Studies have reported that posterior fossa syndromes such as headache, ataxia and drop attacks were more likely to improve following hindbrain decompression. This finding was demonstrated in 70–100% of cases [18, 19, 23, 24]. Radiological resolution or improvement of the syrinx was seen in 88% of cases. Improvement or stabilization of syringomyelia occurred in 65–88% of cases following hindbrain decompression [17, 21, 25, 26]. With regard to scoliosis, a lot of patients showed resolution or stabilization of the curvature following hindbrain decompression. Up to 71% of children showed improvement [18, 27, 28].

Conclusion

Chiari malformation is being increasingly diagnosed but remains little understood in terms of its aetiology and symptomatology. Restoration of cerebrospinal fluid flow dynamics at the foramen magnum by surgical decompression, demonstrated both at operation and on postoperative imaging, does not consistently result in resolution of symptoms in all patients. Identification of predictors of a successful outcome following decompression, coupled with early intervention and an appropriate choice of procedure, may result in improved outcomes. Many studies have suggested that bone-only decompression should be reserved for patients with isolated headache symptoms.

In our experience the surgical technique we have adopted is a good strategy to treat CM, the results are good, and the improvements observed during follow-up are encouraging, arguing against a greater surgical commitment. The operation used in these patients poses significant technical difficulties related to the need to keep the arachnoid intact while opening the dura mater, but this method has the advantage of preventing complications related to contamination of non-bacterial liquor that may occur with some frequency with speeches in which it is planned to open the dura mater.

As noted in the present experience, improvement of syringomyelic cavities possibly present is constant.

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Surgery: Decompression—Lateral Approaches

The High Cervical Anterolateral Retropharyngeal Approach

Nabeel S. Alshafai and V. R. N. Gunness

Abstract The first high cervical anterolateral retropharyngeal (HCALR) approach was reported by Stevenson et al. for a clivus chordoma in 1966. Anterior approaches to the spine have often been developed in response to problems presented by tuberculous spondylitis. This approach is indicated in anterior high cervical spine cases such as tumour resection, abscess drainage, atlantoaxial subluxation; decompression and stabilization. To our knowledge, only 21 papers in the literature have mentioned this approach. Its main advantage over posterior approaches is easy positioning and minimal need for soft tissue dissection. The HCALR approach provides wide exposure (of the anterior upper cervical spine, lower clivus and brainstem region) and feasibility for instrumentation. The limited space in which important neurovascular and visceral structures course and overlap contributes to the complexity of the anatomy. Navigating this intricate anatomy is essential for the safety of this approach and has been a drawback for utilization of the retropharyngeal corridor. This approach is one of the safest and most effective methods available to access the craniocervical junction. The benefits clearly outweigh the risks and complications.

Keywords Craniocervical junction · Anterior approach · Anatomical corridors · Minimally invasive

Introduction

This paper aims to describe the anterior approach to the high cervical spine through the anatomical corridor of the retropharyngeal space. Different authors have used different terminology for this approach. Terms such as 'anterolateral' [1], 'submandibular' [1, 2], 'anterior high cervical' [3, 4] and 'retropharyngeal prevascular' [5, 6] have been used to describe this approach between the carotid sheath lateral to the high cervical spine and the pharyngeal constrictor muscles medial to it. Anterior approaches have principally been developed for tuberculous spondylitis. In 1956, Hodgson and Stock reported their experiences with anterior decompressive surgeries [7]. Cloward [8] and Robinson and Smith [9] have been credited with implementing the anterior approach to the cervical spine for management of disc herniation.

The retropharygeal prevascular approach, which was described by McAfee et al. [5], used the same fascial plane that was illustrated by Southwick and Robinson [10].

In a paper published by Soo-An Park et al. in 2013, they tried to develop a suitable anterior cervical approach to the C2–C3 level [6].

Materials and Methods

Indications for and Importance of This Procedure

The anterior retropharyngeal approach provides wide exposure of the anterior upper cervical spine, lower clivus and brainstem [11, 12], with feasibility for instrumentation [1]. In comparison with other anterior approaches, the retropharyngeal route offers lower rates of infection (with no contamination by nasopharyngeal flora) and lower morbidity by forgoing mandibular osteotomy or tongue division [13]. This approach is indicated in anterior high cervical spine cases of tumour resection, abscess drainage in infections, fixed atlantoaxial subluxation; decompression and stabilization, and arthrodesis minus the necessity of second-stage posterior stabilization [13]. In addition to ventromedial skull base

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lesions [14], C1–C4 involvement in patients with short necks as a part of Klippel–Feil syndrome or other pathologies [3].

A systematic search was performed on PubMed, without a temporal limitation, to identify all clinical reports in which the high anterior cervical approach was mentioned. The following search strategy was used '[(high cervical) OR (high anterior cervical) OR (submandibular) OR (anterolateral) OR (retropharyngeal prevascular)]'. The search was performed independently in parallel by one junior neurosurgery resident and one medical student.

The inclusion criteria included any cases of the aforementioned search terms.

Results

We found 28 papers that matched the search terms, but only 21 papers matched our inclusion criteria. We grouped the sample into four categories: 114 cadavers, seven case reports, 584 patients and one vertebrae (Fig. 1).

Discussion

Several studies have shown the usefulness of the high cervical anterolateral retropharyngeal (HCALR) approach in providing wide exposure from the clivus and the anterior rim of the foramen magnum to the rostral cervical spine up to C4 [5, 15].

The aims of treatment of upper cervical spine lesions are to decompress nervous structures and provide stability.

A superior extension of the anterior approach was used by Smith and Robinson to expose the atlas to the subaxial cervical spine. They did not dissect the carotid sheath or expose the vertebral artery [5].

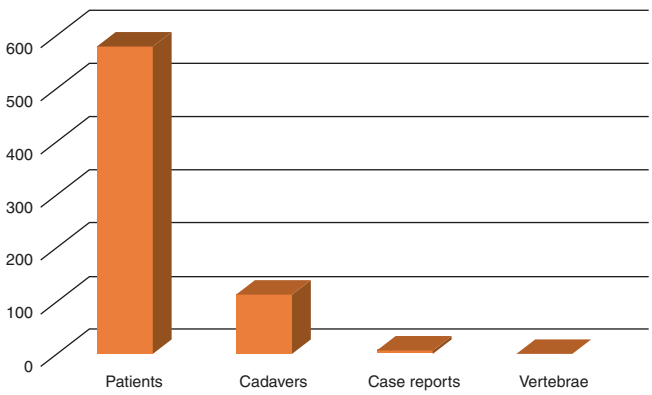

Fig. 1 Numbers of patients, cadavers, case reports and vertebrae

This prevascular extraoral retropharyngeal approach gives good and safe access in the region of the upper cervical spine. Furthermore, anterior osteosynthetic stabilization, including C2–C3, can be performed using this technique, with low morbidity [16].

One of the main advantages of this approach is that it can be performed using traditional instrumentation, and surgeons familiar with the anterior cervical spine will not find it too difficult.

However, this approach can be challenging for the following reasons:

1. Broad dissection of the nerves and vascular structures is required, which is a weakness of this approach.
2. This approach carries a small risk of injury to the marginal mandibular, hypoglossal and superior laryngeal nerves. These nerves cross the operative field and may look like blood vessels. They can be cauterized or ligated.
3. The HCALR approach may cause hypoglossal nerve palsy and superior laryngeal nerve palsy because of excessive retraction [17].

The common postoperative complications due to this approach are dysphagia and dysphonia, which are often transitory, lasting for only a few days, but they can also last for several months.

The dissection is blunt through soft tissues, and finding a plane is of utmost importance to allow us to reach the spine while avoiding the likelihood of nerve damage [17].

Laus et al. published a review of ten cases of anterior extraoral surgery of the upper cervical spine. The authors reported four cases of transient paralysis of the mandibular branch of the facial nerve [18]. Park et al. did not report any paralysis of the facial nerve in their series [17].

The possibility of performing this procedure especially in patients with a short neck, patients with Klippel–Feil syndrome or patients suffering from upper cervical disc herniation—for example, C2–C3 or C3–C4—was reported by Behari et al.

The HCALR approach gives broad exposure to the spine. It avoids the potential contamination that can accompany the transoral approach. It also allows us to do a simultaneous fusion with instrumentation [19].

There are more complications at the C2–C3 level than at any other level. This approach has the highest potential for morbidity, which includes voice abnormalities and aspiration pneumonia due to injury to the superior laryngeal nerve [20].

In the literature, several other risks with this approach have been mentioned, such as injury to the submandibular gland, the recurrent laryngeal nerve and the content of the carotid sheath [17].

The procedural steps in the exposure and the structures at risk during the procedure are listed in Table 1.

Table 1 Procedural steps and structures at risk during the procedure

Step	Structures at risk
Step 1: Submandibular incision from the midline to the anterior border of the sternocleidomastoid	Marginal mandibular branch of the facial nerve
Step 2: Subplatysmal dissection	Marginal mandibular branch of the facial nerve
Step 3: Submandibular gland dissection and retraction	Facial artery and vein, and marginal mandibular branch of the facial nerve
Step 4: Mobilization of the fibrous sling of the digastric muscle	Hypoglossal nerve
Step 5: Tracing of the hypoglossal nerve and superior retraction	Lingual artery and ranine venous plexus[a]
Step 6: Tracing of the internal superior laryngeal nerve and inferior retraction	Superior laryngeal artery
Step 7: Blunt dissection of the facial plane of the retropharyngeal space	Vein draining the retropharyngeal space
Step 8: Lateral retraction of the sternocleidomastoid and dissection around the carotid sheath	Hypoglossal nerve (at the level of bifurcation or superior to it)
Step 9: Palpation of the midline and longitudinal opening of the prevertebral fascia	Ipsilateral vertebral artery (during dissection); it should also be noted that the longus colli muscle is attached until C1

[a]Also called the sublingual vein

Conclusion

The high cervical anterolateral retropharyngeal (HCALR) approach should be learned by every neurosurgeon interested in spine practice because it provides broad bilateral exposure and a safe approach. It avoids the oropharyngeal cavity and at the same time gives the surgeon the option to do an arthrodesis and instrumentation during the primary surgical procedure without the need for a second posterior stage.

With detailed knowledge of the anatomy of the submandibular triangle and the retropharyngeal space, this approach can be easily mastered and added to the neurosurgeon's armamentarium.

The superomedial trajectory of the approach makes it difficult for the surgeon to maintain orientation of the midline during bony drilling. Both neuronavigation and neuromonitoring are helpful to avoid nerve damage.

The HCALR approach should be used strictly for midline lesions restricted to the craniocervical junction.

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Compression Syndromes of the Vertebral Artery at the Craniocervical Junction

Jan Frederick Cornelius, Raoul Pop, Marco Fricia, Bernard George, and Salvatore Chibbaro

Abstract Compression syndromes of the vertebral artery that occur at the craniocervical junction are extremely rare causes of haemodynamic insufficiency of the posterior cerebral circulation. The aetiology of the compression syndrome may be a malformation, trauma, tumour, infection or degenerative pathology. This may lead to dynamic vertebral artery occlusion where the vessel courses around the atlas and the axis—the so-called V3 segment. This in turn may result in insufficient collateral flow to the posterior fossa. The clinical picture is a vertebrobasilar insufficiency syndrome of variable expression ranging from vertigo to posterior fossa stroke. The typical clinical presentation is syncope occurring during rotation of the head, also known as 'bow hunter's syndrome'. The workup is based on dynamic angiography and computed tomography angiography. The treatment of choice is surgical vascular decompression, resulting in a good clinical outcome. However, in some instances, atlantoaxial fusion may be indicated. Alternatively, conservative and endovascular options have to be considered in inoperable patients.

Keywords Bow hunter's syndrome · Extrinsic compression of the vertebral artery · Craniocervical junction · Surgical management · Vertebrobasilar insufficiency and stroke

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Introduction

Compression syndromes of the vertebral artery can occur at different levels: on the segment between the subclavian artery and the cervical spine (the V1 segment), on the segment running through the cervical transverse foramina (the V2 segment), at the loop around the atlantoaxial complex reaching the posterior fossa dura (the V3 segment) and, finally, on the segment after the dura has been penetrated up to the vertebrobasilar junction (the V4 segment) [1, 2]. In this chapter we focus on compression syndromes of the vertebral artery at the craniocervical junction, i.e. the so-called V3 segment (Fig. 1). The associated clinical presentation has been described as bow hunter's syndrome (BHS).

Patients and Methods

As this disease is extremely rare, the medical records and radiological images of eight patients with a vertebral artery compression syndrome at the craniocervical junction, managed over the last 14 years at three different centres, were collected and retrospectively analysed. In the present analysis, three new patients were able to be added to the previous case series of five patients reported by our group in 2012 [3]. The study was conducted according to the Ethical Principles for Medical Research Involving Human Subjects, stated in the 2004 revision of the Declaration of Helsinki (the most recent version of the document at the time when the present study began). The patients signed standard informed consent forms approved by the respective institutional research boards at each university hospital (Strasbourg, Lariboisiere and Dusseldorf). Furthermore, a thorough review of the pertinent English literature was performed on Medline to explore the role of surgery versus both endovascular and conservative management.

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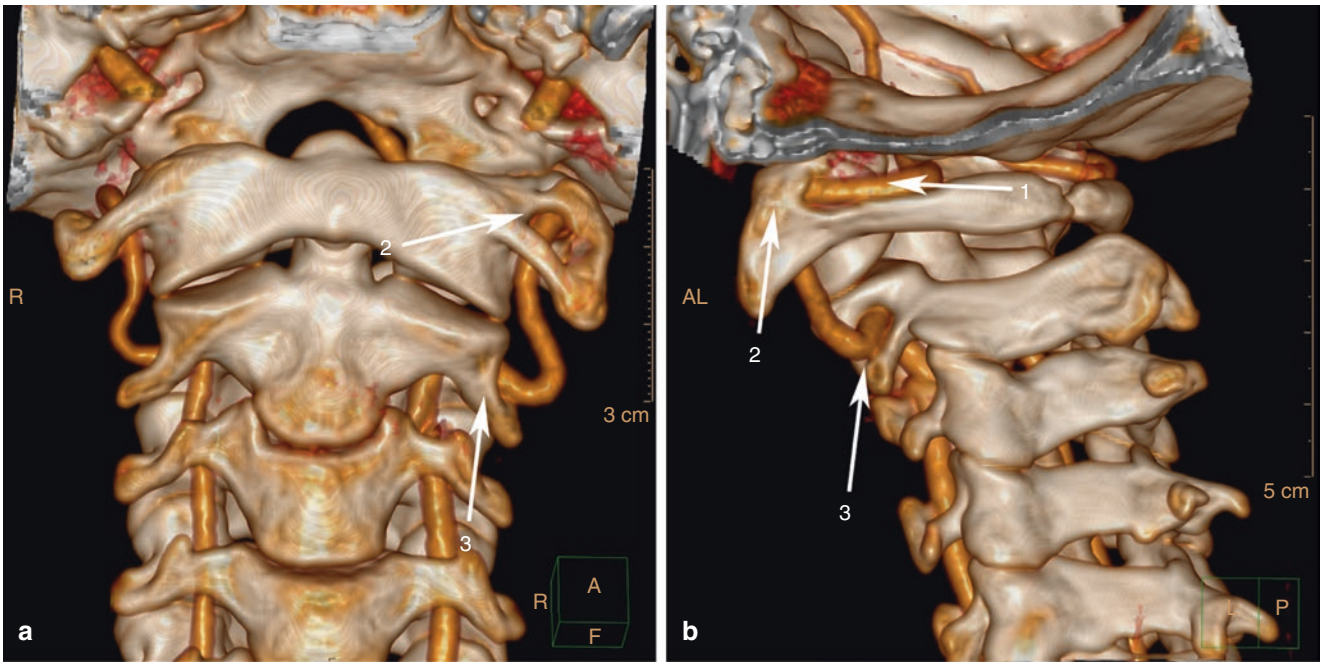

Fig. 1 Possible sites of dynamic vertebral artery compression. Computed tomography (CT) angiogram of the craniocervical junction, showing volume-rendered reconstructions in (**a**) the anterior view and (**b**) the lateral view. The three main sites where the vertebral artery can be compressed during contralateral head rotation are posterior to the lateral mass of the atlas (1) and within the foramen transversarium of the atlas (2) and axis (3)

Results

Of the eight patients studied, six were men and two were women, with a mean age of 24.4 years (ranging from 8 to 46 years). Clinically the most frequent symptoms and signs were vertigo and visual problems occurring during rotation of the head. Extreme head movements—especially rotation and extension—caused symptoms such as fainting, falling and even a posterior circulation infarction in a young boy. The detailed patient demographic characteristics and clinical features are summarized in Table 1. To verify a clinical suspicion of BHS, the patients regularly underwent dynamic digital subtraction angiography (DSA). The patients were asked to actively perform head movements in all directions (left and right rotation, left and right lateral inclination, and inclination and reclination in the sagittal plane) to detect the exact location of the compression and determine the degree of stenosis (Figs. 2 and 3). Furthermore, collateral flow from the anterior circulation could be examined during the examination. The angiograms showed that in all patients in the present series, the vertebral artery was being compressed at the loop around the axis and atlas (the V3 segment). The artery that caused symptoms upon occlusion was always the dominant vessel. Interestingly, the posterior communicating arteries were generally either hypo- or aplastic and the collateral flow from the anterior circulation was insufficient.

Seven patients were treated via an anterolateral cervical approach, which allowed direct decompression of the vertebral artery (Figs. 2 and 3). In one patient (patient no. 2) a posterior approach with decompression and craniocervical osteosynthesis had to be performed because the craniovertebral junction was judged to be unstable. In one patient (patient no. 3) the anterolateral cervical approach had to be performed bilaterally because he presented with a complex malformation. The mean follow-up period was 96 months (ranging from 48 to 144 months). Clinically all patients showed improvements in comparison with their preoperative clinical status; in particular, they became free of syncope. Postoperative dynamic DSA and/or computed tomography (CT) angiography (CTA) were performed to verify the restored vertebral artery flow in the formerly compressing head position (Fig. 4). Perioperative blood loss was negligible in all cases. No wound infections or vascular complications were documented. There was no recurrence, and there were no deaths.

Discussion

Because the first documented clinical case of a vertebral artery compression syndrome was seen in a man drawing a bow, the syndrome was named ‘bow hunter’s syndrome’

Table 1 Demography and clinical features of the series of eight patients

Patient number, sex, and age (in years)	Past medical history	Time to diagnosis (in months or years)	Symptoms	Diagnosis	Treatment, approach	Outcome at last follow-up
No. 1, M, 8	TIA	6 months	Ataxia, vertigo, nausea, hemiparesis, gait disturbance, bilateral pyramidal tract signs, when head to the right	– Cerebellar CVA – Bony malformation C1–C2 with stenosis at left for transv. of C2 – Fibrous band C1–C2 – Bilateral VA loops at C4–C5	– Vascular decompression, left anterolateral	– No syncope – Residual hemiparesis
No. 2, M, 9	None	1 year	Vertigo, nausea, nystagmus, torticollis (with head bended to the right and turned to the left), when head bended to the left	– Bony malformation C0–C1 – Bilateral bony compression of VA at sulcus of atlas – Spinal instability	– Vascular decompression, posterior bilateral – Fusion C0–C2	– Symptom-free after 7 years
No. 3, M, 16	Arnold's neuralgia	1 year	Neck pain and loss of consciousness during extreme head rotation (to either side)	– Bilateral bony stenosis at for transv. of C2 – Fibrous band on left – Right-sided VA duplication passing through two distinct foramina (aberrant course)	– Vascular decompression, anterolateral bilateral	– No syncope
No. 4, M, 42	None	6 months	Vertigo, during rotation to the right and head extension, loss of consciousness during extreme movement	– Fibrous band at sulcus of atlas	– Vascular decompression, left anterolateral	– No syncope – Less vertigo
No. 5, F, 46	Car accident, chiropractic manoeuver	>1 year	Neck pain, visual impairment, dysphasia; dizziness, dysphagia,	– Fibrous bands C1–C2	– Vascular decompression, left anterolateral	– No syncope – Residual psychosomatic symptoms
No. 6, M, 29	None	2 years	Vertigo and loss of consciousness during extreme head rotation (to the left-hand side)	– Fibrous band at sulcus of atlas	– Vascular decompression, left anterolateral	– Free of symptoms at latest FU (6 years)
No. 7, M, 24	None	1 year	Vertigo, nausea, and visual disturbances (during head bending to the right while playing tennis), when head bended and turned to the right-hand side	– Fibrous bands C1–C2	– Vascular decompression, right anterolateral	– Free of symptoms at latest FU (5 years)
No. 8, F, 21	None	3 years	Vertigo during extreme head rotation (to left side while playing volley)	– Bony stenosis at for transv. of C2 – Fibrous band on VA	– Vascular decompression, left anterolateral	– Free of symptoms at latest FU (7 years)

in the medical literature [4]. Compression syndromes of the vertebral artery at the loop around the atlantoaxial complex may have different aetiologies such as a bony, dural or arterial malformation; instability of degenerative, traumatic or inflammatory origin such as rheumatoid arthritis; or a tumour or pseudo-tumour [5–16]. In rare, so-called idiopathic cases, with no evident compressing element on imaging, surgical exploration has shown fibrous connective tissue (bands or rings) as the only possible cause of arterial constriction [9, 13, 17–20]. There is either a thickening of the complex atlantoaxial ligaments or some fibrosis of small neck muscles. Most often these compression syndromes present in middle-aged adults (being acquired), except for malformations that are generally encountered earlier in life, in a paediatric population (being congenital).

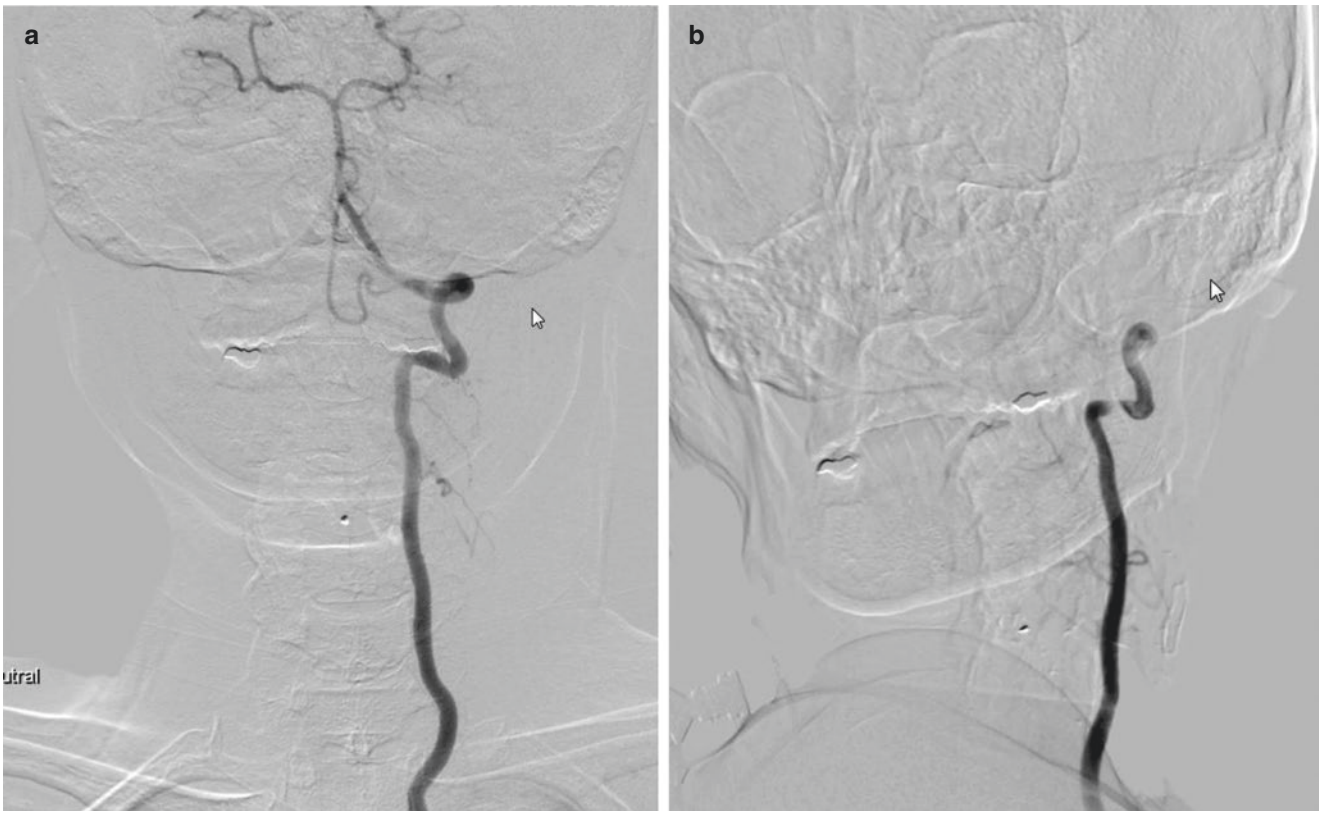

Fig. 2 Illustrative case of typical bow hunter's syndrome. (**a**) Normal left vertebral artery angiogram, anteroposterior incidence. (**b**) The same artery with head rotation 90° to the right, demonstrating occlusion at the atlantoaxial level

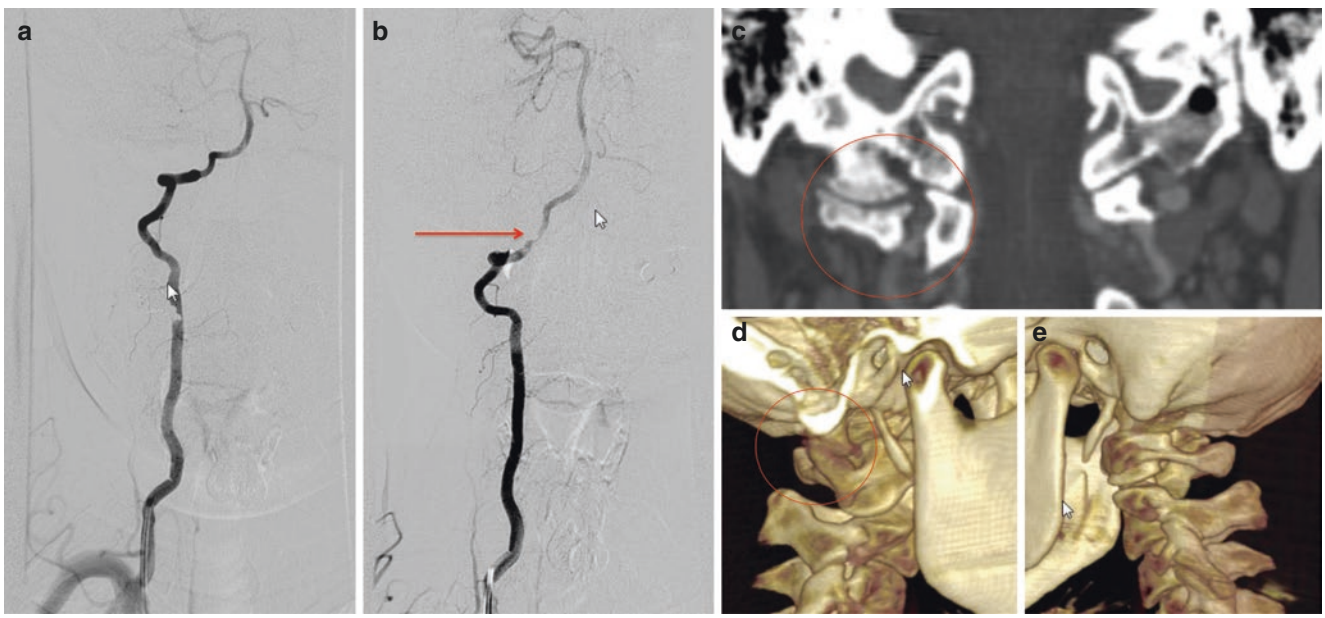

Fig. 3 Illustrative case of extrinsic compression by osseous malformation. (**a**) Normal right vertebral artery angiogram, anteroposterior incidence. (**b**) The same artery with contralateral head rotation, demonstrating tight stenosis at the atlantoaxial level (*red arrow*). (**c**–**e**) Computed tomography (CT) angiogram, bone window views: (**c**) coronal multiplanar reconstruction (MPR) and (**d**, **e**), volume-rendered reconstructions demonstrating abnormal lateral bone formations at the C1 and C2 level on the right side (*red circle*)

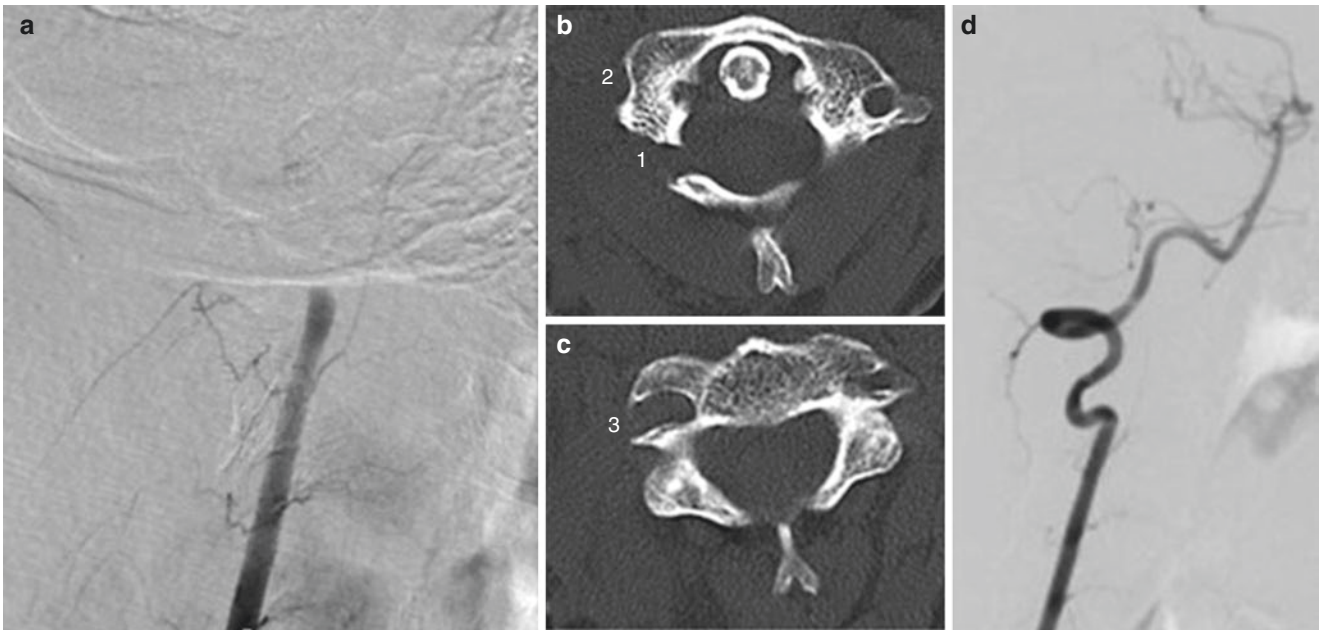

Fig. 4 Illustrative case of C1–C2 decompression. (**a**) Preoperative right vertebral angiogram with contralateral head rotation, showing occlusion at the atlantoaxial level. (**b**, **c**) Postoperative computed tomography (CT) scan, bone window axial views, demonstrating the three sites of decompression: behind the massa lateralis of the atlas (1), partial transversectomy of the atlas (2) and the axis (3). (**d**) Postoperative angiogram showing good patency in the same provoking position

The vertebral artery is stretchable to a large degree and is well adapted to follow the important and complex movements between the atlas and axis. If there is an element (e.g. an osteophyte) externally compressing the vertebral artery, this may completely occlude the vessel when the head reaches an extreme position [21, 22]. In most cases, collateral flow through the contralateral vertebral artery and via the posterior communicating artery down to the basilar artery is sufficient. However, arterial variations such as hypoplastic arteries or posterior inferior cerebellar artery (PICA)–ending vertebral arteries may cause haemodynamic vertebrobasilar insufficiency [23, 24]. Another pathomechanism is thromboembolic stroke due to repetitive microtrauma of the intima [25].

A third cause of vertebrobasilar insufficiency is compression of the vertebral artery below C2, which typically occurs during ipsilateral rotation of the head [9, 19, 26–31].

In fact, there are two different subtypes of BHS. These were defined in an earlier publication as ‘atlantoaxial BHS’ and ‘subaxial BHS’ [3]. Also, a ‘mixed type’ of atlantoaxial *and* subaxial vertebral artery compression with bilateral hypoperfusion has been documented [32].

The symptoms and signs of BHS comprise a spectrum from dizziness and vertigo to visual signs, syncope and cerebellar stroke [25].

Usually, BHS has a long delay between the initial symptoms and diagnosis. Typical symptoms occur during extreme head movements, especially during rotation and simultaneous extension of the head. In general, the symptoms immediately regress when the head is back in a normal position. A thorough diagnostic workup is necessary to verify or rule out extrinsic compression, which can then be treated accordingly.

The diagnostic workup starts with classical neuroimaging modalities: magnetic resonance imaging (MRI) or magnetic resonance angiography (MRA), and CT or CTA [33–36]. These will identify bony malformations or tumours. Dynamic x-rays may detect luxation or any other instability. This may be completed by transcranial Doppler (TCD) and neurophysiological examinations, including examination of the long tracts (motor evoked and somatosensory evoked potentials) and brainstem pathways (e.g. the brainstem auditory evoked response) [30, 37, 38].

Although it is invasive, dynamic angiography remains the gold standard to verify a positional vertebral artery occlusion [5, 13, 24, 39]. With the head extremely rotated and extended, a vertebral angiogram may exactly indicate the location of the vertebral artery occlusion. A four-vessel angiogram has to be performed to assess collateral flow into the posterior fossa [13, 40–42].

In most instances, decompression of the vertebral artery in its V3 (atlantoaxial) segment is a reasonable treatment [13, 31]. In fact, all but one patient treated at our centres underwent arterial decompression via an anterolateral cervical approach. All details of this technique can be found elsewhere [2, 21, 41, 43–46]. In summary, the vertebral artery is accessed from an anterolateral cervical approach and has to be controlled below the axis and above the atlas (the atlanto-

axial loop). For safe identification, micro-Doppler imaging may be very helpful in most cases [23]. The next step is to open the transverse foramina of the atlas and the axis. In some cases, additional sharp disruption of fibrous 'bands' or 'rings', or drilling of bony spurs compromising the vertebral artery, is necessary. Such tight fibrous structures are 'invisible' on standard imaging modalities. Consequently, they may be overlooked and may not be sufficiently addressed with a simple posterior approach [20, 47, 48]. On the other hand, a posterior approach with osteosynthesis may be necessary if the main or only cause of vascular compression is spinal instability (as seen in patient no. 2 in the present series). Finally, bilateral anterolateral approaches may be required (extremely rarely) if there are compressions on both sides (as seen in patient no. 3 in the present series).

While decompression of the compressed artery seems an obvious strategy, osteosynthesis of C1 and C2 to prevent vertebral artery occlusion has only been considered subsequently [7, 8, 20, 22, 25, 49, 50]. First, it was indicated only for atlantoaxial instability but then also as a treatment for BHS per se [8, 20]: osteosynthesis of the atlas and axis prevents any movements that could result in vertebral artery occlusion. Although osteosynthesis may show good relief of typical BHS symptoms, the main disadvantage is the major restriction in movements of the head. When considering osteosynthesis in BHS, especially when opting for screw-based techniques, it is crucial to keep in mind that the vertebral artery course may be highly variable. There are safer options without screws, but they generally achieve lower fusion rates [51].

The result of any surgical procedure—the aim of which is sufficient vertebral artery decompression—may be controlled by one of the following methods: micro-Doppler, indocyanine green angiography, CTA or DSA (Fig. 4d) [30, 38, 52–55]. In centres with a hybrid operating room the angiogram may be obtained intraoperatively, giving the possibility of immediate correction if necessary. At the latest, DSA should be performed within the first few postoperative days.

Concerning alternative treatment options, rare reports about treating BHS with endovascular devices (stents) exist, but the results have been inconsistent [42, 56]. However, as yet, the data are too sparse to allow general recommendations [57]. Also, the natural history is largely unknown. Some authors believe that repetitive mechanical occlusion and endothelial lesions finally lead to embolic strokes. For these reasons, many reported conservative options include cervical immobilization (e.g. a soft collar or orthesis) or resemble stroke prevention treatments [4, 35, 42, 58, 59]. Because of the small patient numbers and very heterogeneous treatment strategies, no clear recommendation for conservative treatment can be given yet.

Conclusion

Compression syndromes of the vertebral artery that occur at the craniocervical junction are extremely rare. In some cases, there are only subtle and misleading symptoms. The typical symptoms are vertigo and loss of consciousness with extreme turning of the head. A posterior fossa stroke is a rare but potentially fatal situation. The diagnosis is confirmed by dynamic computed tomography angiography (CTA) or dynamic angiography. Surgical decompression of the vertebral artery achieves good results. In some cases, atlantoaxial fusion may be indicated. Alternatively, conservative and endovascular options exist for inoperable patients.

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Competing Interests The authors declare that they have no competing interests.

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The Far Lateral Approach to the Craniovertebral Junction: An Update

Nabeel S. Alshafai and Tomasz Klepinowski

Abstract *Introduction:* Since 1972, when Hammon first described the far lateral approach (FLA) for treatment of vertebral artery aneurysms, it has undergone numerous modifications, including drilling of the occipital condyle, removal of the laminas of upper cervical vertebrae and so on. Also, the range of indications has increased exponentially.

Objective: In this paper we discuss state-of-the-art advances in the FLA, such as promising minimally invasive variants where an endoscope is used, and many others.

Methods: We reviewed all articles touching upon the FLA in the modern era (from the year 2000 onward) and selected those that presented a significant contribution to the development of the relevant approach. The database used was PubMed.

Results and Conclusion: We found several new caveats not mentioned in other reviews or book chapters. The FLA is an ever-changing field of battle where the common and ultimate goals are to minimize the risk of injuring the major vessel in the region—the vertebral artery—and to provide such an angle of attack upon the tumours in the anterior and anterolateral foramen magnum that it is feasible to ensure gross total resection. This paper is an update on the knowledge about this approach, which we feel is necessary.

Keywords Far lateral approach · Foramen magnum Vertebral artery · Occipital condyle · MIST

Introduction

Starting from the 1970s, many surgeons developed and introduced into their clinical practice new skull base approaches to the lesions of the anterolateral foramen magnum, with each of them bringing their own variations and modifications [1]. Hammon et al. in 1972 and thereafter Roberto C. Heros in 1986 described a true lateral suboccipital approach for vertebral and vertebrobasilar aneurysms [2, 3]. Heros described the combination of lateral suboccipital craniotomy, C1 laminectomy and drilling of the occipital bone at the level of the posterior occipital condyle (OC). However, Seeger was the first to describe, in 1978, partial resection of the OC to reach the anterior surface of the medulla oblongata [1]. In his variants, published in 1985, Koos suggested a way to increase the exposure of the ventral foramen magnum without drilling the OC [1]. After these initial reports, the far lateral approach (FLA) was popularized by many surgeons [1]. George described vertebral artery (VA) medial mobilization from C2 to its dural entrance point, with ligation of the sigmoid sinus and without condyle drilling [1]. Conversely, Spetzler, Bertalanfy and Seeger mobilized the VA from C1 to the dural entrance point by drilling the C1 facet, posterior C1 arch and posterior lateral third of the OC [1]. Also the range of indications has increased, and they currently encompass basically any lesion, including vascular malformations, in the anterolateral foramen magnum, lower clivus and upper cervical spinal cord [4–10]. Recently, in the twenty-first century, a number of great inventions have been implemented so as to improve the safety and efficacy of the FLA [11]. These are thoroughly covered in this paper, which constitutes an update on this procedure.

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Methods

Using the PubMed Medline database (accessed via https://www.ncbi.nlm.nih.gov/pubmed), we searched for the keyword term 'far lateral approach'. The search engine responded with 659 results. Those referring to a far lateral approach (the same technique name) but for lumbar disc herniation and other unrelated topics were omitted. We included cadaver, biomechanical and clinical papers produced since the beginning of the current millennium. Forty articles were found that were technically contributory, plus five book sections were added, which gave a total of 45 papers. They were analysed in terms of FLA modifications and upgrades.

Results

Minimizing Skin Incisions

Previous trends to perform a large U-shaped or hockey-stick incision are being gradually replaced with a tendency to make a small lazy S-shaped cut (see Fig. 1) or a linear incision placed about 2 cm medially from the mastoid tip [12, 13]. Such a small entry point turns out to be enough to dissect muscles, even layer by layer, detaching them from their attachments [13]. However, the ultimate length of the incision is always dictated by the inferior extension of the tumour and, if necessary, it might go down as far as C3 or lower [14, 15].

The Muscular Stage

To date, two major methods for accomplishing this step successfully have been introduced. They are:

- The piecemeal method [16, 17]
- The single-flap method [13, 18, 19]

Not surprisingly, each of them has pros and cons. The piecemeal method, in which each muscle is dissected separately, is safer in terms of protecting the VA. It also minimizes the risk of postoperative cerebrospinal fluid (CSF) leakage [17]. Nevertheless, it is vastly time consuming and thus it prolongs the entire procedure. The VA sits in the depth of the suboccipital triangle, which is made up of the obliquus capitis superior, obliquus capitis inferior and rectus capitis posterior major. Therefore, the piecemeal method ensures

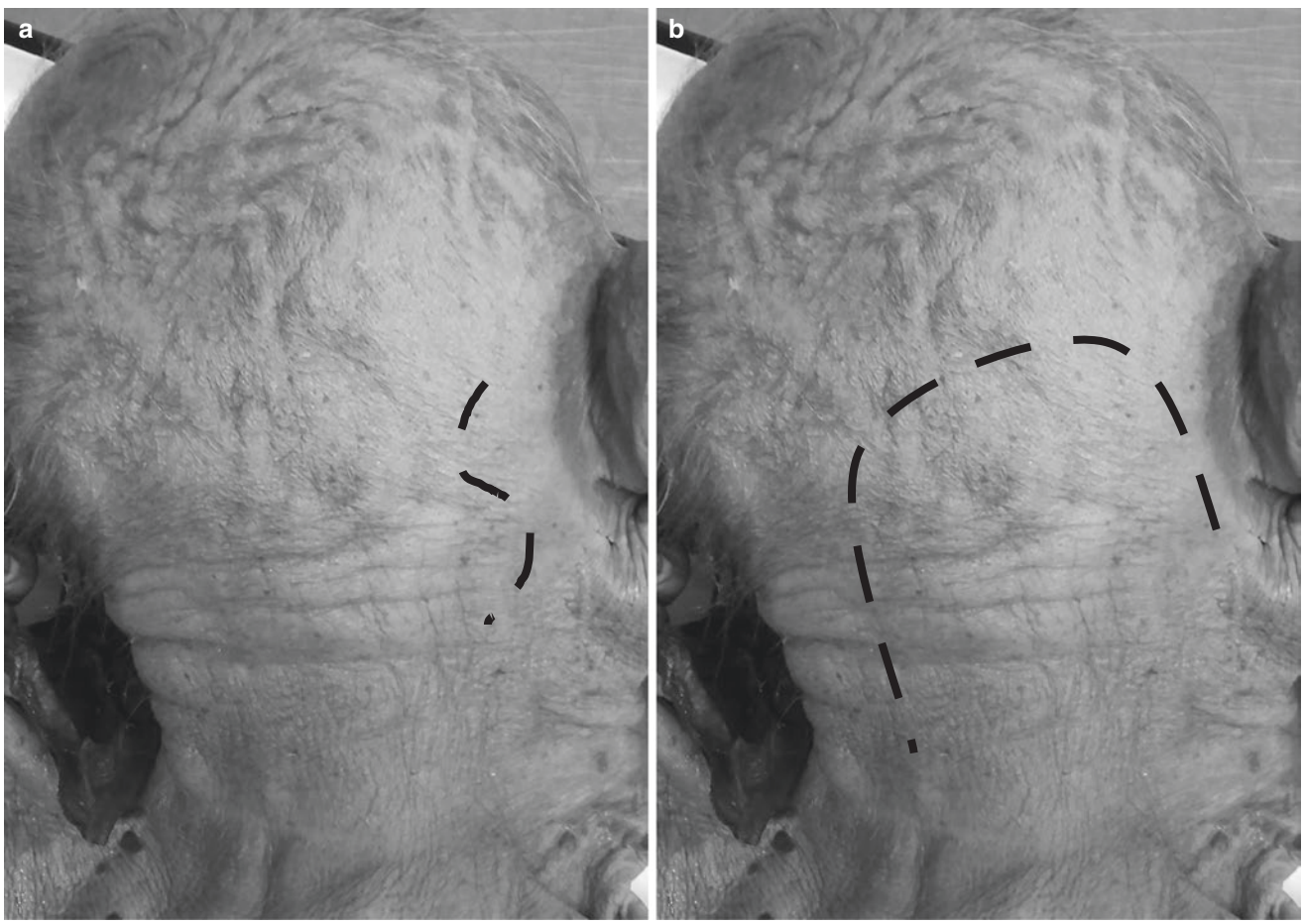

Fig. 1 (**a**) A short S-shaped incision positioned roughly one finger breadth medially from the mastoid process. It is often employed in new minimally invasive variants of the far lateral approach. (**b**) A classic inverted U-shaped skin incision, which consists of three limbs: midline vertical; horizontal, which runs just above the sigmoid sinus; and vertical, reaching just below the mastoid tip. (Photograph courtesy of the Department of Anatomy, Collegium Medicum, Uniwersytet Jagielloński (CMUJ), Kraków, Poland)

safe identification of the VA simply by meticulous anatomical dissection [17]. The single-flap method, on the other hand, entails cutting all muscles in the relevant region simultaneously. This provides quick access to the skull base. The potential threat is inadvertent injury of the VA. To minimize the risk of this serious complication, one should choose the side of the non-dominant VA on the basis of preoperative radiological assessment. In up to 40% of people the left and right VA differ: one is normal whereas the other is hypoplastic and of less significance [20]. This of course will not be valid if the tumour has a large lateral extension toward the dominant VA. The single-layer method also creates a bulk of muscles on one side of the surgical field, which might impair the corridor [21].

Handling the Vertebral Artery

Early control of this major vessel is imperative [22]. An easy and efficient four-step method has been invented to systematically locate the VA [13]:

1. Find the inferior–posterior point of the sigmoid sinus after having it unroofed [23].
2. Move posteriorly to identify the opisthion.
3. Crawl down to palpate the bony landmark—the posterior tubercle of the C1 arch [24].
4. Slide laterally to face the J-groove of the VA.

Note that the pulse of the VA at times may not be palpable, especially when the vessel is calcified or small [13, 21]. Therefore, thorough knowledge of the regional anatomy is obligatory. The artery is surrounded by fatty tissue and the venous plexus, which might be a source of profuse bleeding. The venous plexus is carefully dissected in three short steps:

1. Apply direct tamponade pressure.
2. Divide it with microscissors.
3. Coagulate with bipolar cautery.

This process exposes the arterial wall. Packing with haemostatic agents might be considered if the venous bleeding cannot be controlled with direct pressure.

VA transposition is usually part of the extreme lateral approach (ELA) [20, 25] but has also been used in the FLA. [21, 26]. We, however, disagree with mobilizing the VA in the FLA as this step is an inherent part of the ELA. The rationale for VA mobilization is that it widens the surgical corridor and hence minimizes manipulation of the low cranial nerves and the brainstem [25]. However, the issue of when and if it should be done at all remains controversial. The VA can be transposed at two sites: either at the C1 transverse foramen (extradural VA transposition) or at the dural entry point of the artery (intradural VA transposition) [15]. The former option is reserved for purely extradural tumours or intradural ones with a significant extradural component [27]. The latter option, on the other hand, is more beneficial for purely intradural tumours or extradural tumours with intracranial invasion [25, 28, 29].

The Occipital Condyle: To Drill or Not to Drill?

Probably, thorough preoperative evaluation with imaging is the key here [30]. Bone-windowed computed tomography (CT) carries details required to decide whether a patient's condyle will obscure vision, since surgery on a wider condyle might be more technically demanding [31, 32]. The same goes for a condyle whose medial lip protrudes into the foramen magnum [32]. Thus, drilling it increases the angle of attack [33, 34]. The chief objection stopping neurosurgeons from condylectomy is a potential risk of instability entailing subsequent fusion (see Table 1).

Fusion After Drilling the Condyle: Where Are We Now?

How the FLA destroys the atlanto-occipital joint and when or if fusion is needed are still a matter of debate. Earlier, to avoid craniocervical instability after condylectomy, some authors suggested that if more than 50% of the condyle is removed, CO–C1 stabilization must follow prophylactically [21, 35]. However, there have been reports of condylectomy reaching 75% without any clinically or radiologically evident instability [36]. Biomechanical studies have shown that the range of motion after the transcondylar approach increases significantly, but this has been proved on cadaveric specimens lacking action of the relevant muscles [37]. Stabilization of movement in the normal individual is largely affected by the

Table 1 Advantages and disadvantages of occipital condylectomy [17, 28, 36, 37, 43–47]

Advantages of occipital condyle drilling	Disadvantages of occipital condyle drilling
• Obviates the need for traction on the neuraxis • Visual exposure increases by ~15° in both adult and paediatric populations; this enables the surgeon to reach parts of the tumour adjacent to the anterior medulla and potentially facilitates gross total resection	• Risk of instability: fractures on follow-up have been reported • Significantly increases the range of movement between C0–C1 and C1–C2—and hence cumulatively between C0 and C2 (10.1° to the left 7.8° to the right for right occipital condyle drilling)—if two thirds is drilled • Increased risk of hypoglossal nerve damage • Increased risk of vertebral artery injury • Increased operating time • Increased postoperative pain

support of the relevant musculature. Currently, it is prudent to believe that occipitocervical instability is a cause of disturbing neck pain following a transcondylar approach, particularly if this pain is of a progressive nature or if there are concomitant transient neurological signs, or a new deformity. These patients might be candidates for stabilization, but so far there is no class I or II evidence to support it as prophylaxis following condylectomy reaching beyond a certain percentage [37]. On the basis of these findings, fusion should be considered on a case-by-case basis, depending on the bony destruction by the pathology, the amount of OC resection, the histology of the disease, and the patient's status [36].

Newly Emerging Variants

The Transcondylar Fossa Approach

In the transcondylar fossa approach (TCFA), there is no condyle drilling at all. The condylar fossa, which also serves as the posterior part of the jugular tubercle, is resected using a high-speed drill. The drilling is extradural. The key point of this approach is the bony removal along the condylar canal anteriorly, resulting in removal of the posterior part of the jugular tubercle. The atlanto-occipital joint can be kept intact [38].

The Minimally Invasive Supracondylar Transtubercular Approach

The minimally invasive supracondylar transtubercular (MIST) approach is very similar to the TCFA, as it also targets the condylar fossa. The difference between the two is that the MIST approach includes a significantly smaller craniectomy and minimal condylar resection; only about 10% of the condyle is drilled (see Fig. 2). Tuberculectomy is done in the same fashion described for the TCFA above. Here, an endoscope might be introduced via three disparate routes [12, 39]:

1. The inferomedial trajectory, inferior to the eleventh cranial nerve: This provides great vision of the anterior and anterolateral foramen magnum. It constitutes a useful alternative to a classic open FLA.
2. The transtubercular trajectory: This corridor is obtained once the jugular tubercle has been drilled. To the side of the endoscope there is the retracted dura, the accessory nerve is below, and the glossopharyngeal and vagus nerves are above. This angle of attack allows access to areas of the lower and middle clivus.
3. The superior trajectory: This necessitates superior medial retraction of the tonsils and the lower lateral cerebellum, which certainly is a limitation of this approach. However, the endoscope is inserted above the vagus nerve, just inferolateral to the flocculus. Therefore, it grants excellent vision of the cerebellopontine angle. This approach might be of particular value if a tumour has a significant extension into this region.

Tips and Tricks

Closure must be done meticulously. These are the ways to ensure it is watertight:

- A free abdominal fat graft, placed over the dura: The abdomen should be draped beforehand. It might be difficult in the modified park bench position, but still feasible. The autologous fat is also used to aggressively obliterate the mastoid and its antrum [28]. It might also be placed between the dura and titanium mesh [21].
- Fibrin glue [21].
- Suboccipital aponeurosis: This may serve as a dural patch [40].

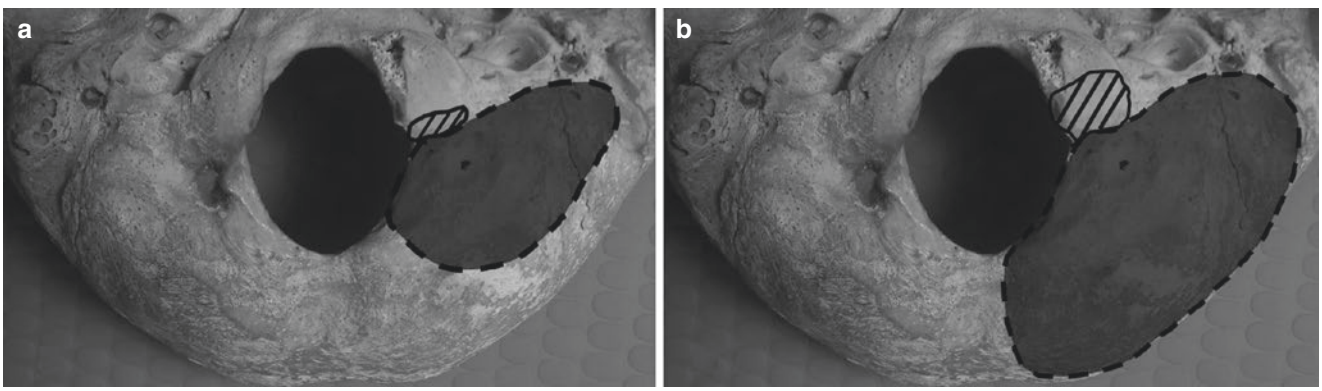

Fig. 2 Comparison of craniotomy (shown by the *shaded area*) (**a**) for the minimally invasive supracondylar transtubercular (MIST) approach and (**b**) for the classic far lateral transcondylar approach. In the MIST approach, only up to 10% of the condyle is drilled, whereas in the classic approach, usually at least one third of the condyle is resected (shown by the *oblique lines*)

- Fascia overlying the sternocleidomastoid: This might be harvested to serve as a dural graft at the time of dural closure [13].
- Fascia lata: This may serve as a dural patch. The lateral surface of one thigh should be draped in advance [41].
- Bone wax: This might be used to obliterate mastoid air cells.

Positioning pearl: In either a modified park bench position or a semi-sitting position, check motor and sensory function (of adequate nerves) before and after positioning to ensure that there are no adverse changes from baseline once the positioning is complete. If it cannot be conducted (as abnormal potentials appear even though the positioning has been checked several times), then awake surgery has been described as an option [42].

Conclusion

The far lateral approach has evolved remarkably in recent years, and all of the changes that have been implemented so far seem to have three major goals in common: first, to protect the vertebral artery and therefore minimize the risk of potentially dangerous complications; second, to avoid the risk of instability by not resorting to drilling much of the occipital condyle; and third, to ensure gross total resection with minimal or no retraction, which might potentially cure the patient completely. This paper is an update and checkpoint on the path to the perfect version of the far lateral approach.

Compliance with Ethical Standards No financial support was received for this work.

Competing Interests The authors declare that they have no competing interests.

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Endoscope-Assisted Far Lateral Approach to the Craniovertebral Junction with Neuronavigation: A Cadaver Laboratory Experience

Francesco Signorelli, Marco Pace, Vittorio Stumpo, Pasquale Ciappetta, Alessandro Costantini, Alessandro Olivi, and Massimiliano Visocchi

Introduction

The far lateral approach (FLA) is a technique performed nowadays to gain access to and remove intradural lesions located ventrolaterally to the brainstem and to the craniovertebral junction (CVJ).

The FLA represents an extension of the suboccipital approach with additional removal of a variable amount of occipital bone. During the 1980s, Heros and George separately performed the first attempts at the FLA [1, 2], followed shortly afterward by Spetzler and Grahm [3]. Since then, several variants of the approach have been proposed, which can be considered as a continuum in which the extent of bone removal is guided by the localization and dimensions of the lesion to be removed.

In recent years, endoscopy has gained wide popularity and became a fundamental tool in the neurosurgical armamentarium. Endoscopy has been used extensively for treating skull base lesions, and several studies have reported the indications and advantages of endoscopy-assisted techniques for operating on the CVJ [4, 5]. The neuronavigation system provides real-time information, which allows accurate localization, better identification and precise resection of these lesions, thus significantly increasing the safety of the approach [6].

Few anatomical studies of neuroendoscopy combined with neuronavigation in the ventral craniocervical junction have been reported to date [6–8].

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Our cadaver study was conducted with the purpose of investigating the benefits, potential and limitations of neuronavigation and endoscopy in the FLA to the craniocervical junction.

Materials and Methods

Materials

Two adult formalin-fixed cadaver specimens (four sides) were examined in stepwise dissection. The following instruments were used: a multidetector 128-layer computed tomography (CT) scanner, binocular lenses (visual magnification 3.5×), 420 mm; 0° and 30° rod lens endoscope (Storz, Tuttlingen, Germany); neuronavigation (Medtronic StealthStation Treon Plus); high-speed drill (Storz); vacuum aspirator (Super Vega Battery); digital camera (EOS 7D telescopic lens image stabilizer ultrasonic macro 100 mm; Canon, Tokyo, Japan); operating microscope (Zeiss, Oberkochen, Germany); microsurgical instruments; and stainless steel headholder.

Methods

The thawing, irrigation, fixation and perfusion phases were performed according to a protocol developed at our research centre. The formalin-fixed samples thus underwent a high-definition CT scan after being injected with the monomeric iodinated contrast medium Iomeprol (Iomeron®), and the vascular system was later perfused with coloured silicone solutions: blue for the venous system and red for the arterial system. The imaging data (saved in DICOM [Digital Imaging and Communications in Medicine] format) were stored on compact disc (CD) and imported into the neuronavigation workstation to create three-dimensional (3D) reconstructions.

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The neuronavigation system, binocular lenses, microscope and endoscope were used during simulation of the four FLAs (one on each side of the two cadaver specimens).

Results

After an inverted hockey-stick incision, stepwise dissection of all muscular layers was performed; the occipital artery was identified and isolated from its distal to proximal end until the corner of the jaw, together with the greater occipital nerve, which was also dissected and isolated along its superficial course. Close to the artery, the occipital vein was identified, together with the emissary mastoid vein and its foramen. Once this latter vein was sectioned, the previously isolated occipital artery and vein were overturned posterolaterally and preserved. The suboccipital triangle was then identified, housing the vertebral artery on its floor, wrapped by its venous plexus and surrounded by the C1 nerve. The muscular branch of the vertebral artery was also identified along its posterior surface, and the distance between the midline and the artery entry point into the dura mater was measured (1.9 cm and 2.1 cm in specimen A, and 1.8 cm and 2.3 cm in specimen B). After subperiosteal vertebral artery mobilization, a lateral suboccipital craniectomy was performed, followed by C1 semiarch removal. Subsequently, with the aid of neuronavigation, the occipital condyle was identified and its posterior third was removed. The dura mater was then incised approximately 1 cm from the vertebral artery entry point and reflected laterally. On both sides of the two specimens, the posterior inferior cerebellar artery (PICA) displayed an intradural origin. During the intradural phase, optimal fixation of the cerebellar parenchyma in terms of trophism and consistency, complete arterial and venous perfusion of the intracerebral vessels (demonstrated by the filling of the cerebellar cortical vessels) and arachnoid preservation were evident (Fig. 1). After the arachnoid incision, endoscopic exploration by means of the 0° and 30° endoscope lenses was performed, allowing identification of the lower cranial nerves, PICA and intradural vertebral artery (Fig. 2). In one case, during the endoscopic exploration of the inferior corridor, tearing of the emerging roots of the eleventh cranial nerve occurred. With the aid of neuronavigation, it was possible to identify the posterior skull base structures, the clivus and its condyles (Fig. 3).

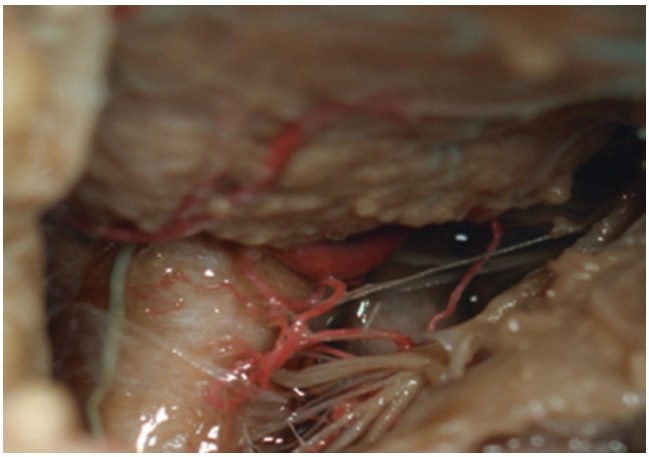

Fig. 1 Intradural vision of the bulbomedullary junction, brainstem, lower cranial nerves and posterior inferior cerebellar artery (PICA) obtained with the far lateral approach

Discussion

The FLA nowadays represents a mainstay for the surgical treatment of intradural pathologies of the ventral craniocervical junction. Since the first description by Heros and George, extensive discussion and modifications of this approach have been reported in the literature.

Overall, the crucial steps in this approach are the positioning, skin incision, muscular dissection, vertebral artery (VA) management, suboccipital/retrosigmoid craniotomy, lateral foramen magnum rim removal and drilling of the condyle, dural opening, intradural microsurgery, and closure. In the extensive literature on the FLA, execution of each step is described in several ways by different authors. The crucial issues with regard to the extradural steps of the FLA are the extent of condyle resection (with its potential for CVJ instability) and VA management, which have both been described with different strategies ranging from the most aggressive to more conservative ones.

The ventrolateral bulbomedullary junction—the target of the FLA—is a complex anatomical region characterized by dense distribution of nerves and blood vessels, an irregular skull base and a narrow operating space; therefore, execution of each step requires accurate topographical anatomy knowledge and extensive experience in both spinal and skull base surgery. Endoscopy-assisted techniques exploit the potential of the endoscope to provide a wider, brighter, magnified and multi-angle panoramic view through narrow surgical corridors. Overall, these features result in improved visualization of the surgical field while minimizing brain retraction and trauma to neurovascular structures that need to be identified and preserved while one is working in the space around and beyond them.

Several cadaver studies have demonstrated the use and benefits of endoscopy in the FLA [4, 6, 9]. In one study [9] the surgical corridors for the insertion of the endoscope were divided into superior, middle and inferior corridors. Cranial nerves VII and VIII, IX and X, and XII are the roofs and floors, respectively, of the three corridors, with access to and observation of aspects of the brainstem and posterior

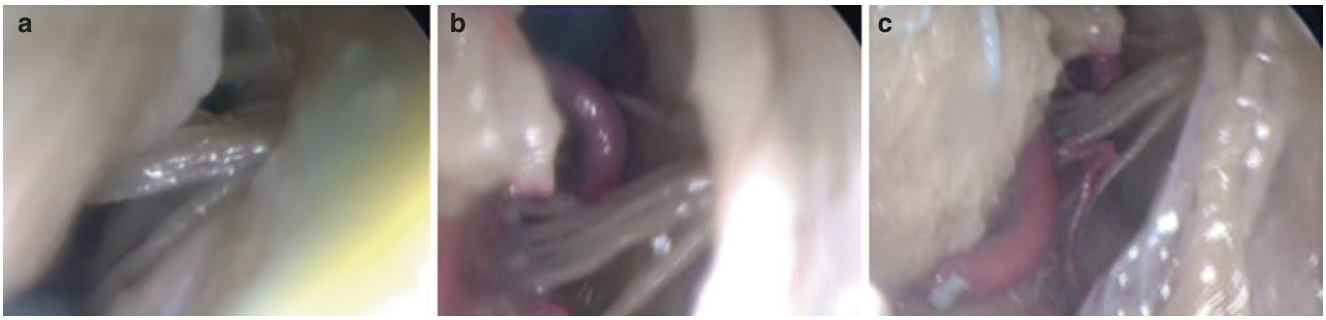

Fig. 2 Endoscopic intradural exploration through (**a**) the upper corridor, (**b**) the middle corridor and (**c**) the inferior corridor, according to Chotai et al. [9]

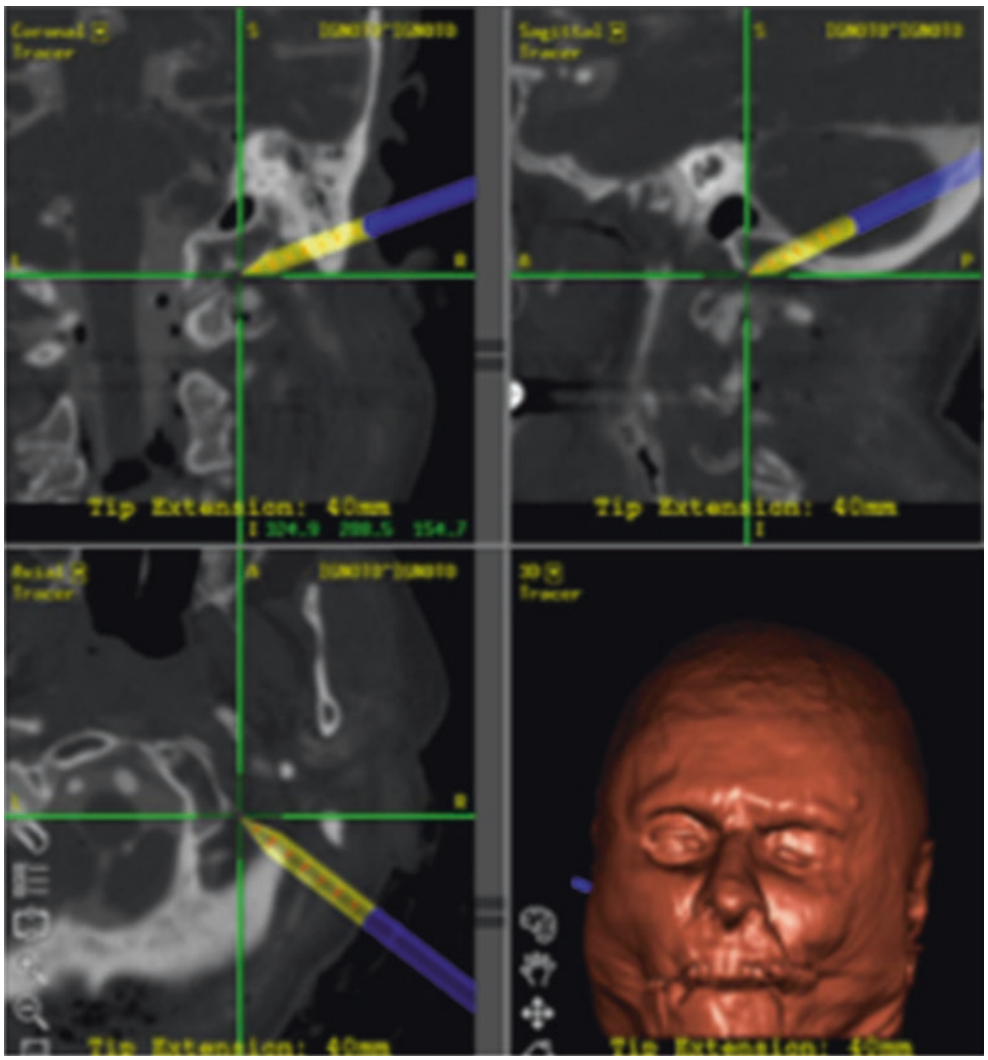

Fig. 3 Neuronavigated computed tomography (CT) scan: identification of the occipital condyle with the probe

circulation provided by means of the 0° lens (for the superior and middle corridors) and the 30° lens (for the inferior corridor).

Another cadaver study compared the 3D endoscopic and microscopic views in the FLA after partial condylectomy and resection of the jugular tubercle [10]. In that study the 3D endoscope provided a better view of the bulbopontine segment of the anterior inferior cerebellar artery (AICA), the subarcuate artery, the labyrinthine artery, the origin of the PICA, the vertebrobasilar junction, the anterior spinal artery and the lateral aspect of the lower cranial nerves. However, the study concluded that the 3D endoscopic probe was too large and, although there was reasonably safe and adequate access for visualization of the deeper structures, the surgical manoeuvrability was significantly hampered. On the other hand, in a more recent study [6] the authors claimed that the endoscopy-assisted FLA has the potential to reduce exposure of the surrounding brain tissue and shorten the neck incision, thus avoiding excessive injury to the muscles and ligaments. Several authors have reported similar benefits with use of the endoscope in clinical series. These studies reported a significant benefit from the ability of the endoscope to identify any tumour that was adherent to the brainstem or clivus and was amenable to resection [9, 11–13].

Nonetheless, there are several limitations of endoscopy-assisted techniques that warrant awareness. First, there is a learning curve associated with endoscopy-assisted techniques. In addition, the endoscope occupies space in the surgical corridor, leaving less room for instruments. Moreover, insertion and removal of the endoscope increase the risk of damaging neurovascular structures and must be done under microscopic visualization. Finally, the endoscope can sometimes provide superior visualization without equivalent surgical manoeuvrability, especially in a narrow curvilinear corridor between delicate neurovascular structures, as in the case of the CVJ [9].

In our anatomical study, the endoscope allowed exploration of the three main access routes as described, and the intradural anatomy was extensively observed, but in one case we noticed tearing of some lower cranial nerve fibres, particularly of cranial nerve XI, which is especially fragile because of the discrete cerebellar rigidity.

The endoscopy-assisted FLA with navigation guidance combines the advantages of neuronavigation and neuroendoscopy. Neuronavigation could overcome the risk connected with incorrect introduction of the instrument and the inability of the endoscope to see straight ahead, resulting in possible injury to adjacent structures [6, 14, 15]. In addition, neuronavigation can be employed before surgery to determine the direction and extent of jugular tubercle and occipital condyle drilling needed for the FLA. Real-time monitoring of the drilling size allows the surgeon to avoid injuring the hypoglossal canal.

In our study, neuronavigation was useful to confirm the identity of clearly recognizable structures and, more crucially, provided essential support for safe identification of deep structures in the preliminary and intermediate phases of dissection, such as the occipital condyles, the dens and the clivus.

Conclusion

Anatomical dissections constitute irreplaceable training for young surgeons and have further importance in approaches to one of the most complex anatomical regions in the entire human body: the craniovertebral junction. With the aid of endoscopy and image guidance, it is possible to maximize the dissection experience. Furthermore, in the clinical setting, use of neuronavigation during endoscopic procedures provides the surgeon with constant orientation in the surgical field, thus increasing the accuracy and the safety of the approach.

Compliance with Ethical Standards No financial support was received for this work.

Competing Interests The authors declare that they have no competing interests.

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Extreme Lateral Approach to the Craniovertebral Junction: An Update

Nabeel S. Alshafai and Tomasz Klepinowski

Abstract *Introduction:*The term 'extreme lateral approach' (ELA) was first introduced by Sen and Sekhar relatively recently (in 1990). Its definition varies and remains controversial, but it generally entails more aggressive bony removal than the far lateral approach (FLA).

Goal: In this paper we review the relevant literature and weigh up the advantages and disadvantages of this approach. We propose methods to manage the complications resulting from the more invasive character of the ELA. Some modern trends regarding how to definitely distinguish the ELA from the FLA are also presented.

Methods: Using the PubMed database, literature was collected on the relevant topics and subsequently reviewed. All up-to-date tips and tricks were carefully gathered, and current morbidity and mortality rates were obtained, as well as further perspectives.

Results and Conclusion: The morbidity associated with the ELA remains higher than that associated with the FLA, but the mortality nowadays is comparable. The ELA undoubtedly is a challenging procedure requiring deep insight into the relevant anatomy and its normal variants.

Keywords Extreme lateral approach · Meningiomas · ELITE Vertebral artery transposition · Craniovertebral junction

Introduction

The extreme lateral approach (ELA) is generally considered a more aggressive extension of the far lateral approach (FLA). The term 'extreme lateral approach' dates back to 1990, when Sen and Sekhar depicted an alternative way to handle meningiomas and schwannomas located anteriorly at the cervicomedullary junction [1]. The rationale behind this procedure was to enable gross total resection of lesions with significant lateral extensions that would otherwise be inaccessible, e.g. via the anterior or classical far lateral approaches. Essentially, the ELA involves a greater extent of bony removal, as it sometimes aims for nearly total condylectomy [2] or skeletonization of the jugular bulb along with the sigmoid sinus (in the transjugular variant of the ELA); more often, vertebral artery (VA) transposition is performed. The exact delineation of craniotomy and other nuances lie within a spectrum of variation under the umbrella of the ELA, as reported by Salas and Ziyal [3, 4]. Thus, it renders the surgical corridor wide open, but on the other hand it is inherently associated with higher rates of morbidity and mortality [5–7].

Methods

A literature search was performed by means of the PubMed Medline database (accessed via https://www.ncbi.nlm.nih.gov/pubmed). The phrase that constituted the subject of our search was 'extreme lateral approach'. Initially, as many as 259 results were produced. Forty-two papers pertaining to the craniocervical ELA were found, issued between September 1990 and July 2016. Since the present paper is an update on what is already known, only contemporary literature after the year 2000 was included in our analysis. Thus, ultimately, 24 articles were included, published from November 2001 until July 2016.

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Results

We found that the literature pertaining to the ELA is rather sparse, with the mean production of new articles being less than 2/year (1.62/year since 1990) worldwide (see Fig. 1 for the last 9 years). The year 2016 was the most productive since 2004. All of the papers included in this review then underwent critical scrutiny to identify modern trends, obstacles, incidence rates of complications, innovations, and future perspectives.

Modern Indications

A variety of lesions might be considered appropriate surgical candidates for the ELA. Cases involving the following pathologies have been reported to be operable via the ELA, according to our literature review [6, 8–21]:

- Large meningiomas.
- Distal VA aneurysms.
- Prepontine epidermoids.
- Superior cervical nerve sheath tumours (neurofibroma and schwannoma).
- Hypoglossal neurinomas.
- Chordomas, chondrosarcomas.
- Paragangliomas.
- Neuroepithelial cysts.
- Congenital and acquired abnormalities of C0–C1, such as hypertrophy of the atlanto-occipital joint and a lateral mass, or osteochondroma; the ELA also allows for reduction of atlantoaxial rotatory fixation, which has been done so far in 13 paediatric patients.
- Spinal dural fistula.
- Extracranial VA-to-intracranial VA bypass.
- Dens pathologies (e.g. rheumatoid arthritis).

However, when it comes to tumours, it is often not the histopathological diagnosis that influences the choice of ELA but, rather, the precise location of the lesion. Zozyulia et al. described detailed criteria for choosing a surgical approach for lesions in the ventral and ventrolateral regions of the craniovertebral junction [21]. It was suggested that the ELA might be taken into account in cases of a large tumour anterior to the spinal cord, extending beyond the midline, that pushes the cord posteriorly and takes over its anterior space. In other scenarios, less invasive procedures should be pondered, such as the FLA. Currently the ELA is used to treat approximately 17% of tumours of the ventral and/or ventrolateral craniovertebral junction [21].

Mastering Vertebral Artery Transposition

Since mobilization of this vessel is an inherent part of this wide approach, mastering the technique is essential for potential final success. It can be accomplished in two ways: either at the C1 foramen transversarium or at the dural entry point of the VA [22], but rarely at both. There has been much controversy around this step, with a number of neurosurgeons refuting its usefulness, having realized the potential risks it involves and that this might actually be an unnecessary step—for instance, when the tumour itself sets the neuraxis aside, automatically opening the corridor. Table 1 shows all of the current top arguments for and against this stage of the ELA [22].

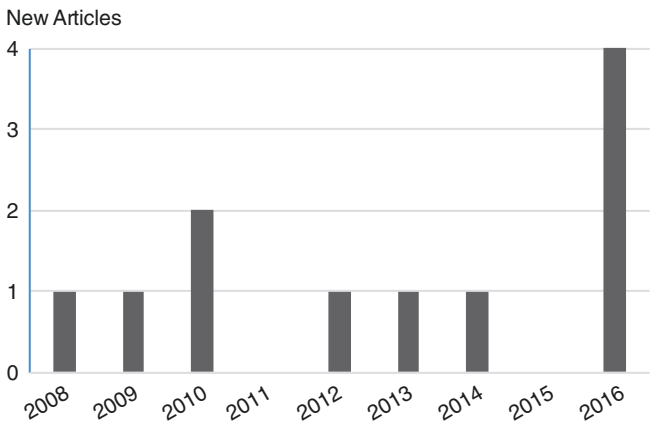

Fig. 1 Numbers of new articles on the extreme lateral approach that have been issued each year over a 9-year period

Table 1 Arguments advocating and negating mobilization of the vertebral artery (VA) [22]

Arguments in favour of VA transposition	Arguments against VA transposition
• It facilitates condylar drilling for unimpeded access to the tumour • Meningiomas of the deep foramen magnum may otherwise be difficult to handle • It is helpful if the tumour diameter is <3 cm • It is useful in anterior foramen magnum meningiomas and those above the VA • It minimizes manipulation of the neuraxis	• Large tumours displace the neuraxis back, creating a working space that makes VA transposition and condylar drilling unnecessary • VA transposition may lead to troublesome venous plexus engorgement and bleeding • It is unnecessary in neither lateral foramen magnum meningioma nor those below the VA • There is a risk of formation of a pseudoaneurysm or an arteriovenous fistula

A Brand New Concept: Extreme Lateral Inferior Transcondylar–Transtubercular Exposure

In the extreme lateral inferior transcondylar–transtubercular exposure (ELITE) approach, the occipital condyle is drilled parallel to the articular surface but—notably—sparing the articular facet of the atlas [23]. This manoeuvre exposes the hypoglossal canal. The structure seen just above it, which forms its roof, is a jugular tubercle, which is then resected. The rationale behind this step is that the convex surface of the tubercle obstructs the view of the most anterior and contralateral anterolateral area of the foramen magnum. The jugular tubercle also makes up a medial wall of the jugular foramen; thus, care must be taken to not injure the neurovascular structures of it. Drilling the tubercle extradurally in such a manner produces excellent exposure [23]. To preoperatively predict whether a jugular tubercle will affect the scope of vision, computed tomography (CT) is performed and measurements of the jugular tubercle are taken: the width on the coronal section, and the thickness and length on the axial section. These measurements on CT are very close to the ones taken on dry skulls and hence may be safely used [24].

In children it is advised against drilling much of the condyle, since this structure grows as the entire skeletal system does, and the influence of such a manoeuvre upon the stability is even harder to predict in this paediatric population [25].

Current Complications of the Extreme Lateral Approach

For the time being, the situation is still not satisfactory, as the morbidity and mortality related to the ELA remain high (see Fig. 2). It is most often associated with deficits of the lower cranial nerves, of which the incidence has been reported to be 17.9–50% [6, 22]. The nerve that is injured most frequently, even temporarily, is the hypoglossal nerve (14.2–50%), followed by the vagus/glossopharyngeal complex (4–37.5%). The accessory nerve is at the bottom of this list (0–25%). Other sequelae of this aggressive approach include [6, 26]:

- Development or exacerbation of hydrocephalus (~13%).
- Cerebrospinal fluid (CSF) leaks (22%).
- Central nervous system infection (5–25%).
- Transient or permanent hemiplegia or tetraplegia (12.5%).
- Instability: this is controversial. This supposedly might occur after 50% of the condyle is drilled [27], but a number of cases have been published where the drilling exceeded 75% and still there was no sign or symptom of instability [28].

The general incidence of complications might vary depending on the nature of the lesion, as well as which variant of ELA is chosen. Thus, the incidence appears to be higher in more invasive transjugular ELA (tjELA) when the surgeon is aiming for paragangliomas and schwannomas [27, 29]. The classic tjELA is possibly the most aggressive posterolateral approach to the foramen magnum, as it also entails a total mastoidectomy with transposition of a facial nerve from the facial canal. Also, some critical venous structures are exposed: the jugular bulb and vein [30]. This, altogether, might result in a higher complication rate.

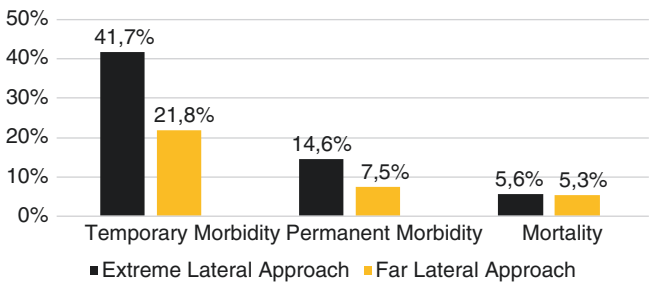

Fig. 2 Comparison of temporary morbidity, permanent morbidity and mortality rates between the extreme lateral approach (ELA) and the far lateral approach (FLA) [6]

Conclusion

The extreme lateral approach (ELA) has been given little attention over the last 15 years, especially in comparison with its less invasive sister, the far lateral approach (FLA). Therefore, one might ask what future the ELA has with current tendencies to assume a minimally invasive attitude. The ELA, however, still appears to be a reasonable option for large anterior and anterolateral lesions when greater exposure is desperately required. It could also still be utilized as an approach in complex vascular lesions in this region. However, it goes along with a higher complication rate than other posterolateral approaches to the foramen magnum region. This morbidity and mortality may also be attributed to the fact that the lesions themselves that qualify for the ELA are larger and of a more complex nature.

Compliance with Ethical Standards No financial support was received for this work.

Competing Interests The authors declare that they have no competing interests.

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The Extreme Lateral Approach to the Craniovertebral Junction: An Anatomical Study

Francesco Signorelli, Walter Pisciotta, Vittorio Stumpo, Pasquale Ciappetta, Alessandro Olivi, and Massimiliano Visocchi

Abstract *Background*: The extreme lateral approach is a direct lateral approach which allows a good control of the entire length of the vertebral artery (VA), the jugular foramen, the lowest cranial nerves, and the jugular–sigmoid complex. Herein we try to exploit the variants of the approach and we identify indications, advantages, and disadvantages.

Methods: All phases of the study were conducted at the Institute of Public Health Section of Legal Medicine and Insurance of the University. We performed the extreme lateral approach in four subjects, who died between 24 and 48 h before in non-traumatic circumstances (three men and one woman).

Results: The great auricular nerve, the spinal accessory, the branches of the first ventral spinal nerves, the jugular vein, and the vertebral artery were identified in all the cadavers. In all cases the right VA exited from the transverse foramen of C1. The site of SCM piercing the accessory nerve was at a distance from the tip of the mastoid between 3 and 4 cm (3.3 in one case, 3.4 in 2 cases, and 3.7 in one case). No vessels and nerves have been damaged after being identified and isolated.

Conclusions: Extradural lesions at the ventro-lateral aspect of the CVJ may require an extreme lateral approach, a procedure more aggressive comparing with far lateral approach, which represents a reasonable option for large anterior and anterolateral lesions when greater exposure is required.

Keywords Extreme lateral approach · Craniocervical junction · Craniovertebral junction

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Introduction

Lesions of the ventrolateral aspect of the craniovertebral junction (CVJ) still represent a challenge to neurosurgeons. Several lateral approaches have been adopted to manage these lesions while ensuring minimal manipulation of the spinal cord and careful control of the main vascular structures.

The indications, technical descriptions, nuances, advantages and drawbacks of these approaches have long been debated by neurosurgeons [1–14]. A plethora of terminology has been adopted since the early 1980s, broadly summarized as the posterolateral or far lateral approach and the anterolateral or extreme lateral approach (ELA).

The ELA, as originally described, is a direct lateral approach to the deep anterior portion of the sternocleidomastoid muscle (SCM), behind the internal jugular vein and anterior to the vertebral artery (VA). This approach allows a wide domain of the VA, the jugular foramen, the lowest cranial nerves and the jugular–sigmoid complex.

The main application of this approach is the treatment of extradural lesions, but in some circumstances it can also be used to remove intradural lesions. Key points of this approach are (1) spinal accessory nerve preservation during SCM overturning; (2) VA transposition by subperiosteal dissection along the sulcus arteriosus on the posterior arch of the atlas; (3) drilling of the lateral mass of the atlas; and (4) removal of the lower surface of the occipital condyle, if necessary. Different combinations of drilling of the occipital condyle, the facets and laminae of C1 and C2, and the tubercle and jugular process, along with different amounts of suboccipital craniectomy, provide the surgeon with different ways to approach the CVJ anterolaterally.

Occipitocervical fusion and stabilization are warranted after removal of more than the posterior third of the occipital condyle.

In this cadaver study we aimed to better define the potentials and limitations of the extreme lateral approach to the craniocervical junction.

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Materials and Methods

Materials

The following instruments were used: binocular lenses (visual magnification 3.5×), 420 mm; high-speed drill (Storz, Tuttlingen, Germany); vacuum aspirator (Super Vega Battery); digital camera (EOS 7D telescopic lens image stabilizer ultrasonic macro 100 mm; Canon, Tokyo, Japan); operating microscope (Zeiss, Oberkochen, Germany); microsurgical instruments; and a stainless steel headholder.

Methods

The ELA was performed at the Section of Legal Medicine and Insurance, Institute of Public Health, at the Catholic University School of Medicine in Rome, using four cadavers of individuals (three male, one female) who died between 24 and 48 h beforehand in non-traumatic circumstances. The Institute is provided with two dissection stations, which simulate the operating room environment. In all four cases a diagnostic examination was required, and the neck was an area of interest. Special authorization had already been obtained from the ethics committee (protocol no. P663/EC/2010 approved on 28 July 2010; subsequent amendment no. P437/CE 2012 approved on 2 May 2012).

Results

A right approach was performed on every specimen. The great auricular nerve, the spinal accessory nerve, the branches of the first ventral spinal nerves, the jugular vein and the VA were identified in all four cadavers, and in all cases the right VA passed through the transverse foramen of C1. The site where the accessory nerve pierces the SCM was found at a distance of 3–4 cm from the tip of the mastoid (3.3 cm in one case, 3.4 cm in two cases and 3.7 cm in one case).

No vascular or nervous structure was damaged after being identified and isolated.

Discussion

The ELA involves opening of the C1 transverse process and VA transposition. George [15] was the first to report medial transposition of the VA. This technique reduces the risk of VA injury during drilling of the medial atlanto-occipital joint.

The ELA provides good access to the bone and the extradural anterior and lateral space. It can be extended caudally to the subaxial cervical spine, and it offers simultaneous control of the VA, the cervical segment of the internal carotid artery (ICA), the lower cranial nerves and the sigmoid–jugular complex [15].

The main indications for this approach are lesions located in the anterolateral aspect of the CVJ or in the spinal cord, with a large extradural component, such as meningiomas and dumbbell neuromas; it is also indicated for osseous lesions such as chordomas or metastases of the lateral portion of the CVJ. Moreover, this approach can be extended anteriorly to reach the jugular foramen, allowing removal of simple glomus tumours that are not invading the carotid artery (CA) and also removal of neuromas growing in this anatomical region.

By performing an inverted L incision, we could conservatively identify the SCM through incision of its anterior margin and sectioning of its occipital and temporal insertions.

The inferolateral reflection of the SCM and cranial nerve XI allows direct lateral access to the anterior aspect of the CVJ between the internal jugular vein and the dural sac. The fundamentals of this procedure are similar to those described by Verbiest, who used this technique to remove osteophytes causing compression of the VA and the nerve roots of the lower cervical spine [16]. This technique is also similar to oblique corpectomy, which was described by George et al. [17].

During ELA, the muscles are detached from their insertion on the transverse process of the atlas. Great attention must be paid to avoiding damage of the VA, internal jugular vein and spinal nerves, which are found just underneath these muscles.

Our dissection experience further confirms that ELA is able to provide exposure of the whole odontoid process, the inferior clivus and the medial surface of the contralateral atlanto-occipital joint.

The ELA can thus provide an alternative to the transoral approach for extradural lesions of the CVJ, with the advantages of a shorter surgical route and avoidance of rhinopharynx contamination. Furthermore, we have been able to confirm that the ELA can be further extended downward to deal with lesions located at C2, C3 or lower levels.

Conclusion

The anterolateral or extreme lateral approach is very effective for treating anterolateral craniovertebral junction lesions with a relevant extradural component. This approach can easily be combined with a posterolateral procedure and can be extended to the lower cervical spine (Figs. 1 and 2).

Compliance with Ethical Standards No financial support was received for this work.

Competing Interests The authors declare that they have no competing interests.

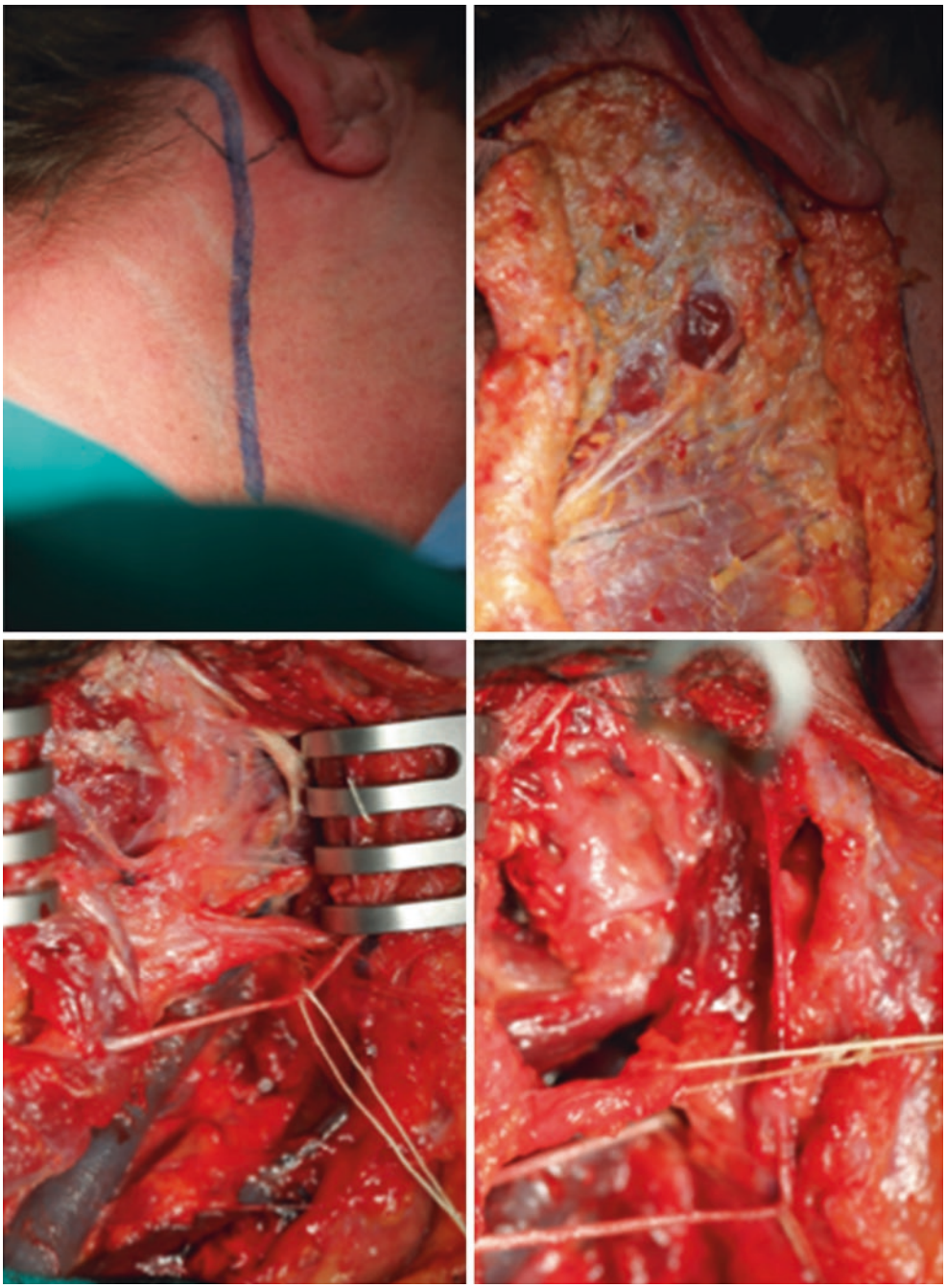

Fig. 1 Stepwise extradural dissection in the extreme lateral approach

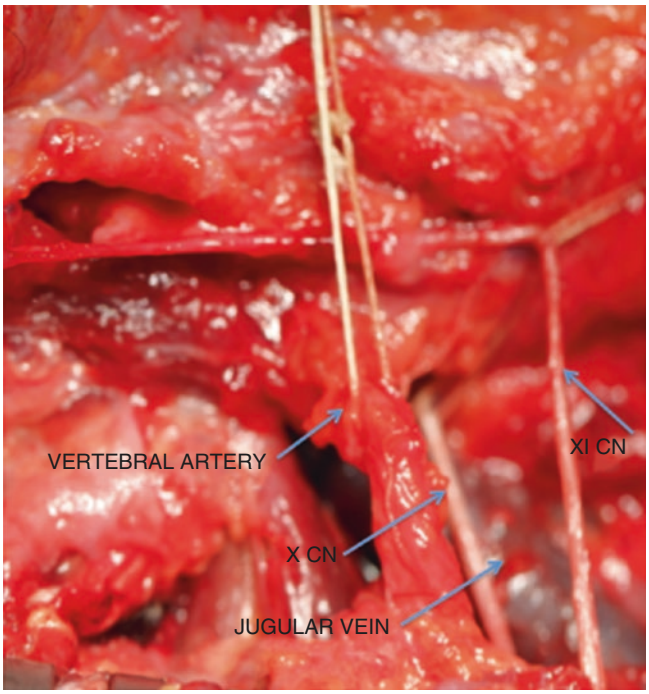

Fig. 2 Vertebral artery transposition in the extreme lateral approach

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Direct Approaches

Transoral Versus Transnasal Approach for Craniovertebral Junction Pathologies: Which Route Is Better?

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Abstract *Background:* Several pathologies that affect the craniovertebral junction (CVJ) can be treated by means of a microsurgical transoral approach (TOA) or, alternatively, with an endoscopic endonasal approach (EEA), which is potentially able to overcome some complications associated with the former approach. In this paper, after discussing updates in the recent literature, to which we add our own surgical experience, we critically analyse these procedures with the aim of demonstrating that the TOA still deserves to be considered a viable alternative and that, in selected cases, it can even be considered superior to the EEA.

Methods: Our experience involves 25 anterior procedures in 24 paediatric and adult patients (18 TOA and seven EEA). The TOA group (13 male and five female patients) encompassed three tumours, three rheumatoid arthritis cases, one condylus tertius, three basilar invaginations, four impressio basilaris cases, one developmental anomaly of C0–C1, one os odontoideum, one posttraumatic C1–C2 compression and one C2 fracture. The EEA group (three male and four female patients, median age 39 years, operated on over a 7-year period) comprised four tumours, two impressio basilaris cases and one case of impressio basilaris with platybasia.

Results: In the TOA group, all but one patient were discharged after posterior procedures within 2 weeks and improved or remained unchanged after surgery and during the follow-up period. No major complications occurred in the TOA group. In the EEA group, two patients who developed a cerebrospinal fluid (CSF) infection died, one from disease progression and the other from myocardial infarction.

Conclusion: Our data, in agreement with those from previous reports on other series, suggest that no clear superiority of the EEA over the endoscopic TOA can be postulated so far; in fact, the EEA can produce complications similar to those observed with the TOA in CVJ surgery.

Introduction

According to the current literature, the congenital, developmental and acquired pathologies that affect the anterior craniovertebral junction (CVJ) can be targeted through several surgical approaches:

1. The transoral approach (TOA), which provides the widest and shortest [1] route
2. The endoscopic endonasal approach (EEA), which was recently introduced by Kassam [2–5] with the goal of overcoming some technical challenges and surgical complications secondary to soft tissue swelling
3. The transcervical approach

In fact, the indications for the transcervical approach are not recognized worldwide, although it may still be considered in highly selected cases; as to whether the EEA is superior to the TOA in the treatment of similar CVJ pathologies, this is still a matter of debate.

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In this paper we describe our surgical experience with the TOA and EEA for different CVJ pathologies and discuss the relevant literature, seeking to demonstrate that despite the wide dissemination of the EEA, the TOA deserves and should receive greater consideration in the surgical decision-making process, and that in some cases it can even be considered superior to the EEA.

Material and Methods

Among 25 anterior procedures (in 24 patients) performed from 2011 to 2017, a consecutive series of seven patients with different CVJ pathologies were treated with the EEA and 18 with the TOA by a mixed team of neurosurgeons and rhinolaryngologists, who are expert in microneurosurgery and endoscopic video-assisted microsurgery (Table 1).

An occipitocervical instrumentation and fusion procedure was completed according to our protocol for CVJ tumours and platybasia—and to prevent cranial settling of the C2 vertebral body—1 week later, using a titanium implant (DePuy Synthes Spine Mountaineer®) in accordance with our patients' wishes. All patients underwent magnetic resonance imaging (MRI), computed tomography (CT) scanning and standard/dynamic X-ray evaluation of the CVJ before surgery, immediately after surgery and every 6 months thereafter.

Results

Our recent institutional experience is summarized in Table 1. All except one patient were discharged after the posterior instrumentation and fusion procedure within 2 weeks after admission and improved or remained unchanged (with a good neurological status) after surgery and during the follow-up period. No major complications were observed in the TOA group. On the other hand, in the EEA group, two patients who developed a cerebrospinal fluid (CSF) infection died, one from disease progression and the other from myocardial infarction.

Discussion

Among the seven patients who underwent the EEA, six had an uncomplicated postoperative course, but one developed an intraoperative CSF leak and subsequent meningitis, and eventually died 5 weeks after the surgery; none of the TOA group experienced the same complication. *Such an event was probably due to the limitation of the bidimensional view afforded by the oblique surgical view in the EEA, as opposed to the tridimensional transoral microsurgical view afforded by the straightforward surgical view in the TOA.*

According to the current literature, a total of 120 patients (including our seven) affected by CVJ disease and treated with the EEA have been reported so far. In a previous analysis [6] published in 2017—at which stage, the literature included a total of 107 patients treated with the EEA—we noted that CSF leakage (intra- and/or postoperative) had been reported in 13 patients (12.4%); transient velopharyngeal incompetence, variably associated with nasal speech and swallowing impairment, in 6 patients (5.6%); postoperative epistaxis in 2 patients (1.86%); and respiratory dysfunction requiring a tracheostomy in 2 patients (1.86%). Moreover, a recent systematic review and meta-analysis [7] showed increased prevalence rates of several complications in the EEA group in comparison with the TOA group (30-day mortality 4.4% versus 2.9%, intraoperative CSF leakage 30% versus 0.3%, postoperative CSF leakage 5.2% versus 0.8%, meningitis 4% versus 1%, need for prolonged intubation or re-intubation 6% versus 5.6%, need for reoperation 5.1% versus 2.5%, velopharyngeal insufficiency 6.4% versus 3.3% and sepsis 7.7% versus 1.9%). Conversely, the same paper also showed increased prevalence rates of other complications in the TOA group in comparison with the EEA group (arterial injury 1.9% versus 0%, wound infection 3.3% versus 1.9% and need for tracheostomy 10.8% versus 3.4%). However, none of these differences was statistically significant, except for the difference in the need for postoperative tracheostomy [7].

According to the available literature and our personal experience, the presumed improved safety of the EEA in comparison with the TOA requires reassessment [8, 9] even though it is regarded worldwide as being "minimally invasive" [10] .

Table 1 Personal series of patients operated by means of an anterior approach to the CVJ

Pts	Age (sex)	Primary disease	Radiology	Pre-op C1–C2 shift (X-rays)	Treatment	Post-op shift	Frankel scale and Di Lorenzo grade changes	External orthosis	Follow-up (months)
1 SO	26 (F)	CVJ chordoma	C0–C2 anterior compression CVJ instability	Virtual	1. Transoral decompression 2. C0–C3 reduction, lateral masses screws instrumentation and heterologous bone fusion	No	E/E, I/I	Philadelphia (1 month)	88
2 FF	33 (M)	CVJ chordoma	C0–C2 anterior compression CVJ instability	Virtual	1. Transoral decompression 2. C0–C3 reduction, C2 pedicles, and C3 lateral masses screws instrumentation and heterologous bone fusion	No	E/E, I/I	Philadelphia (1 month)	99
3 CO	68 (F)	Rheumatoid arthritis	Anterior C1–C2 compression C1–C2 instability	>5 mm	1. Transoral decompression 2. C0–C2 pedicles and lateral masses screws instrumentation	No	D/E, II/I	Soft collar (1 month)	82
4 CL	15 (M)	Developmental anomaly C0–C1	C0–C1 anterior compression C1–C2 instability	>5 mm	1. Transoral decompression 2. C1 laminectomy, C0 double vertical screws, C2 pedicles, and C3 lateral masses screws instrumentation	No	D/E, II/I	Soft collar (1 month)	77
5 CA	78 (M)	Chordoma (chondroid)	CVJ instability C0–C2 Ant. compression	Virtual	Transoral C1-odontoidectomy and clivectomy, C0-double vertical screws, C2, C3, C4, and C5-lateral masses screws instrumentation	No	D/E, II/I	Soft collar (1 month)	74
6 EA	11 (M)	Impressio basilaris Os odont. (Downs.)	C1–C2 anterior compression	Virtual	Transoral C1-odontoidecotmy and clivectomy in C0–C2–C3 screwing instrumentation and heterologous bone fusion (previously implanted)	No	D/E, II/I	Soft collar (1 month)	73
7 RR	14 (M)	C2 fracture and dislocation	C2 fracture and C1–C2 dislocation with cervic. contusion	>7 mm	1. Transoral C1–C2 decompression 2. C0–C3–C5 screwing instrumentation and heterologous bone fusion	No	D/E, II/I	Soft collar (1 month)	70
8 MP	8 (M)	Impressio basilaris	C0–C1–C2 compression	No	1. Transoral C1–C2 decompression 2. Staged C0–C2–C3 screwing instrumentation and fusion	No	D/E, II/I	Halo + Philadelphia (2 months)	66
9 LC	15 (M)	Third condyle	C0–C1 compression	No	1. Transoral C0–C1 decompression 2. Staged C0–C2–C3 screwing instrumentation and fusion	No	D/E, II/I	Halo + Philadelphia (3 months)	103
10 CP	67 (F)	Rheum. arthritis C2 fracture + dislocation 2 cm	C2 compression + myelopathy	Yes	1. C1–C2 decompression combined 2. C0–C2–C3 screwing instrumentation and fusion	No	C/E, III/I	Philadelphia (2 months)	8

(continued)

Table 1 (continued)

Pts	Age (sex)	Primary disease	Radiology	Pre-op C1–C2 shift (X-rays)	Treatment	Post-op shift	Frankel scale and Di Lorenzo grade changes	External orthosis	Follow-up (months)
11 PP	50 (M)	Basilar invagination	C2 anterior compression	Virtual	1. One stage transoral decompression 2. C0–C3 instrumentation and fusion	No	D/E, II/I	Soft collar (1 month)	15
12 SP	7 (M)	Basilar invagination	C0–C1–C2 compression	Virtual	1. Transoral C0–C1–C2 decompression 2. C0–C2–C3 screwing instrumentation and fusion	No	D/E, II/I	Soft collar (3 months)	68
13 SS	9 (M)	Os odontoideum	C2 compression	Virtual	1. Transoral C1–C2 decompression 2. C0–C2–C3 screwing instrumentation and fusion	No	D/E, II/I	Halo + Philadelphia (2 months)	121
14 MM	68 (F)	Rheumatoid arthritis + ED C2–C3	C2–C3 compression	Virtual	1. Transoral C0–C1–C2 decompression 2. C0–C2–C3 screwing instrumentation and fusion	No	D/E II/I	Halo + Philadelphia (3 months)	12
15 AE	12 (M)	Basilar invagination + ED C3–C4 myelopathy	C2 compression	Virtual	1. Transoral C0–C1–C2 decompression 2. Anterior C3–C4 discect. and titanium graft 3. C0–C2–C3 screwing instrumentation and fusion	No	D/E, II/I	Halo + Philadelphia (3 months)	3 (death for heart attack)
16 GC	(F)	Impressio basilaris + Chiari	C0–C1–C2 compression	Virtual	1. Transoral C0–C1–C2 decompression 2. C0–C2–C3 screwing instrumentation and fusion	No	II/I	Halo (3 months)	23
17 MS	(M)	Impressio basilaris	C0–C1–C2 compression	Virtual	1. Transoral C0–C1–C2 decompression 2. C0–C2–C3 screwing instrumentation and fusion	No	II/I	Miami (2 months)	5
18 ML	56 (M)	Posttraumatic C1–C2 compression	C1–C2 compression	Virtual	1. Transoral C0–C1–C2 decompression 2. C0–C3 screwing instrumentation and fusion	No	D/E, II/I	Soft collar (1 month)	6
19 SO	35 (F)	Chordoma	CVJ infiltration (previous TOA and C0–C3 fusion)	NP	Transnasal decompression (0°–30°)	No	D/D, I/I	Previously performed	39

20 FP	66 (F)	Impressio basilaris	CVJ anterior compression	Yes	1. Transnasal decompression (0 °–30°) 2. C0–C3 reduction, lateral masses screws instrumentation and heterologous bone fusion	No	D/E, I/I	Halovest (1 month)	3 (meningitis and death)
21SR	47 (F)	Impressio basilaris + platybasia	CVJ anterior compression	Yes	1. Transnasal C1 decompression and odontoidectomy 2. C0–C3 reduction, C2 pedicles, and C3 lateral masses screws instrumentation and autologous bone fusion	No	D/E, I/I	Halovest (1 month)	52
22 PLP	14 (F)	Impressio basilaris + platybasia	CVJ anterior compression	No	Transnasal C1 partial decompression and partial odontoidectomy	No	D/E, I/I	No	54
23 NR	22 (M)	Recurrence of clivus chordoma	CVJ destruction and compression	NP	1. Transnasal C1 decompression and odontoidectomy 2. C0–C3 reduction, lateral masses screws instrumentation and heterologous bone fusion	No	D/E, I/I	Halovest (1 month) Hard Collar (2 months)	37
24 MG	58 (M)	Myeloma	Odontoid infiltration	NP	Transnasal and odontoid biopsy	No	E/E, I/I	No	30
25 RC	36 (M)	Isolated plasmacytoma	CVJ anterior compression	NP	Transnasal C1 decompression, and odontoidectomy and C0–C3 reduction, lateral masses screws instrumentation and heterologous bone fusion	No	D/D, I/I	Soft collar (1 month)	3

PTS patients, *CVJ* craniovertebral junction, *NP* not performed

Conclusion

No clear superiority of the endoscopic endonasal approach (EEA) over the endoscopic transoral approach (TOA) is recognized by the current literature so far; in fact, the EEA has been shown to result in complication rates similar to those observed with the TOA in craniovertebral junction surgery.

In normal anatomical conditions, superior surgical freedom is provided by the TOA in comparison with the EEA. The respective roles of the two different strategies is still a matter of debate, requiring more cohort and prospective studies, along with detailed and conclusive meta-analysis.

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Competing Interests The authors declare that they have no competing interests.

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The Endonasal Endoscopic Approach to Pathologies of the Anterior Craniocervical Junction: Analytical Review of Cases Treated at Four European Neurosurgical Centres

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Abstract Supported by preliminary anatomical and clinical studies exploring the feasibility and usefulness of approaching many ventral pathologies of the craniocervical junction (CCJ) using the endoscopic endonasal approach, four European centres have joined forces to accumulate and share their growing surgical experience of this advanced technique. By describing the steps that led to the development and continuous refinement of this approach to the CCJ, this article delves deeply into an analysis of the cases operated on since 2010 at these four institutions, and discusses in detail the operative nuances that so far have allowed achievement of successful outcomes with excellent perioperative patient comfort and satisfactory long-term quality of life.

Keywords Endoscopy · Endonasal · Transoral · Odontoidectomy · Basilar impression/invagination · Odontoid pannus

Introduction

Over the years, many ventral approaches to the craniocervical junction (CCJ) have been developed to address bulbomedullary compression [1, 2] caused by several degenerative, traumatic, congenital and neoplastic spinal pathologies. These include, but are not limited to, (1) chronic inflammation of the CCJ osteoligament complex, mostly related to rheumatoid arthritis and metabolic disorders; (2) traumatic C1–C2 dislocations, resulting in basilar invagination; (3) congenital malformations, causing instability and/or stenosis at the level of the CCJ, such as those resulting from collagenopathies, osteogenesis imperfecta, Down's syndrome and achondroplasia; and (4) neoplastic lesions (i.e. primary and secondary spinal tumours) and paraneoplastic lesions (i.e. Paget's disease), usually affecting the body and dens of C2. Although the standard transoral approach provides wide access to this anatomical region, some complications have highlighted the need to redefine the surgical strategy and find alternative routes. This technique, in fact, requires splitting of the soft palate and provides a wide but deep working channel; furthermore, it is affected by the risks of teeth traumatism, cerebrospinal fluid (CSF) leakage, bacterial contamination, tongue swelling and nasopharyngeal incompetence, often requiring prolonged intubation and nasogastric tube feeding. In summary, the more invasive the pathology, the longer the operative time and the riskier the extended procedure [3–12].

Since 2005 a new endoscopic technique exploiting the natural nasal corridor has been described and implemented for surgical management of extradural or intradural pathologies involving the ventral CCJ. Fairly rapidly, the endoscopic endonasal approach (EEA) appeared to be potentially promising in overcoming previous technical challenges and surgical complications, gaining wide attention with overwhelmingly positive opinions. Following a detailed description of the anatomical principles behind the development of this approach to CCJ pathologies, in this article we provide a critical review of the cases operated on since 2010 at four European institutions, and we delve deeply into the operative nuances that so far have allowed achievement of successful outcomes with excellent perioperative patient comfort and satisfactory long-term quality of life.

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Anatomical Aspects of the Endoscopic Endonasal Approach

Following the success of endoscopic approaches to the anterior cranial fossa [13, 14], initial anatomical studies on the implementation of the endoscopic endonasal approach to the CCJ were conducted by many groups, so this surgical technique rapidly gained wide attention and enthusiastic support even before the translation of these laboratory findings into clinical practice (see Fig. 1) [5, 15–26].

Fries and Perneczky were the first to envisage the possibility of using video endoscope instrumentation at the craniocervical junction, and they reported some cases treated with this surgical strategy among 380 endoscope-assisted microneurosurgical operations performed in a 4.5-year series [21]. In 2002, Alfieri et al. performed a cadaver study of exclusively transnasal endoscopic odontoidectomy through one or two nostrils; the surgical landmarks leading to the CCJ were the inferior margin of the middle turbinate, the nasopharynx and the Eustachian tubes. The authors described a line drawn between the Eustachian tubes as the line indicating the juncture between the clivus and atlas, and they demonstrated that the EEA could provide unlimited access to the midline clivus and a potential surgical window for decompression at the CCJ without the risk of causing C1–C2 instability [15]. Three years later, Cavallo et al. confirmed such observations in a cadaver study [27], and in 2005, Kassam et al. described the first case of rheumatoid arthritis treated with EEA-based odontoidectomy [22], suggesting that—being above the level of the soft palate and therefore characterized by the less aggressive microbic flora of the nostrils rather than the oral microbic flora—the EEA exposed patients to a lesser degree of bacterial contamination.

Later, de Almeida et al. defined the radiological concept of the nasopalatine line (NPL), a line drawn on the midsagittal plane and connecting the inferior margin of the nasal bone, anteriorly, and the border of the hard palate, posteriorly, to calculate the inferior limit of the endoscopic approaches to the spine [5]. This concept was then reappraised and more reliable surgical concepts such as the nasoaxial line (NAxL) and the rhinopalatine line (RPL) were introduced to overcome the imprecision of the NPL, which does not take into account the dorsal nose skin between the nostrils and the nasal bones [23, 28].

More recently, some pioneering clinical series based on the EEA have pinpointed the feasibility of successfully approaching the CCJ through the natural nasal corridor. Because of its intrinsic features, this route provides a panoramic and multiangled view of the region, allowing a close-up view without the need for prolonged tongue compression or mouth retractors. Eventually, this approach became widely recommended as a valid alternative to address CCJ pathologies in both adult and paediatric populations, mostly because it is more respectful of the oropharynx and adjacent structures, and it is not associated with dysphonia and dysphagia, which are usually reported after transoral approaches.

Besides the anatomical studies, the introduction of the EEA into the surgical armamentarium was favoured also by the widespread use of technological aids such as neuronavigation systems, which are helpful in visually reconstructing magnified three-dimensional anatomy and permitting better orientation during the whole surgical procedure. Although the error associated with spinal shift is not completely eliminated, at present the calculated accuracy is less than 1 mm [19, 26].

Patients and Methods

A prospective multicentre study was conducted from June 2010 to May 2016 on a cohort of 24 consecutive patients undergoing pure endoscopic endonasal odontoidectomy for primary surgical management of various CCJ diseases responsible for irreducible atlantoaxial dislocation. All patients had pre- and postoperative clinical and imaging assessments (cervical computed tomography [CT] and

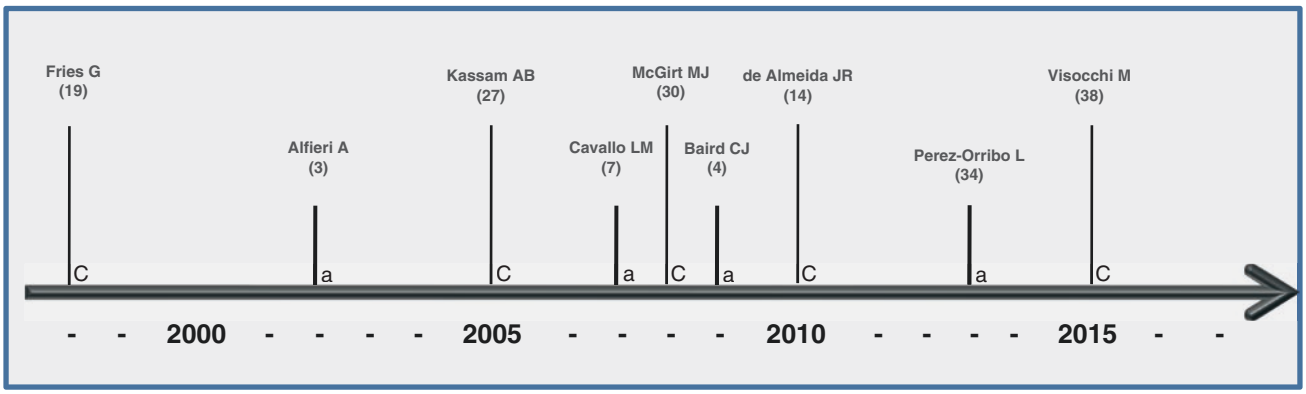

Fig. 1 Time-line of clinical and anatomical landmark papers. *c* clinical; *a* anatomical

magnetic resonance imaging [MRI] scans). The initial follow-up was scheduled at 6 months and repeated once every year. The study was reviewed and approved by the institutional research boards (IRBs) of the Strasbourg, Foch (Suresnes–Paris) and Catholic University of Rome hospitals, and suitable surgical candidates were informed about the various surgical options and invited to participate in the present study; this involved provision of information about the study design, the intended benefits and the expected outcomes of the study itself. Informed consent forms, specifically prepared by the authors and approved by the aforementioned IRBs, were signed by all patients participating in this prospective trial.

Surgical Technique

Our surgical strategy for the endoscopic endonasal approach to the CCJ has been described elsewhere [29]; nonetheless, it seems appropriate to highlight some operative tips and tricks, which are useful for all those who decide to consider this technique and possibly introduce it into their surgical practice. It is important to pay attention to the position of the patient on the surgical table and to personally set up all instruments beforehand. We always prefer a recumbent and slight Trendelenburg position (20°) with the head fixed in a Mayfield three-pin headholder; the screen of the neuronavigation system, for which we always recommend use of CT and MRI image fusion, can be positioned cranially to the patient and just laterally to the endoscope monitor. Neuromonitoring with somatosensory evoked potentials, although not always mandatory, can play an important role, depending on the degree of bulbomedullary compression.

Preoperative antibiotic prophylaxis is routinely administered 20 min before the start of the operation; it may be wise to administer a second dose whenever long and complex debulking of space-occupying lesions (SOLs) at the CCJ protracts the surgery beyond 4 h. The nostrils are routinely prepared with iodine 5% and naphazoline. In our practice, we use 0° and 30° angled endoscopes with a HD camera (Storz, Tuttlingen, Germany) and dedicated endoscopic tools (Storz), with either a four-hand or two-hand technique coupled with a fixed endoscope holder; this choice relies solely on the surgeon's skills and preferences, and it can be adapted to the pathology being treated.

The surgical corridor is created through an inferior septectomy, removing no more than 2 cm of vomer bone at its junction with the hard palate; whenever additional space for surgical manoeuvres is needed, a proper sphenoidotomy can be considered.

Besides the intraoperative guidance of the neuronavigation pointer, we rely on a few fundamental anatomical landmarks: (a) the clivus–septum junction superiorly; (b) the Eustachian tubes laterally; and (c) the RPL inferiorly. To provide a mucopharyngeal flap, we create an inverted U-shaped flap, which can be done by use of a diode laser or monopolar electrocautery. This U-shaped flap goes from the level of the sphenoid floor to the level of the soft palate and can be caudally reflected into the oropharynx during the bony removal and replaced at its original site at the end of the procedure. A high-speed drill can be safely used to facilitate the surgical decompression, which requires a great deal of attention to avoid CSF leakage, vascular damage and, in cases of remarkable stenosis, anterior herniation of the medulla following odontoidectomy. The brainstem decompression can be considered adequate only when wide exposure of pulsatile dura is achieved. In this regard, Goldschlager et al. [30] have proposed recording pre- and postoperative T2-weighted MRI sequences of the CSF thickness at the dens midline level, showing that good decompression is achieved when an increased thickness of 2.34 ± 0.43 mm is obtained.

Reconstruction is always fundamental to an uneventful healing process: following odontoidectomy (with or without removal of SOLs) it is recommended to create a multilayered closure, including a Gelfoam/Surgicel pack, overlaid by a pedicled regional paraspinal muscle and the mucopharyngeal flap, sealed together by fibrin glue. This closure can be supplemented and reinforced with fascia lata and fat whenever the risk of postoperative CSF leakage is deemed high by the surgeon. Finally, a Doyle splint placed inside each nostril at the end of the procedure can be kept in place for 4–5 days and is helpful in preventing endonasal mucosal adherence.

Results

Among the 24 patients included in this study, 13 were men and 11 were women, with a mean age 51.7 years (range 14–95 years). The mean follow-up period was 30.2 months (range 3–78 months); 12 patients presented with basilar impression, seven had a degenerative pannus, four had CCJ chordomas (one was a recurrence) and one had a myeloma. The patients' demographic, clinical, radiological, complication and outcome features are summarized in Table 1). All patients had satisfactory neurological improvement, and the quality of decompression was excellent in all cases. Posterior stabilization was performed in 20 of the 24 cases during the same operation. In one patient (a 95-year-old) we opted for a rigid collar for 3 months, with an uneventful postoperative course and excellent outcome. Stability was preserved in three patients who underwent either a partial odontoidectomy (case no. 21), a simple biopsy (case no. 23) or limited debulking (case no. 24).

Table 1 Operative series of 24 patients: demographic, clinical, radiological, complication, and outcome features

No.	Age and sex	Primary disease	Radiology	Pre-op C1–C2 shift (X-rays)	Pre-op Frankel scale	Management	Post-op shift (X-rays)	Post-op Frankel scale	Post-op external orthosis	F-U mths pre-/postoperative complications
1	20 M	Basilar impression	Brain stem compression	CCJ malformation	*C*	Endonasal endoscopic C0–C2 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	15
2	36 M	Basilar impression	Brain stem compression	CCJ malformation	*D*	Endonasal endoscopic C0–C2 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	12
3	48 M	Basilar impression	Brain stem compression	CCJ malformation	*C*	Endonasal endoscopic C0–C2 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	D	No	30
4	56 M	Basilar impression	Brain stem compression	Large shift with brain stem compression and posterior deviation	*C*	Endonasal endoscopic C0–C2 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	48 Preoperative CSF leak
5	58 M	Basilar impression	Brain stem compression	Large shift with brain stem compression and posterior deviation	*B*	Endonasal endoscopic C0–C2 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	D	No	30 Preoperative blood loss needing transfusion
6	67 F	Basilar impression	Brain stem compression	Large shift with brain stem compression and posterior deviation	*D*	Endonasal endoscopic C0–C2 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	24
7	68 M	Degenerative disease	Brain stem compression	Large shift with brain stem compression and posterior deviation	*C*	Endonasal endoscopic C0–C2 decompression with odontoidectomy *and* C1–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	16
8	73 F	Degenerative disease	Brain stem compression	Large shift with brain stem compression and posterior deviation	*D*	Endonasal endoscopic C0–C2 decompression with odontoidectomy *and* C1–C2 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	78 Serous otitis needing ENT drainage—good outcome

No.	Age and sex	Primary disease	Radiology	Pre-op C1–C2 shift (X-rays)	Pre-op Frankel scale	Management	Post-op shift (X-rays)	Post-op Frankel scale	Post-op external orthosis	F-U mths pre-/postoperative complications
9	84 F	Degenerative disease	Brain stem compression	Large shift with brain stem compression and posterior deviation	*C*	Endonasal endoscopic C0–C2 decompression with odontoidectomy *and* C1–C2 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	9 Pneumonia—good outcome
10	95 F	Degenerative disease	Brain stem compression	Large shift with brain stem compression and posterior deviation	C	Endonasal endoscopic C0–C2 decompression with odontoidectomy leaving anterior C1 arch intact, no posterior instrumentation/fusion	No	E	Yes	30 Pneumonia—good outcome
11	28 F	CVJ skull base chordoma	Brain stem compression by tumor invasion of C0–C2	Brain stem compression, unstable on dynamic X-rays	C	Endonasal endoscopic C0–C2 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	48
12	59 F	Basilar impression	Extensive brain stem compression by C2 odontoid process	Large shift with brain stem compression and posterior deviation	D	Endonasal endoscopic C1 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	36
13	72 M	Degenerative compression	Extensive brain stem compression by hypertrophied "C2 odontoid process"	Large shift with brain stem compression and posterior deviation	D	Endonasal endoscopic C1 decompression with odontoidectomy *and* C1–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	D	No	32
14	85 M	CVJ skull base chordoma	Brain stem compression	Large shift with brain stem compression and posterior deviation and instability	C	Endonasal endoscopic C1 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	26 Preoperative CSF leak
15	38 M	Basilar impression	Extensive brain stem compression by C2 odontoid process	Large shift with brain stem compression and posterior deviation	C	Endonasal endoscopic C1 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	24
16	77 M	Degenerative compression	Extensive brain stem compression by hypertrophied "C2 odontoid process"	Large shift with brain stem compression and posterior deviation	C	Endonasal endoscopic C1 decompression with odontoidectomy *and* C1–C2 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	D	No	22

(continued)

Table 1 (continued)

No.	Age and sex	Primary disease	Radiology	Pre-op C1–C2 shift (X-rays)	Pre-op Frankel scale	Management	Post-op shift (X-rays)	Post-op Frankel scale	Post-op external orthosis	F-U mths pre-/postoperative complications
17	51 F	Degenerative compression	Brain stem compression	Large shift with brain stem compression and posterior deviation	D	Endonasal endoscopic C1–C2 decompression with odontoidectomy *and* C1–C2 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	26
18	27 M	Basilar impression	Brain stem compression	Large shift with brain stem compression and posterior deviation	C	Endonasal endoscopic C0–C3 decompression with odontoidectomy *and* C0–C3 reduction, lateral mass screw instrumentation and autologous (iliac crest) bone fusion *at the same stage procedure*	No	E	No	31
19	66 F	Basilar impression	CVJ anterior compression	Large shift with brain stem compression and posterior deviation and instability	D	1) Transnasal decompression (0°–30°) 2) C0–C3 reduction, lateral mass screw instrumentation and heterologous bone fusion	No	E	Yes	3 Death (meningitis)
20	47 F	Basilar impression and platybasia	CVJ anterior compression	Large shift with brain stem compression and posterior deviation and instability	D	1) Transnasal C1 decompression and odontoidectomy 2) C0–C3 reduction, C2 pedicles, and C3 lateral mass screw instrumentation and autologous bone fusion	No	E	Yes	46
21	14 F	Basilar impression and platybasia	CVJ anterior compression	Not performed	D	Transnasal C1 partial decompression and partial odontoidectomy	No	E	No	48
22	22 M	Recurrence of clivus chordoma	CVJ destruction and compression	Not performed	D	1) Transnasal C1 decompression and odontoidectomy 2) C0–C3 reduction, lateral mass screw instrumentation and heterologous bone fusion	No	E	Yes	31
23	58 M	Myeloma	Odontoid infiltration	Not performed	E	Transnasal and odontoid biopsy	No	E	No	24
24	35 F	Chordoma	CVJ infiltration previously operated: (1) transoral and (2) C0–C3 reduction, lateral masses	Not performed	D	Transnasal decompression (0°–30°)	No	D	Previously performed	36

Pre- and Post-op pre- and postoperative, *F-U mths* follow-up months

One patient (case no. 19) died from meningitis 3 months after the surgery. In the remaining 23 patients, neither new neurological deficits nor postoperative CSF leakage were observed, despite two cases (case nos. 4 and 14) requiring intraoperative repair of dural tears; no patient required a tracheostomy or gastrostomy.

The postoperative Frankel grade was 'E' in 19 patients and 'D' in the remaining five. No postoperative X-ray listhesis was recorded; in four patients an external orthesis was required, while in the other 20 patients who were stabilized, no postoperative external orthesis was deemed necessary.

Discussion

The agglomerated data from our centres confirm that the endoscopic endonasal approach to the CCJ can be safe and effective in many different pathologies and surgical scenarios. In our experience it is possible to address pathologies as high as those involving the midclivus without extensive soft- or hard-palate manipulation. We previously confirmed that the NAxL and RPL can be reliable preoperative predictors of the maximal extent of inferior dissection for the EEA [26] and pointed out that in the case of very lateralized lesions, or whenever anatomical variations with an aberrant course of the internal carotid artery (ICA) and vertebral artery are preoperatively identified, the EEA is either contraindicated or very difficult to perform, with a high morbidity risk [22, 29, 31, 32]. We noted that in the case of quite a low junction, generally far below the level of the hard palate, it is almost impossible to remove the dens via the endonasal route; on the other hand, in the presence of a higher CCJ, the dens is more easily reachable and safely removable via the nasal route [15, 25, 27, 29, 32–38]. Furthermore, it is important to mention that more specific indications for preferring the EEA over the transoral route are the following: children (with a narrow mouth) [24]; cases of micrognathia and macroglossia (bearing in mind that the mouth opening for a transoral approach should always be at least 25 mm); and patients with osteogenesis imperfecta, as 25% of them have basilar impression [35]. In Table 2 we highlight the most important advantages, risks and limitations of the endoscopic endonasal approach to the CCJ; it seems appropriate, however, to delve deeper into the management of complications: haemostasis, CSF leakage and CCJ instability.

Achieving satisfactory haemostasis during the EEA could constitute a major concern because it is often difficult to control bleeding through the endonasal surgical corridor, especially in the case of SOLs; for this reason we believe that this procedure requires dedicated bipolar forceps and use of haemostatic products such as Gelfoam and activated thrombin matrix.

Table 2 Advantages, risks, and limitations of EEA

Advantages	Ref.
Better and wider angled views, preservation of palatal function	Kassam et al. [22]
Reduced frequency of tracheostomy, decreased postoperative pain, and decreased risk of meningitis following CSF leakage	Kassam et al. [22]
Prevention of velopharyngeal incompetence, edema or tongue necrosis, dysphagia, odynophagia, teeth injuries, and pharyngeal cellulitis which can complicate transoral approaches to CCJ	Zenga et al. [39]
Risks	
ICA injury (which could result in severe stroke)	Gardner et al. [31]
Limitations	
Spheric aberrant artifacts with loss of stereoscopic perception when using 2D endoscopes	Ponce-Gomez et al. [37]
Steep learning curve with required anatomical training in wet laboratory	Alfieri [15] Kassam et al. [22] Cavallo et al. [17]

Intraoperative identification of CSF leakage can be easier under endoscopic guidance because it allows closer-up and multiangled vision, and the degree of dexterity to stitch up the dura can be much greater. For this purpose, we have already stressed the importance of creating a well vascularized muscle–mucosal flap overlying the ventral CCJ, and considering a lumbar drain during the initial postoperative days whenever the risk of CSF leakage is too high and can compromise the healing process of the multilayered closure flap.

Finally, it is fundamental to bear in mind that removal of the anterior arch of C1 and the dens of C2 can cause destabilization of the CCJ, obliging the surgical team to plan for an occipitocervical arthrodesis. Leaving intact the anterior arch of C1 is usually quite demanding, and some authors [34, 39, 40] have proposed technical variations to keep it intact during odontoidectomy. Although we reckon that this choice can be considered in paediatric or elderly populations, in selected rheumatoid pannus cases and in low-lying lesions involving the dens and body of C2, we believe that posterior occipitocervical instrumentation with C1–C2 or C1–C3 stabilization should be considered as the safest way to ensure postoperative fusion and preserve the range of occipitocervical rotational movements.

Conclusion

In our series, the endoscopic endonasal approach (EEA) allowed for fast recovery with excellent perioperative patient comfort and a satisfactory long-term quality of life. Our experience seems to support the leading theory that the EEA provides a direct surgical corridor to the craniocervical joint (CCJ) and allows a degree of decompression similar to that offered by the more invasive transoral route. Nonetheless, prospective randomized studies are certainly warranted to establish whether the EEA can be definitely considered superior whenever an approach to the ventral CCJ is needed.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

All procedures performed in the study were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

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Endoscopic Endonasal Odontoidectomy and Posterior Fusion in a Single-Stage Surgery: Description of Surgical Technique and Outcome

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Introduction

An anterior approach to the craniovertebral junction (CVJ) and, particularly, to the odontoid process of the second cervical vertebra has classically been performed, in neurosurgical settings, via a transoral route. Such a technique is still considered the gold-standard treatment for odontoid process diseases.

However, the advent of endoscopy in neurosurgery and the development and refinement of endonasal approaches to the entire midline skull base [1–5] have meant that this field, once dominated by microsurgery, has become a territory of exploration for neurosurgeons, who have dedicated clinical and scientific efforts in this direction. In fact, the endoscopic endonasal approach to the craniocervical junction, and to the odontoid process, is among the areas of most interest for which endoscopic techniques have been developed.

Several anatomical and/or clinical studies have been reported, showing the interest of approaching the craniocervical junction (CCJ) through the nasal corridor [6, 7]. The availability of new technologies—such as endoscopes, high-definition endoscopic cameras, navigation systems, ultrasound micro-Doppler, dedicated endonasal instruments and bipolar forceps—has opened new horizons for management of pathologies involving this complex region, using the natural nasal corridors; this way/approach has demonstrated remarkable improvements in the quality of disease resection and in functional outcomes with lower morbidity.

The endonasal route provides direct access to the surgical field, minimizing the mucosal and neurovascular manipulation: it follows a natural path that goes from the nostrils to the mucosa covering the rhinopharynx, the rhinopharyngeal muscles, the anterior arch of C1 and, finally, the odontoid process. As a consequence, the endoscopic endonasal approach is less invasive and does not require additional surgical manoeuvres such as (1) mouth retraction; (2) tongue compression or even splitting; (3) possible injury to the teeth; (4) injury to the uvula and/or the soft palate and velum pendulum; or (5) neurovascular manipulation through the oropharynx. Theoretically, such facts imply a lower rate of postoperative complications related to invasiveness, with lower rates of postoperative dysphagia and respiratory complications, possibly due to the fact that with the endoscopic approach, extubation coincides with the end of the procedure. This allows more rapid mobilization and a reduction in the recovery time for natural feeding, which is then reflected, of course, in the hospitalization time. Seen in this light, the endoscopic endonasal approach offers a viable alternative to the more established transoral approach, especially for the clear advantages that the endoscopic technique offers in cases where there is a full indication to execute it. On the other hand, in cases of dural opening the endonasal approach is associated with difficulty of dural closure, with associated higher risks of postoperative cerebrospinal fluid (CSF) leakage and meningitis. Given the intrinsic features of the endoscope, the endonasal route provides a wider, panoramic and multiangled view of the region and also allows close-up views of the relevant anatomical structures on the surgical field.

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Anterior Versus Posterior Approach

The decision making between an anterior or a posterior approach depends on different particular aspects: (1) the direction of the compression; and (2) the surgeon's confidence and experience with the approaches, and thus the possibility to perform reduction of the compression with an anterior, posterior or combined approach. In general, irreducible anterior subluxation associated with spinal cord compression requires an anterior approach, whereas reducible posterior compression requires a posterior surgical route. However, different complex diseases, acquired or congenital, can cause an alteration of atlantoaxial relationships and anterior cervicomedullary junction compression. In these cases, fixation or posterior stabilization may be not sufficient to resolve the ventral compression. In fact, in recent years, the option of a combined anterior and posterior approach has become the best choice according to many authors.

Transoral Approach and Transnasal Approach

Several surgical routes have been described for the craniovertebral junction (CVJ) region because of its complex anatomy and vital surrounding structures. During recent decades, the transoral approach with microscopic assistance has been proposed as the standard procedure for performing anterior odontoidectomy, considering the aetiology of the disease, the mechanism of compression and, finally, its reducibility [8–11]. The transoral approach has been considered the gold-standard approach for the surgical treatment of pathologies of the anterior CVJ. Specifically, in the absence of spinal cord contusion or progressive myelopathy, posterior decompression and fusion are sufficient to achieve an acceptable outcome. Odontoidectomy is necessary when there is irreducible bony compression of the spinal cord or soft tissue pannus causing severe ventral compression and resulting in progressive myelopathy.

The risk of bacterial contamination, need for prolonged postoperative intubation and nasogastric tube feeding, tongue swelling and nasopharyngeal incompetence after transoral surgery have led authors to identify alternative routes to approach this region.

The anterior aspect of the craniocervical region can be exposed also via a transnasal approach, although some anatomical limitations exist. In the transnasal route, exposure of the C2 body below the odontoid process is limited by the posterior part of the hard palate; however, angled endoscopes, drills and dedicated instruments provide access downward to the lower edge of the C2 body [12–15]. On the other hand, the transoral approach is limited by the degree of mouth opening, the size of the patient's tongue, and the position of the uvula and the soft palate. The inferior limit of the access, usually the C3 vertebra, is determined by the degree of mouth opening, the size of the patient's oral cavity and the prominence of the incisors. However, also for the transoral approach, the use of angled endoscopes and instruments directs the approach superiorly, increasing the rostral access above the anterior arch of the atlas to the lower clivus and C2 [16, 17]. One of the main anatomical landmarks to consider, especially in the transoral route, is the course of the vertebral artery (VA). After ascending through the transverse foramen of the axis and atlas, approximately 15 mm from the midline, the VA courses medially along the upper surface of the posterior arch of the atlas to reach its dural entrance. It is mandatory to preserve the segment of the VA ascending between the C1 and C2 transverse processes.

Once the anterior arch of C1 is exposed, it must be drilled to expose the odontoid process of C2. Another difference between the transoral and transnasal approaches is the visualization of the ligamentous complex. For instance, the apical ligament is easily visualized directly ahead of the endoscope in the transnasal route, but in the transoral approach it is not seen until later, after removal of the odontoid process. The main step in anterior odontoidectomy is represented by the drilling of the dens. In the transnasal approach, the dens is seen directly ahead. The anterior cortical surface and core of the dens are drilled, and the cortical shell is removed. On the other hand, the base of the dens is more easily accessed for drilling by the transoral route. In addition, a different view is offered by these two approaches regarding the exposure of the upper, middle or lower clivus. The standard endoscopic transnasal transsphenoidal approach allows one to reach the upper clivus, which corresponds to the posterior wall of the sphenoid sinus. Thus, the middle and lower clivus are viewed directly ahead in the transnasal approach. The access to the middle and lower clivus generally does not require opening of the sphenoid sinus. On the other hand, in the transoral approach the middle and upper clivus are not usually accessible because this would require soft and hard palate opening, with splitting of the tongue or mandible, to gain upward angulation. However, manoeuvres such as using an angled endoscope, retracting the uvula sufficiently and opening the mouth widely provide safe access to the lower clivus.

Indications

Odontoidectomy is a procedure that is necessary in all cases in which there is impairment of the nervous structures of the craniocervical junction due to an irreducible alteration of the

relations that the odontoid process conducts with neighbouring neurovascular structures.

Various pathologies may cause atlantoaxial misalignment and bulbomedullary junction compression; among them are congenital malformations such as Arnold–Chiari malformation type II, genetic degenerative transformation such as in Down's syndrome, chronic inflammation related to rheumatoid arthritis and/or metabolic disorders, and, finally, post-traumatic alterations (Fig. 1).

The irreducibility is a crucial concept on the path that leads to the indication for surgery. In fact, several studies have confirmed that, when feasible, reduction of the compression by putting the craniocervical junction in traction and subsequent fixation—and in cases of compression due to rheumatoid pannus, posterior stabilization of the craniocervical junction—lead in some instances to improvement or even resolution of the ventral compression.

Therefore, the indications for odontoidectomy arise in all cases in which there is irreducible atlantoaxial subluxation associated with severe brainstem and/or spinal cord compression causing progressive neurological dysfunction. In most cases, the pathological process may be due to (1) irreducible basilar impression [18–23]; (2) ventral compression, as in cases of rheumatoid pannus that is not resolved after posterior stabilization [24–26]; (3) significant retroflexion of the odontoid process or basilar invagination associated with Chiari malformation [27]; (4) the presence of os odontoideum [28–30]; or (5) post-traumatic pseudoarthrosis or misalignment. Several recent experiences have enlarged the indications for endoscopic endonasal odontoidectomy to treat intradural lesions [3, 5, 31–33].

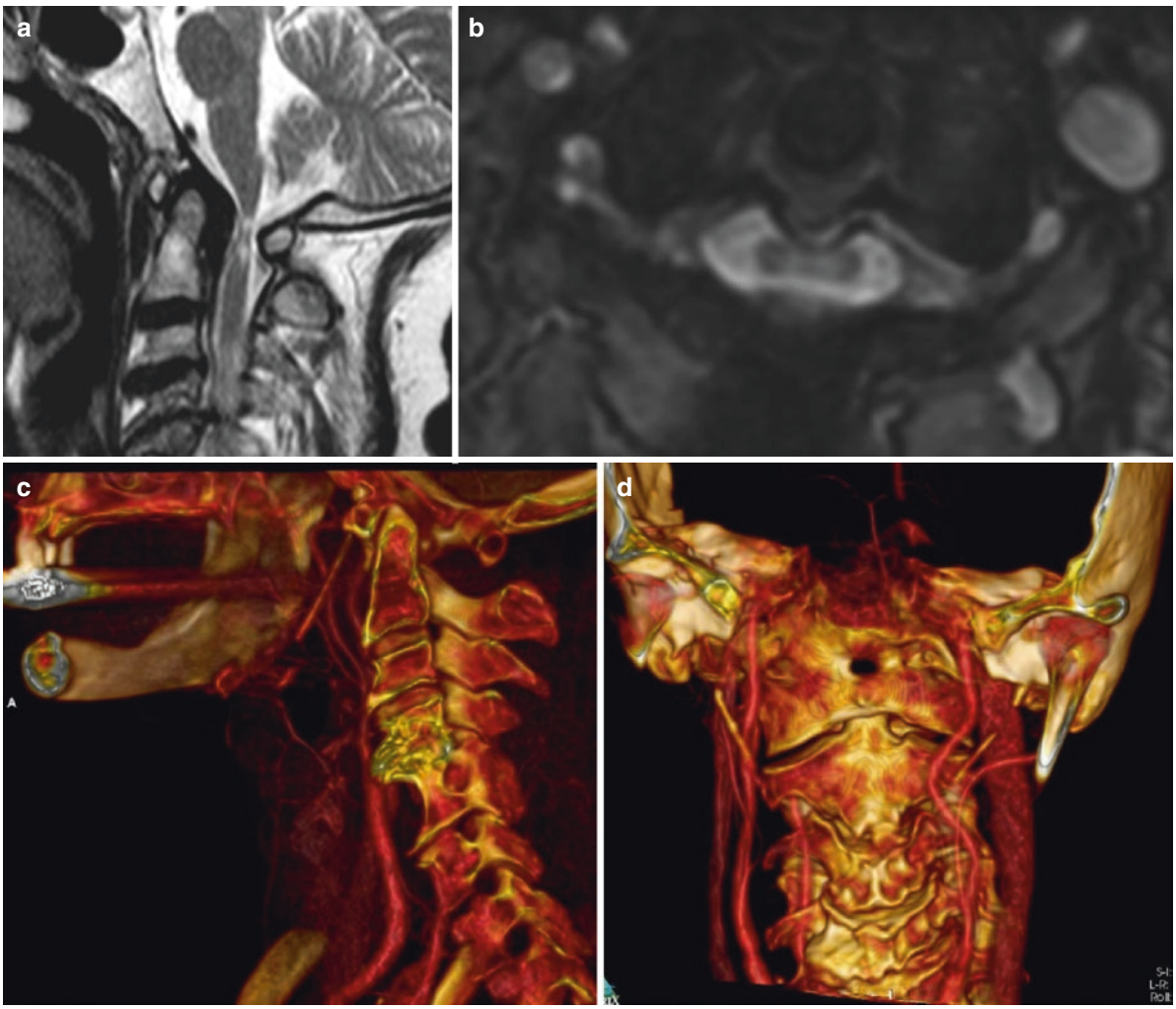

Fig. 1 Preoperative neuroimaging studies. T2-weighted (**a**) sagittal and (**b**) axial magnetic resonance imaging (MRI) of the craniovertebral junction, showing bulbomedullary compression by an extradural mass lesion of the odontoid process (rheumatoid pannus). (**c**, **d**) Three-dimensional reconstruction of computed tomography angiography (CTA) of the same patient

Feasibility of the Endoscopic Endonasal Odontoidectomy

The goal of the surgical operation is to completely remove the odontoid process of C2 and achieve sufficient decompression of the ventral brainstem and CVJ. In the debate between microsurgery and the endoscopic technique, it has been remarked that in the endonasal approach, there can be difficulty in reaching the lower portion of the craniocervical junction and, namely, the base of the dens. To understanding this aspect, numerous studies have been performed on cadavers and on radiological images, with the purpose of delineating the limits and then the indications for the endoscopic approach to odontoid process pathology. Leading authors have widely reported the feasibility of the endoscopic endonasal approach (EEA) to the CVJ [3, 6].

In cases of a low junction, located far below the level of the hard palate, it could be quite difficult, if not impossible, to reach the anterior arch of C1 and the base of the odontoid process. Such cases can represent an indication for the transoral approach. On the other hand, in cases of a higher junction, the dens is more easily reachable and removable by the nasal route.

To preoperatively assess the feasibility of odontoidectomy via an endoscopic endonasal route, on a midline sagittal computed tomography (CT) slice with a bone window it is possible to draw four lines representing possible paths from the piriform aperture of the nasal bones, which target the odontoid process and allow assessment of the inferior limit for surgical exposure. Predicting the inferior limit of the CVJ is crucial to choose the appropriate approach in an area that is considered a transitional area between the endonasal and transoral routes.

Nasopalatine Line

One of the criticisms of the EEA to the upper cervical spine is the limited exposure inferiorly. Endonasal dissection of the upper cervical spine is limited superiorly by the nasal bones and the soft tissues of the nose, and inferiorly by the hard palate and soft palate [34, 35]. The line created by connecting the most inferior point of the nasal bone to the posterior edge of the hard palate in the midsagittal plane is defined as the nasopalatine line (NPL) and is considered a limitation of caudal dissection with straight endoscopic instruments. The angle created by this line and the plane of the hard palate—the nasopalatine angle (NPA)—provides the window of exposure to the skull base and upper cervical spine. The mean nasopalatine angle is 27.1 ± 0.7°. The mean point of intersection between the NPL and the vertebral column is reported to be 8.9 ± 1.8 mm above the base of the C2 vertebral body. The NPL is considered by several authors to be a controversial predictor of the maximal extent of inferior dissection in endoscopic endonasal resection of the odontoid process [34], considering that the inferior limit predicted by the NPL was found to have a mean value of 12.7 mm, below the real inferior extent of surgical dissection. Various pathological factors (basilar invagination) and physiological factors (head positioning) affect the point of intersection of the NPL with the cervical spine. To improve caudal exposure, the use of angled instruments or drills may be of value. Additionally, retraction of the soft palate and drilling of the posterior edge of the hard palate may improve the exposure but may increase the risks of palatal dehiscence and velopharyngeal insufficiency.

Nasoaxial Line

The nasoaxial line (NAxL) is defined as a line constructed on the midsagittal plane using a starting point that corresponds to the midpoint of the distance from the rhinion to the anterior nasal spine of the maxillary bone and a second point at the tip of the posterior nasal spine of the palatine bone. It extends posteriorly and inferiorly to the cervical spine. To predict (more accurately than using NPL) the lower limit of the EEA to reach the CVJ through the correspondence between CT measurements and the real surgical limit, a cadaver study was performed to evaluate the predictive value of the NAxL. The findings supported the close correspondence between the NAxL, drawn on preoperative CT images, and the anatomical surgical extent [36].

Hard Palate Line

The hard palate line (HPL) is defined as a line that passes through the anterior and posterior edges of the hard palate (the anterior nasal spine of the maxillary bone and the posterior nasal spine of the palatine bone, respectively) and intersects with the craniovertebral junction posteriorly. This line represents the long axis of the hard palate [37]. It is considered a reliable marker of the inferior extension of the CVJ especially in congenital abnormalities, such as platybasia with associated basilar invagination, where the tip of the odontoid process is often above the plane of the hard palate [38].

Rhinopalatine Line

The rhinopalatine line is defined as a line constructed on the midsagittal plane, using a starting point that corresponds to the two-thirds point of the distance from the rhinion to the anterior nasal spine of the maxillary bone and a second point at the posterior nasal spine of the palatine bone. The line is extended posteriorly and inferiorly, ending at the cervical spine. There have been great efforts made by different groups to study the inferior limit of the EEA. De Almeida et al. [34] described the nasopalatine (NPL) as a good and accurate predictor of the inferior limit of the EEA, but in their study, the NPL always gave a result below the inferior extent of surgical dissection with a mean value of 12.7 mm. Consequently, the nasoaxial line was reported to predict, more accurately and reliably, the inferior caudal exposure with the EEA. Similarly,

it was found that the NAxL also overpredicted the lower limits of the approach [37]. The rhinopalatine line (RPL) seemed to be the most accurate predictor in several studies.

This predictor accounts also for patient anatomical variability, such as the presence of nasal and palatal osseous and soft structures, together with the hard palate's direction and length, which represent the most significant factors that limit the inferior extension of the EEA. The RPL cannot be used to predict the lateral limits of the EEA in CVJ surgery.

Operative Technique

Depending on individual patients' pathologies, we perform endoscopic endonasal odontoidectomy followed by posterior decompression and fusion in a single-stage surgery.

To accurately choose the correct approach, we consider, on a sagittal CT scan, the relationship between the nasopalatine and rhinopalatine lines and the upper cervical spine.

We routinely use a neuronavigation system (StealthStation S7®; Medtronic, Minneapolis, MN, USA) based on contrast-enhanced magnetic resonance imaging (MRI) with angiographic time-of-flight (TOF) sequences merged with a 1-mm layer CT scan of the brain and cervical spine in a single volume. Generally, we use the optical tracking of the StealthStation S7® merged with the angiographic TOF sequences to provide feasible preoperative images regarding the relationship between the CVJ bony and vascular structures such as the vertebral and carotid arteries. Somatosensory evoked potential neuromonitoring is used routinely.

Patient Positioning and Preparation

Following general anaesthesia and orotracheal intubation, the patient is placed in a supine position with the trunk elevated by about 20°. The head is slightly turned to the right (maximum 10°), not flexed, and fixed in a radiolucent Mayfield-Kees three-pin headclamp. The head is kept parallel to the floor and maintained without flexion or extension during the posterior fusion, when the patient is turned from the supine position to the prone position. In all cases we use the O-arm® system (Medtronic) in the phase of posterior fusion. On this, the optical reference of the neuronavigator is mounted, in case the optical system is used. The magnetic reference is positioned on the patient's head, in case the electromagnetic system is used. We administer antibiotic prophylaxis with cefazolin 2 g 1 h before the procedure.

Nasal Phase

The nose is prepped with cottonoids soaked with diluted iodopovidone 5% solution inside the two nostrils. A 0° angled lens and an 18 cm endoscope associated with a high-definition (HD) camera (Karl Storz, Tuttlingen, Germany) is introduced into the right nostril. Identification of the usual anatomical nasal landmarks is performed (the inferior turbinate laterally and the nasal septum medially). As a standard endoscopic endonasal procedure, above the inferior turbinate, the middle turbinate is identified and luxated laterally, with cottonoids soaked with diluted adrenaline (epinephrine) placed between the middle turbinate and the nasal septum to prevent bleeding of the nasal mucosa. The same manoeuvres are carried out in the left nostril. The endoscope advances parallel to the floor of the nasal cavity until the choana is reached. With the aid of the neuronavigation system, the anatomical landmarks are verified. The mucosa over the posterior and inferior aspect of the nasal septum is cauterized with monopolar coagulation or, better still, with bipolar forceps. We do not routinely perform removal of the anterior wall of the sphenoid sinus since a transsphenoidal corridor is rarely needed unless higher exposure is required in cases where the tip of the dens goes quite high or where more space is required for the surgical manoeuvres because of the patient's individual anatomy. Afterward, an inferior septectomy is performed with sufficient removal of the vomer bone and extending inferiorly down to the hard palate. The most superior limit reached is the clivus–nasal septum junction. At this stage a few important anatomical landmarks should be identified, which guide the surgeon to stay oriented: (1) the clivus–septum junction superiorly; (2) the Eustachian tubes laterally; and (3) the nasal floor/soft palate inferiorly as marked by the hard and soft palate. The neuronavigation will confirm the position of such surgical landmarks and give the correct direction for the subsequent surgical steps.

Nasopharynx Phase

The key points of the nasal phase allow the widest exposure of the rhinopharynx and avoidance of any conflict between instruments during the next surgical steps. The nasopharynx mucosa is incised on the midline (Fig. 2a), and the muscles are dissected bilaterally to expose the anterior arch of C1 (Fig. 2b). Several authors have described a reverse U-shaped nasopharyngeal flap prepared with monopolar electrocautery, elevated and reflected caudally to the level of the soft palate to improve the surgical field. The craniocaudal extension of the flap involves the inferior third of the clivus superiorly and the C2 vertebral body inferiorly; the lateral margins of the operative exposure includes the lateral masses of the C1 vertebra. The U-shaped nasopharynx flap extends the surgical corridor laterally, but on the other hand it increases the risk of injury to the parapharyngeal carotids, which are located laterally to the superior pharyngeal constrictor muscle. We prefer doing a straight midline opening of the nasopharynx because it guarantees sufficient exposure and a lower risk of vascular damage. Then, we proceed with skeletonizing of the anterior arch of C1 and of the odontoid process in a subperiosteal fashion.

C1 Anterior Arch Preservation in Selected Cases

Recently, several authors have reported their experience in the matter of endoscopic endonasal odontoidectomy, focusing on the preservation of the C1 anterior arch during the craniovertebral junction phase, avoiding posterior fixation [32, 39]. Particularly in cases of rheumatoid arthritis or other inflammatory diseases, the anterior arch of the atlas is preserved by drilling the odontoid base, weakening its apex and leading to pulling downward of the dens in the working area. Subsequent removal of the axis with other remaining compressive inflammatory lesions is performed using a combination of a high-speed drill, ultrasonic bone curette and standard Kerrison rongeurs [32, 39]. According to such authors, working above and below the C1 anterior arch and its preservation not only represent an element of stability but also provide an important opportunity for reconstruction and to reinforce the closure. Additionally, the same groups, in cases of inveterate Anderson–D'Alonzo type II fractures or the combination of an odontoid fracture with a fracture of the anterior arch of C1, have proposed a technique of anterior fixation and anterior C1 arch reconstruction [40].

Craniovertebral Junction Phase and Closure

In our technique, the anterior arch of the atlas is exposed and removed using the high-speed drill with diamond burrs and Kerrison rongeurs. Posteriorly, the odontoid process of C2 is exposed (Fig. 2c), separated from the alar and apical ligaments, dissected from the transverse ligament, thinned using

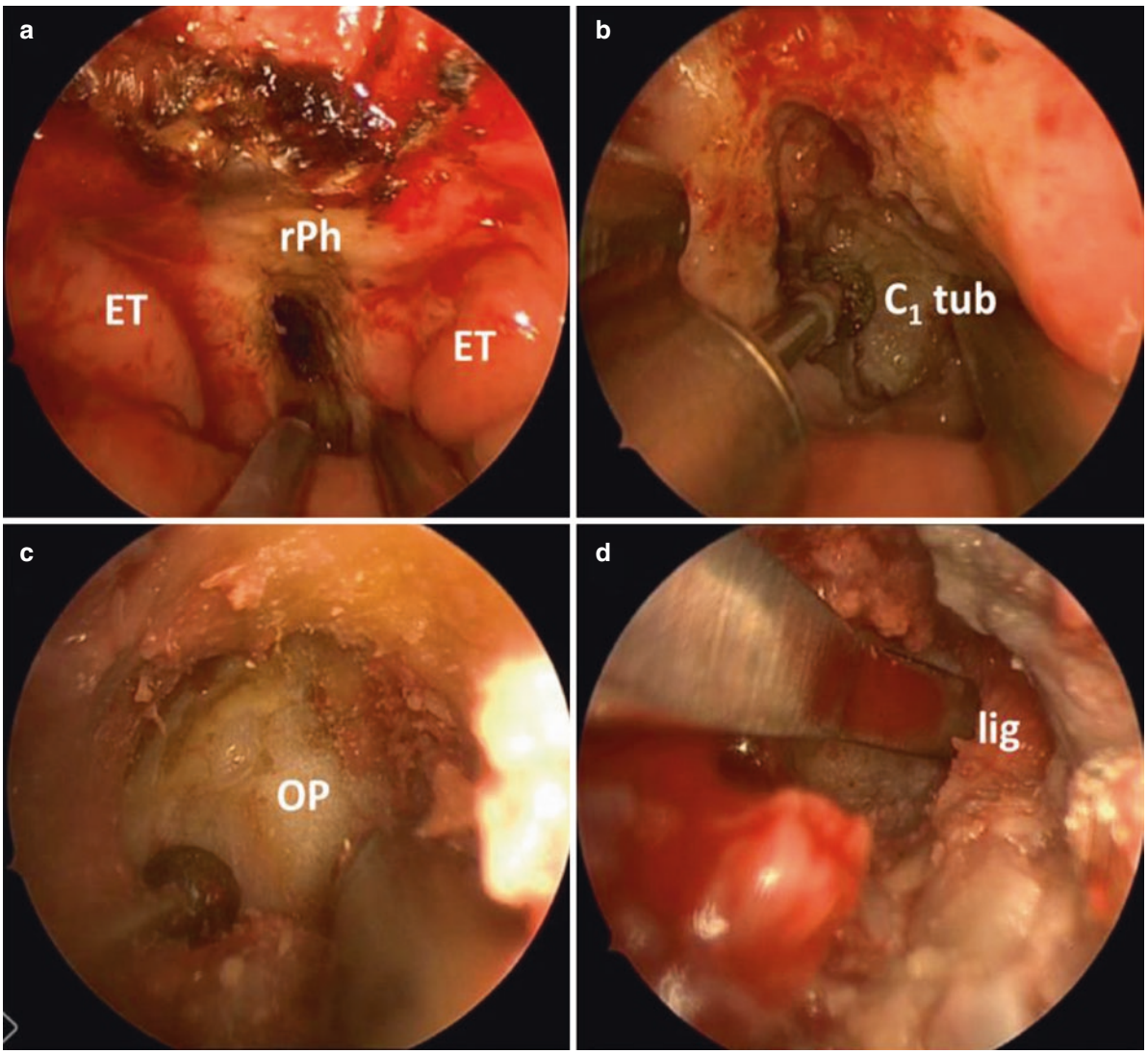

Fig. 2 Intraoperative pictures of the endoscopic endonasal approach. (**a**) Incision into the rhinopharynx (rPh). (**b**) Drilling of the anterior arch of C_1. (**c**) Drilling of the odontoid process (OP) of C_2. (**d**) Freeing of the remaining part of the dens from the ligaments. *C1 tub* anterior tubercle of C1, *ET* Eustachian tubes, *lig* ligaments

the microdrill and finally removed (Fig. 2d). At this point, a wide surgical corridor is created. The odontoidectomy is performed carefully by using the high-speed drill, Kerrison rongeurs and, in cases of lesions with a soft consistency, curettes and pouches or ultrasound aspiration. When the removal is complete, the dural plane appears to pulsate and indicates optimal decompression of the brainstem (Fig. 3a, b).

After satisfactory haemostasis is achieved, the closure is guaranteed with a layer of fibrin glue only in the absence of possible dural tearing (Fig. 3c). In the case of CSF leakage, packing with Gelfoam/Surgicel and fibrin glue is used to reinforce the closure. In these cases we consider the possibility of positioning an extended lumbar drain (ELD) at the end of the operation. We close the nasopharynx mucosa with a single stich because the median opening allows faster closure of the muscles at the end of the endoscopy time. Generally, we position a nasogastric tube under endoscopic control (Fig. 3d).

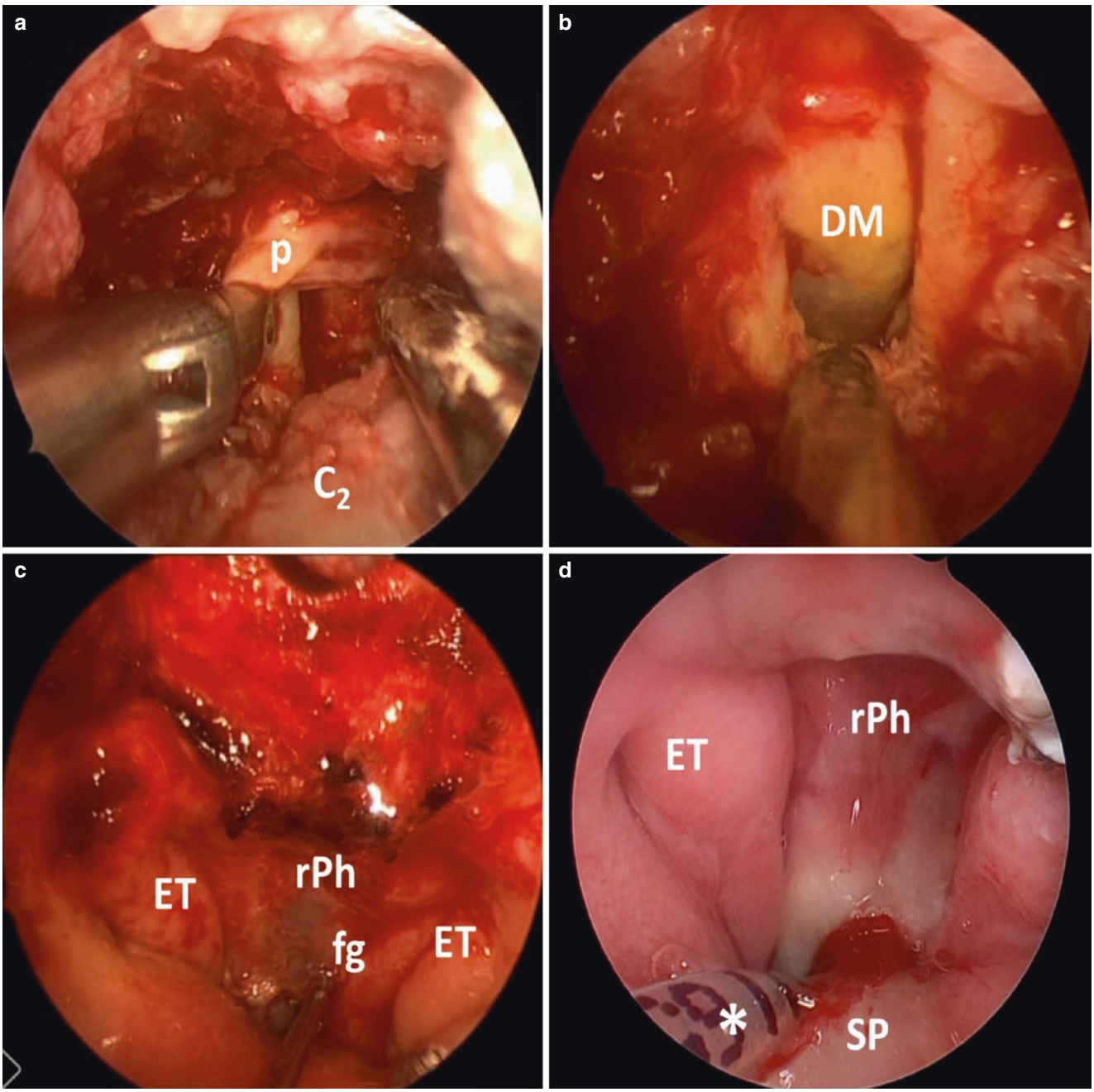

Fig. 3 Intraoperative pictures of the endoscopic endonasal approach. (**a**) Removal of pannus (p) causing compression. (**b**) Dura mater (DM) of the craniovertebral junction. (**c**) Closure of the muscle and mucosal incision with the aid of fibrin glue (fg). (**d**) Endoscopic control of the surgical field 3 days later, showing optimal closure of the incision. The *asterisk* denotes the nasogastric tube. *C2* base of the dens (body of C_2), *ET* Eustachian tube, *rPh* rhinopharynx, *SP* soft palate

Posterior Fusion

The second step of the operation is characterized by the posterior occipitocervical fusion. The patient, already fixed to the Mayfield-Kees three-pin carbon fibre radiolucent headholder, is turned from the supine position to the prone position with the head parallel to the floor and with a slight degree of extension. This position considers the C0–C2 angle, which is formed by the posterior extension of the hard palate and the vertical line passing through the dens, and avoids breathing impairment related to the flexion. A midline incision is performed, starting from the inion, to the spinous process of C6. The fascia is exposed and incised on the midline with monopolar cautery. The muscle dissection is performed along the raphe in a subperiosteal fashion from the basiocciput to the posterior complex of C5. The bone landmarks are clearly visible: (1) the occipital bone; (2) the posterior arch and lateral masses of C1; and (3) the posterior complex from C1 to C5.

Generally, we remove the posterior arch of C1 because in most of our cases it is contributing to the bulbopontine compression. The lateral masses of C3 and C4 are identified and verified through the O-arm® system. The fixation system we use in all cases is the Vertex titanium system (Medtronic). The high-speed drill is used to prepare the position of the screws within the lateral masses of C3 and C4. The polyaxial screws are inserted according to the Magerl technique [41] to avoid vascular injuries. In the basiocciput the monoaxial screws are positioned 2 cm from the inion on both sides and 1 cm above the sinuses. The length of the screws we use is 8 mm. After the screws are positioned, the two rods are pulled to obtain the correct alignment of the cervical spine and finally fixed through the wrench of the wing nuts. The bone fusion is improved with the addition of bone substitutes. The last verification with the O-arm® system is done at the end of the procedure. At discharge we recommend use of a cervical collar for 2 months.

Series Presentation

In the Neurosurgical Clinic at the University of Messina, a series of four endonasal endoscopic odontoidectomies were performed. Demographic, clinical and management details are summarized in Tables 1 and 2.

All four patients were female, ranging between 62 and 82 years of age (mean age 67.75 years). Three patients were admitted with a neurological onset characterized by tetraparesis; in one patient, motor deficits were prevalent in the right arm. Urinary incontinence was present in two patients. One patient presented with severe dysphagia with both solids

Table 2 Details of patient management in our series

Patient no.	Procedures	Operating room set-up	Duration of postoperative hospital stay (days)
1	Endoscopic endonasal odontoidectomy and occipitocervical stabilization at the same stage	StealthStation S7® with optical tracking + O-arm®	17
2	Endoscopic endonasal odontoidectomy and occipitocervical stabilization at the same stage	StealthStation S7® with optical tracking + O-arm®	13
3	Endoscopic endonasal odontoidectomy and occipitocervical stabilization at the same stage	StealthStation S7® with optical tracking + O-arm®	19
4	Endoscopic endonasal odontoidectomy	StealthStation S7® with optical tracking	9

Table 1 Demographic, aetiological and clinical data on the patients in our series

Patient no.	Age (years)	Sex	Aetiology	Symptoms	Postoperative outcome
1	62	Female	Rheumatoid pannus	Right arm weakness Tetrahyperreflexia Urinary incontinence	Improved Oral feeding
2	64	Female	Odontoid process misalignment in patient with previous Anderson–D'Alonzo type II fracture (not stabilized)	Tetraparesis Tetrahyperreflexia Urinary retention	Improved Oral feeding
3	82	Female	Rheumatoid pannus	Tetraparesis	Improved Oral feeding
4	63	Female	Craniocervical junction malformation	Tetraparesis Severe dysphagia Dysphonia	Improved Dysphagia not completely resolved

and liquids. In two patients, the symptoms were related to the presence of a rheumatoid synovial pannus, while the other two cases showed signs and symptoms due to a complex malformation of the craniocervical junction and to misalignment of the odontoid process following a previous non-fused Anderson–D'Alonzo type II fracture, respectively. Interestingly, the patient affected by the complex CCJ malformation had previously undergone occipitocervical stabilization at another institution. She then underwent an attempted transoral odontoidectomy, which failed because of the higher position of the dens. She was subsequently referred to our clinic for anterior decompression performed through an endoscopic endonasal odontoidectomy. In the remaining three patients, anterior decompression and posterior stabilization were performed during the same operation.

The mean length of stay was 14.5 days (range 9–19 days). In all patients there was improvement of the neurological conditions in comparison with the preoperative status. In one patient the swallowing dysfunction resolved, allowing early oral feeding. In two cases, implementation of parenteral nutrition was necessary for a few days. In one case the nasogastric tube was left in place to facilitate enteral feeding.

Postoperative Management

In our practice, according to the general clinical condition of the patient and the length of sedation, we prefer to keep the patient in our intensive care unit for 24 h. This occurred in two of the four cases we treated. In our department, the primary aim is early mobilization of the patient to lower the risks associated with extended bed rest. In addition, use of a nasogastric tube guarantees sufficient caloric intake, with the addition of parenteral nutrition, when required. We perform at least two endoscopic postoperative controls: one in the first 24 h and one before discharge. During such checks we verify the closure of the surgical wound and the possible presence of CSF leakage, and then we remove the nasogastric tube under endoscopic control. This manoeuvre can be performed only after testing of the function of the lower cranial nerves by an otolaryngologist. In our series, removal of the nasogastric tube occurred in three patients: on the eighth postoperative day in two patients, and on the seventh postoperative day in the other one. In our series, before discharge, a CT scan of the head and cervical spine was performed to assess the degree of the odontoidectomy and the correct positioning of the screws and rods of the posterior fusion, and MRI was performed to evaluate the decompression of the neurovascular structures. A further control was performed after 3 months. All patients started a physical rehabilitation programme, which was also continued after discharge (Fig. 4).

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

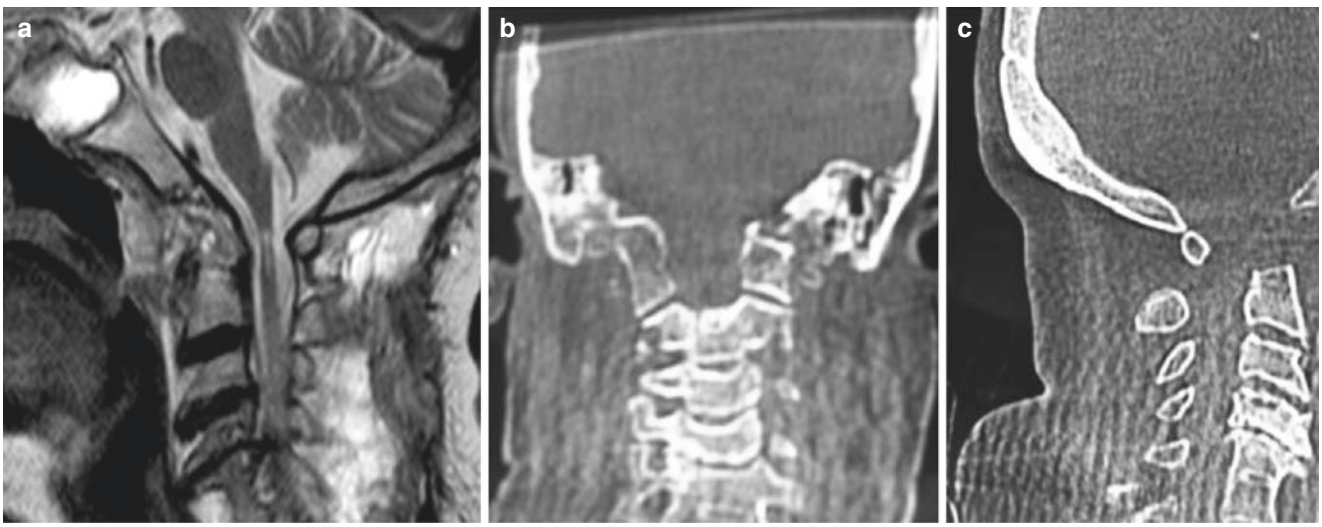

Fig. 4 Postoperative neuroimaging studies of the same patient shown in Fig. 1. (**a**) T2-weighted sagittal magnetic resonance imaging (MRI) of the craniovertebral junction (CVJ) showing optimal decompression of the bulbomedullary junction. (**b**, **c**) Intraoperative O-arm® images showing removal of the odontoid process. (**d**–**f**) Three-dimensional reconstruction of the postoperative computed tomography (CT) scan of the CVJ

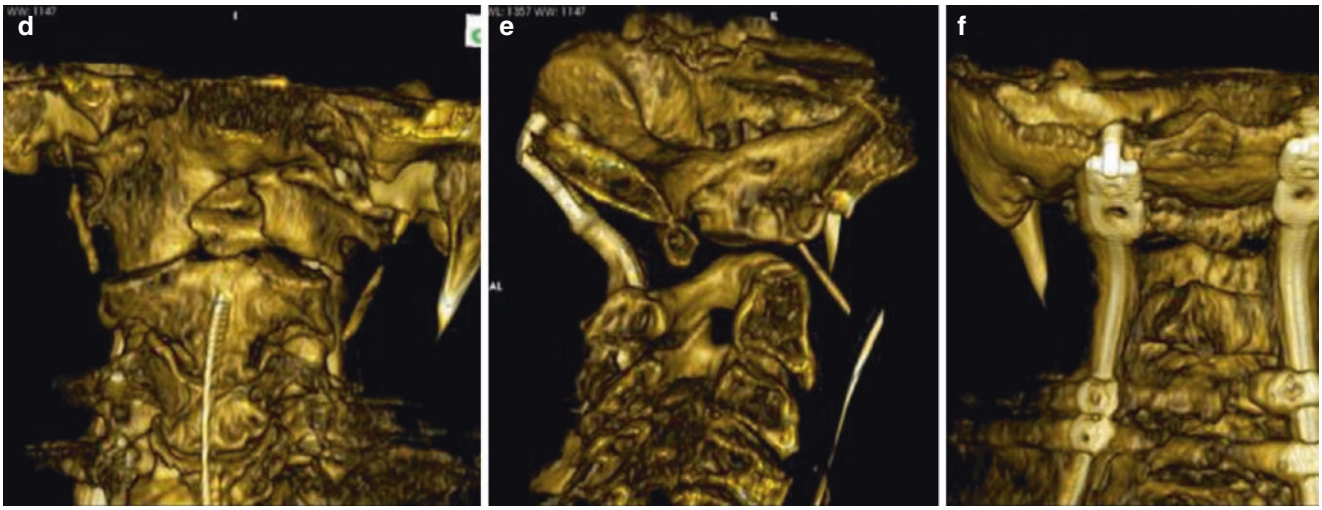

Fig. 4 (continued)

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Endoscopic Endonasal Approaches for Treatment of Craniovertebral Junction Tumours

Davide Locatelli, Apostolos Karligkiotis, Mario Turri-Zanoni, Frank Rikki Canevari, Fabio Pozzi, and Paolo Castelnuovo

Abstract Tumours involving the craniovertebral junction (CVJ) are challenging because of their local invasiveness and high recurrence rates, as well as their proximity to critical neurovascular structures and the difficulty of reconstructing the resulting skull base defect at this site. Several surgical techniques are currently available to access these lesions, including the far lateral, extreme lateral, direct lateral, transcervical, transoral and transnasal approaches. In this paper, application of the endoscopic endonasal approach (EEA) in the treatment of CVJ tumours is analysed. The indications, contraindications, preoperative workup, step-by-step surgical technique, skull base reconstruction options and postoperative management are described. The advantages and limitations of the EEA are also discussed. Finally, a systematic review of the literature is provided to elucidate the levels of evidence supporting the use of the EEA in this field. Employment of this approach to the CVJ has contributed to high success rates in achieving gross total resection of tumours and improvement in neurological symptoms. Intraoperative and postoperative complication rates are acceptable, with cerebrospinal fluid leakage being the major concern (with a 17–25% incidence). Moreover, in comparison with traditional approaches to the CVJ, the EEA provides lower rates of postoperative dysphagia and respiratory complications. Use of the EEA for treatment of CVJ tumours appears to be a rational alternative to the conventional transoral, transcranial and transcervical approaches in selected cases. Multidisciplinary teamwork including different specialists—such as medical and radiation oncologists, radiologists, otorhinolaryngologists and neurosurgeons—is strongly recommended for the purpose of offering the best treatment strategy for the patient.

Keywords Chordomas · Meningiomas · Craniovertebral junction · Clivus · Posterior cranial fossa · Endoscopic transnasal approaches · Multiportal endoscopic approaches · Radiotherapy

Introduction

General Aspects

Different tumours can involve the anterior and anterolateral craniovertebral junction (CVJ), although the more frequently occurring ones are chordomas and meningiomas. The surgical treatment of these tumours is challenging because of their local invasiveness and high recurrence rates, as well as the proximity of the clivus and CVJ to critical neurovascular structures and the difficulty of reconstructing the resulting skull base defect at this site. Thus, the benefits of total removal must always be balanced against the risks related to an extensive and potentially invasive surgical dissection.

The leading objectives of the surgical treatment of CVJ tumours are to achieve gross total resection, decompress the neural structures, and avoid surgical morbidity and mortality [1, 2]. Several surgical approaches can be used to access CVJ

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lesions, including the far lateral, extreme lateral, direct lateral, transcervical, transoral and transnasal approaches, which have been developed over the years to overcome the significant morbidity and mortality rates of the historical midline suboccipital approaches [1].

Traditionally, the transoral approach is the most frequently used corridor for accessing the lower clivus and the anterior CVJ, without the need to mobilize or retract the cranial nerves, the lower brainstem or the upper cervical spinal cord. However, it is associated with a high rate of complications such as velopharyngeal incompetence, hypernasal speech and nasal reflux, dental injury, oedema or tongue necrosis, upper airway obstruction from retropharyngeal oedema, posterior pharyngeal wound dehiscence, dysphagia, odynophagia, pharyngeal cellulitis, meningitis (from cerebrospinal fluid [CSF] leakage) and temporomandibular joint syndrome [3, 4]. Otherwise, the endoscopic endonasal approach (EEA) provides direct and full access to the anterior CVJ, as well as to the lower, middle and superior clivus, offering improved lateral visualization, decreased airway and swallowing morbidity, preservation of palatal function, decreased postoperative pain, and a reduced incidence of tracheostomy [3, 4].

One of the major limitations of the EEA, preventing radical resection of clival–CVJ tumours, is lateral extension of the tumour with adhesions to important neurovascular structures [5]. In these cases, different transcranial and transcervical approaches may be combined with the endoscopic transnasal one to optimize the degree of resection and reduce morbidity [3, 6]. Adjuvant radiation therapy is usually indicated for local tumour control and to prolong progression-free survival, as well as in the case of extensive involvement of neurovascular structures [7–9].

Treatment Planning

Appropriate treatment of CVJ tumours strongly calls for multidisciplinary team management, including neurosurgeons, otorhinolaryngologists, radiologists, medical oncologists and radiation oncologists. The goals of surgical treatment may include (1) obtaining a diagnosis when preoperative imaging is not able to establish it; (2) decompressing neural structures; and (3) maximizing survival outcomes and patient quality of life while minimizing collateral damage to nasal or intracranial structures, as well as complication rates. In this regard, a multidisciplinary evaluation is devoted to defining the best treatment strategy for a given patient, taking into account the tumour biology and extension, as well as the patient's comorbidity and expectations.

When surgery is indicated as the first treatment option, a tumour board discussion is paramount in defining the appropriate surgical approach to achieve gross total resection while minimizing postoperative sequelae. The advantages and limitations of the EEA should be compared with those offered by traditional techniques such as the transoral, transcervical and far lateral approaches to identify the best surgical corridor for the lesion to be treated. In selected cases of multicompartmental lesions, a combination of different surgical approaches can be used to achieve disassembly of the different tumour components in a precise and anatomically oriented fashion.

In the presence of encasement of a major vessel (e.g. the internal carotid, basilar or vertebral arteries) and lower cranial nerve involvement, radical resection of the tumour is precluded and additional treatment strategies should be considered after surgery. In such cases, conventional radiotherapy, stereotactic radiosurgery and the recently introduced hadron therapy may be used. Particle radiation therapy using proton and heavier ion beams provide greater biological effectiveness and improve dose distributions in comparison with photon beams, and thus enable dose escalation within the tumour while sparing normal tissues [10]. When partial or subtotal resection of the tumour is planned, a preoperative multidisciplinary consultation with radiologists and radiotherapists is mandatory to delineate the tumour mass suitable for surgical removal and the tumour volumes suitable for irradiation (Fig. 1). Especially in cases where postoperative radiotherapy or heavy ion therapy is planned, the potential risk of atlantooccipital (AO) instability with the need for CVJ fixation has to be preoperatively anticipated. In selective cases, resection of CVJ tumours requires a lateral extension that can affect AO stability. In particular, it is extremely important to evaluate the extent of condyle resection to determine the immediate and long-term postoperative effects on AO stability [11]. Notably, if the extent of medial condyle resection is less than 50%, patients have a low risk of instability and, in general, only clinical observation is required, without the need for surgical fixation. On the other hand, a high risk of AO instability is observed when medial resection involves 75% of the condyle; in these instances, surgical fixation should be performed. In cases where the extent of condyle resection is between 50% and 75%, the patients are asymptomatic and no AO joint resection is performed, close monitoring should be scheduled [12].

Given these data, the goal of surgery is to maximize tumour resection while minimizing postoperative AO instability. Whereas far lateral and transcervical approaches predominantly dominate the posterolateral segment of the CVJ and often demand drilling out of the C1 lateral mass and condyle to reach the medial portion of the lesions, the EEA permits direct access to the anteromedial part of the CVJ and makes enlargement from the anteromedial to posterolateral sections feasible with removal of the condyle in a progressive fashion, in relation to the specific location of the lesion, thus minimizing the risk of AO instability. However, in some cases the tumour extension to the condyle and to the AO joint requires extensive resection with a subsequently increased risk of AO instability, no matter which surgical approach is used.

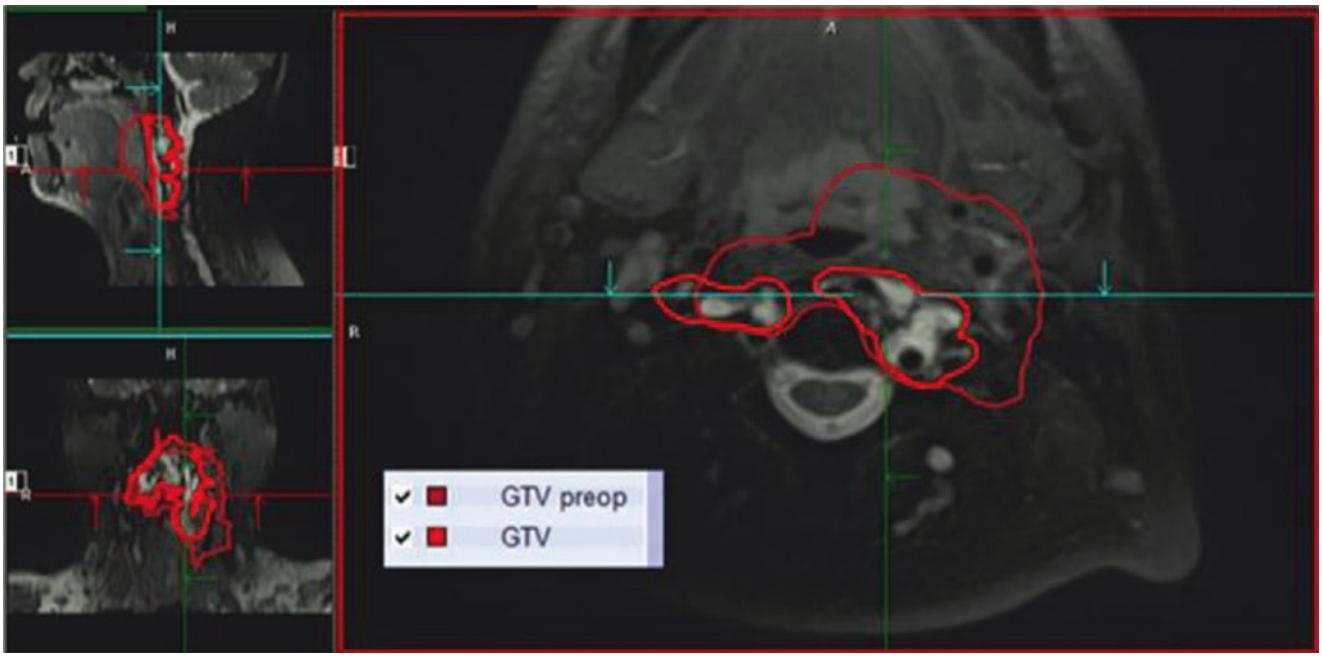

Fig. 1 Preoperative radiation planning to delineate the tumour mass suitable for surgical removal and tumour volumes suitable for adjuvant irradiation

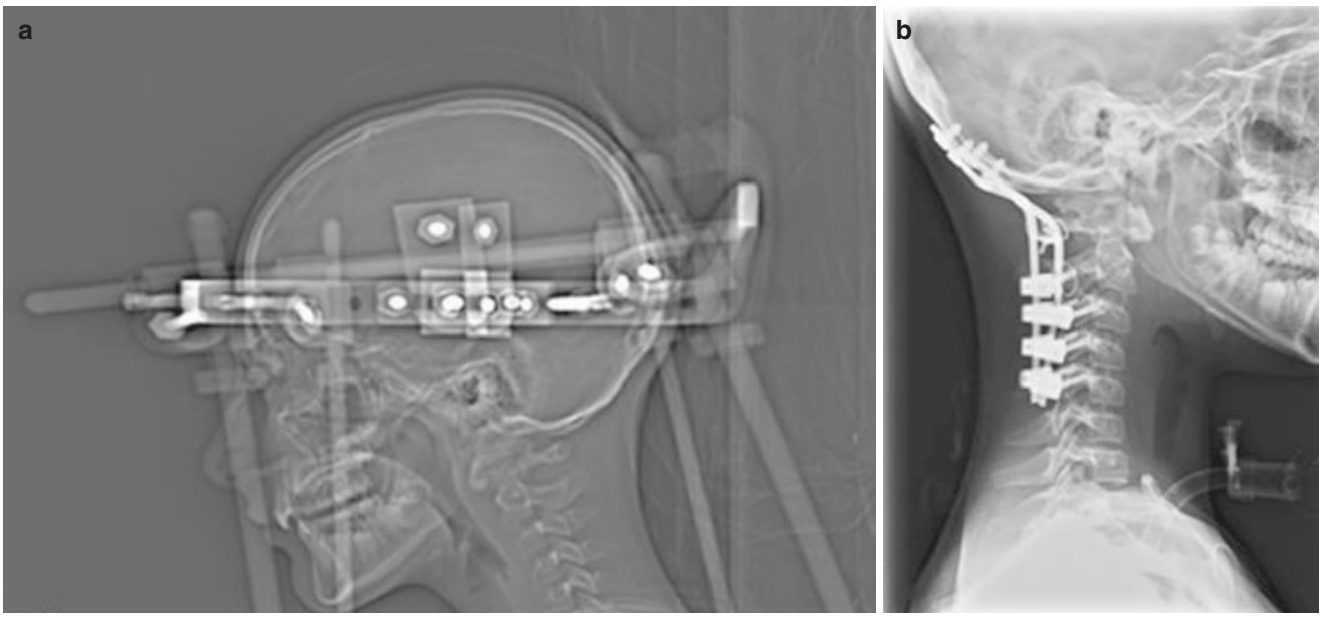

Fig. 2 (**a**) Sagittal computed tomography (CT) scan of a halo vest orthosis used to ensure temporary cervical immobilization. (**b**) Lateral cervical x-ray showing C0–C5 fixation performed before excision of a clival chordoma involving the occipitoaxial junction

For all of these reasons it is mandatory to preoperatively plan the degree of condyle and AO joint resection in order to anticipate the need for CVJ fixation, as well as to plan appropriate cervical stabilization. Posterior arthrodesis with titanium fixation rods and plates is a safe, permanent and effective method to achieve AO stability but may preclude postoperative irradiation by interfering with the gross tumour volume (GTV) and the clinical target volume (CTV) delineation. A rigid cervical collar and a halo vest orthosis can be used to ensure cervical immobilization temporarily, representing a valid option for patients with mild AO instability and for patients in whom postoperative irradiation is planned (Fig. 2).

Preoperative Assessment

The preoperative assessment includes nasal endoscopy, imaging and biopsy. Endoscopic examination is paramount to evaluate the sinonasal anatomy and identify any anatomical variations (septal deviation, spurs or perforation) that may influence the reconstruction. Moreover, any extracranial, nasopharyngeal or intranasal extension of the tumour can be seen. The essential preoperative imaging modalities are computed tomography (CT) and contrast-enhanced magnetic resonance imaging (MRI) scans, which are complementary: a CT scan provides a perfect analysis of bone structures, and an MRI scan provides information on the real extension of the lesion and distinction of different kinds of tissues (tumour tissue versus brainstem). A CT scan of the paranasal sinuses, CVJ and cervical spine is imperative to show bony erosion at the level of the foramen magnum and AO joint, as well as to evaluate the opportunity for occipitocervical fixation. On the other hand, MRI permits evaluation of the tumour characteristics (vascularization), the eventual vessel encasement and the relationship with the lower cranial nerves.

These radiological investigations are essential guiding instruments during surgery. Nowadays the increasingly widespread application of neuronavigation systems with the possibility to obtain real-time tracking images, three-dimensional reconstructions of the neurovascular anatomy and fusion CT/MRI images has definitely improved the accuracy of surgical procedures, and they should always be used when possible.

Moreover, angiographic studies (CT angiography, magnetic resonance angiography or conventional angiography) are necessary for evaluation of intracranial arterial displacement or encasement by the tumour, as well as for evaluation of the course of the vertebral arteries and, above all, of the parapharyngeal segment of the internal carotid artery (ICA), which, in some cases, can show medial kinking behind the posterior wall of the nasopharynx.

In the case of nasopharyngeal extension of the tumour, once the imaging evaluation has been fully completed, multiple biopsies for histopathological examination should be performed to reach a more precise preoperative diagnosis.

Technical Considerations

Endoscopic Resection

The major advantage of the EEA is the possibility of obtaining a multiangled and magnified perspective, which makes it easier to differentiate normal and diseased mucosa. The entity of dissection can be modulated in relation to the extension of the lesion. The anatomical limits of the EEA in CVJ surgery are represented by the brainstem posteriorly, the parapharyngeal ICAs laterally, and the vertebral arteries and lower cranial nerves superoinferiorly [13]. In this specific anatomical region, lesions such as chordomas commonly have an inferior extension with involvement of C1 and, less frequently, C2 [3]. Thus, most of the time a combination of endoscopic transclival and transodontoid approaches is necessary for either extradural or intradural lesions of the CVJ and the posterior cranial fossa.

Chordomas and chondrosarcomas are examples of lesions that can be purely extradural; however, these lesions often have an intradural component. Meningiomas are lesions that mainly occur intradurally in the posterior fossa. The main advantage of the EEA is that through this corridor, the vital structures are located laterally to the tumour and there is no need for neural tissue retraction or dissection in between cranial nerves. Often the tumour growth pushes away the brainstem artery and cranial nerves, allowing better vision and safer resection of the lesion itself [14].

Removal starts usually with intracapsular debulking, with the purpose being to reduce the tumour volume and make extracapsular dissection easier. The feasibility of intradural resection is strictly in rapport to the presence of a preserved subarachnoid plane; otherwise, adhesion of the tumour to neurovascular structures interferes with safe resection.

To accomplish this, the two-nostrils, four-hands technique is the best way to perform careful extracapsular dissection and removal of the lesion: it allows exposure and dissection of the tumour under direct view and employment of countertraction, avoiding blind manoeuvres (especially pulling with grasping forceps). Furthermore, the endoscopic transclival and transodontoid approaches allow devascularization of durally based lesions, such as clival meningiomas, during the tumour exposure, providing more thorough tumour resection.

In any case, it is essential to always take into account the golden rule of the Pittsburgh group: do not cross nerves. Anytime the surgeon judges that the EEA is not sufficient as a single approach (for different reasons such as extension, infiltration, etc.), lateral approaches and staged interventions should be contemplated [14, 15]

Caudal exposure remains an essential limitation of the EEA in CVJ surgery because of the presence of nasal and palatal bony and soft tissues superiorly and inferiorly, respectively. Different lines have been described as predictors of the inferior limit of the EEA in CVJ surgery. Various studies have asserted that both the nasopalatine line and the nasoaxial line overestimate the inferior limit of surgery and do not provide a reliable prediction of actual surgical results [16]. Thus, a new line—the rhinopalatine line—has been proposed recently, which appears to correspond most precisely to the intraoperative situation. The rhinopalatine

line is identified as a line drawn on the midsagittal plane of a CT or MRI scan, with a starting point that corresponds to the two-thirds point of the distance from the rhinion to the anterior nasal spine of the maxillary bone and a second point at the tip of the posterior nasal spine of the palatine bone. The line is then extended posteriorly and inferiorly to end at the cervical spine [16]. The rhinopalatine line seems to predict the caudal limit of the EEA more precisely than previously described lines, and it is essential in supporting surgeons to choose the best approach in relation to the anatomy of the patient and the location of the tumour.

Given caudal extension of the lesion at the level of the C2–C3 interspace, an endoscopic transoral approach should be combined with an endonasal one (Fig. 3) [17]. In recent years the development of multiportal endoscopic approaches combining the transnasal and transoral corridors to access the superior and inferior limits of CVJ tumours has maximized the degree of tumour resection while avoiding splitting of the soft palate and the subsequent complications it may cause [18, 19].

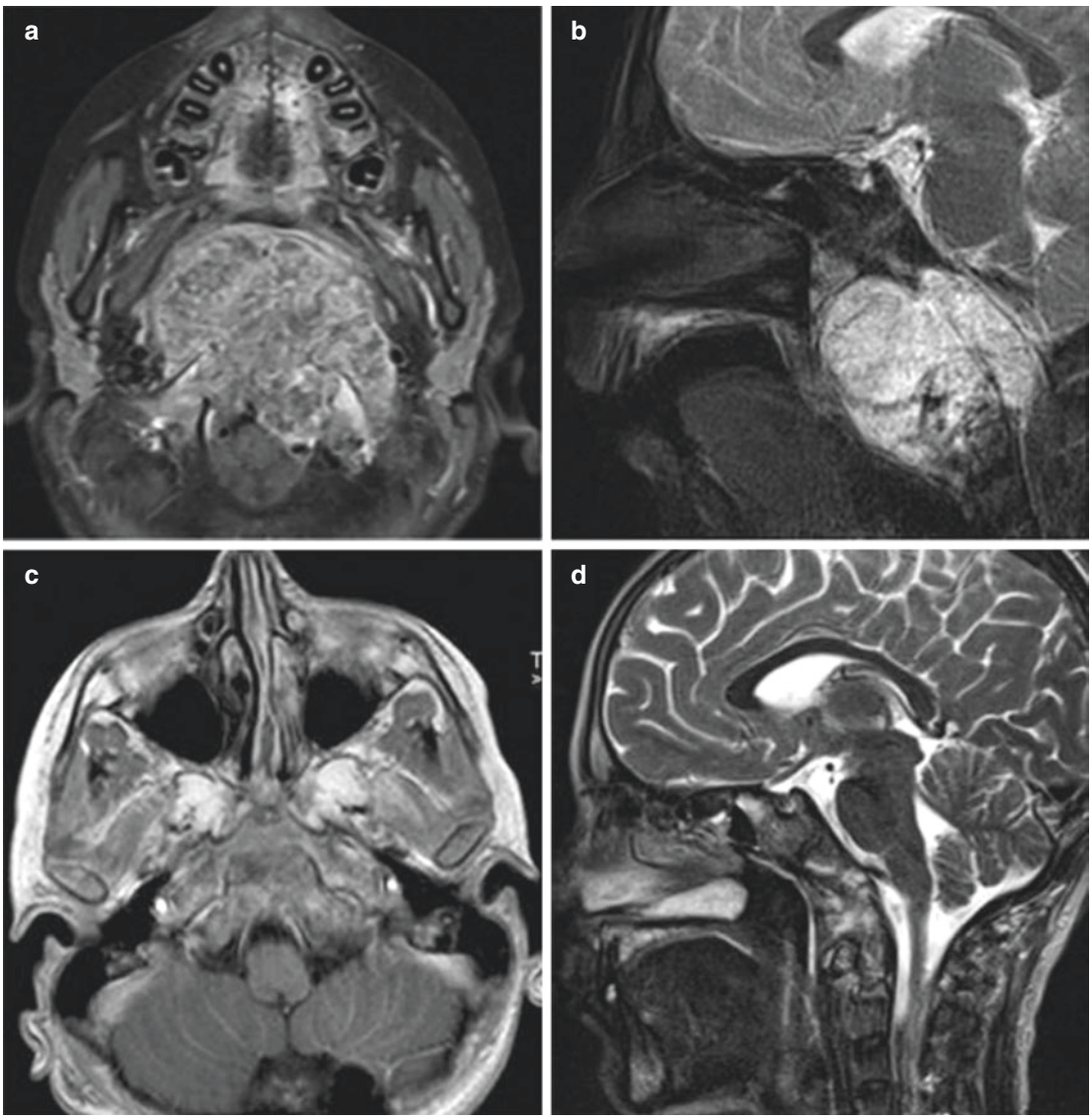

Fig. 3 (**a**) Preoperative axial T1-weighted gadolinium-enhanced and (**b**) preoperative sagittal T2-weighted magnetic resonance images (MRIs) of a posterior cranial fossa chordoma involving the clivus and craniovertebral junction. (**c**) Postoperative axial T1-weighted gadolinium-enhanced and (**d**) postoperative sagittal T2-weighted MRIs after a combined multiportal endoscopic transoral and transnasal approach, showing residual disease involving C1 and C2. (**e**) Postoperative axial T1-weighted gadolinium-enhanced and (**f**) postoperative sagittal T1-weighted MRIs showing extension of tumour resection after second-stage surgery using an exclusive endoscopic transnasal approach

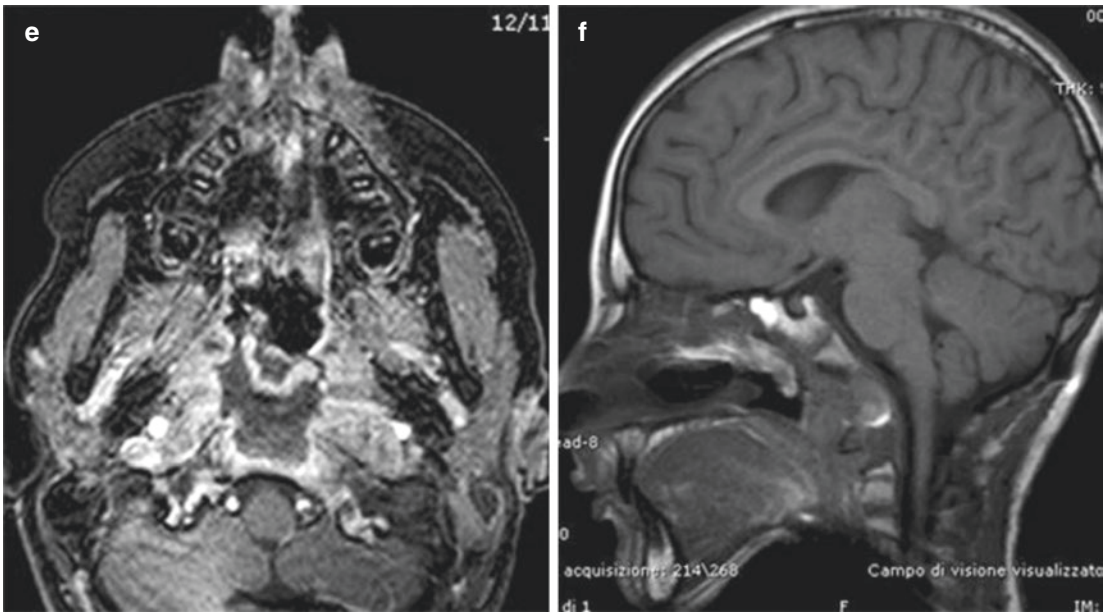

Fig. 3 (continued)

Endoscopic Skull Base Reconstruction

Skull base reconstructions are employed to recreate a separation of the cranial cavity from the sinonasal tract to prevent CSF leakage, pneumocephalus and intracranial infections [20]. A reconstructive technique must be chosen according to anatomomechanical and biological factors: the former are linked to the site and size of the defect, as well as its borders; the latter are linked to the histology of the resected lesion, its diffusion behaviour and the eventual need for adjuvant radiotherapy. It is necessary to emphasize that the size of the defect, as well as the location and its borders, determine the complexity of the skull base reconstruction and can make the endoscopic endonasal procedure either easy, complex or even impossible.

Many different materials have been proposed to aid the closure of dura apertures such as autologous grafts, allogenic transplants and various synthetic biomaterials. However, the ideal material for duraplasty should be (1) autologous, in order to avoid all potential risks of heterogeneous grafts; (2) free from biological hazards to avoid human immunodeficiency virus (HIV) infection, hepatitis and other transmissible diseases; (3) able to facilitate fibroblastic migration and connective tissue deposition; and (4) capable of guaranteeing a good cost-effectiveness ratio [21]. Skull base reconstruction is generally described according to the chosen reconstructive procedure with use of free grafts or vascularized flaps. Nowadays, the autologous tissues most frequently employed with the grafting techniques are fascia lata, fascia temporalis, cartilage, bone and fat. Vascularized flaps are divided into local and regional ones, depending on the area from which they are obtained—whether it is adjacent to or distant from the defect. An ideal flap should be simple to design, be resistant to trauma, have low morbidity, supply an appropriate surface area and have an arc of rotation that allows its transposition without a tendency to return to its original position. The most frequently employed vascularized flaps for posterior skull base and CVJ reconstruction are the nasoseptal flap [also known as the Hadad–Bassagasteguy flap (HBF)] and the temporoparietal fascia flap (TPFF) [22, 23]. Other flaps described in the literature are the posterior pedicle inferior turbinate flap, the Oliver pedicle palatal flap, the salpingopharyngeus myomucosal flap and the microvascular free flap [24–28].

Surgical Techniques

General Principles

Surgery is performed with the patient placed under hypotensive general anaesthesia and placed in a supine anti-Trendelenburg position. The nasal cavities are packed with pledges soaked in 2% oxymetazoline, 1% oxybuprocaine and adrenaline (epinephrine) 1/100,000 solution, for 10 min. Telescopes with 0° and 45° views and 4 mm in diameter are used. To facilitate the surgical procedure, a microdebrider, diode laser, bipolar forceps, and straight, angled and double-ended instruments are recommended. An endonasal pen-

style microdrill equipped with long handles and with diamond or cutting burrs—as well as ultrasonic surgical aspirators, intranasal surgical Doppler and neuronavigation systems—are fundamental for endoscopic procedures.

Neurophysiology monitoring may also prove helpful in selected cases. Somatosensory and motor evoked potentials may be used, along with monitoring of the lower cranial nerves if they are affected by the patient's condition. A neural integrity monitor electromyogram endotracheal tube (NIM 3.0; Medtronic, Minneapolis, MN, USA) allows stimulation of the vagus nerve and assessment of the motor response within the vocal cords. Use of local anaesthetic gels must be avoided with the endotracheal tube and neuromuscular blockade. In addition, it cannot be used with magnetic navigation systems or intraoperative MRI, as it is magnet incompatible. Electrodes may also be placed in the trapezius muscle and tongue to monitor the accessory and hypoglossal nerves, respectively.

Endoscopic Resection

The first step of EEA in CVJ surgery is the entire exposure of the sphenoidal sinus. The approach to the sphenoidal region can be tailored depending on the lateral extension of the tumour, varying from a more conservative parasagittal approach to a more expanded transethmoidal–pterygoidal approach. The final objective is to obtain a whole anterior opening of the sphenoidal sinus, combined with removal of the sphenoidal floor and the posterior third of the vomer, in order to identify the essential anatomical landmarks represented by (1) the paraclival ICAs, medial pterygoidal laminae and pterygoid canals superiorly and laterally; and (2) the Rosenmüller fossae, Eustachian tubes, pharyngobasilar fascia and nasopharyngeal mucosa inferiorly.

If the lesion is located in the lateral portion of the caudal third of the clivus, an endoscopic medial maxillectomy is needed: the opening can employ a combined Denker's approach to achieve better lateral access in removing the medial and lateral pterygoid plates. Another important surgical step is to drill the base of the pterygoid to expose the vidian canal (through which the vidian nerve and vidian artery run), which becomes an important landmark to reach the anterior genu of the ICA, located superiorly to the foramen lacerum, where the paraclival tracts of the ICA are the lateral limits of the superior clival fenestration. Moreover, the vidian canal is an important structure in guiding the surgeon inferolaterally to the anterior ICA, in the petrous bone region, gaining control of the parapharyngeal ICA: in this case the Eustachian tube is used as a superficial anatomical landmark. These approaches provide complete control and mobilization of the ICA (parapharyngeal, horizontal petrous, anterior genu and vertical paraclival segments) when dealing with lesions extending to the foramen magnum, hypoglossal canal, medial occipital condyle and jugular foramen.

Exposure of the clivus, the CVJ and the anterior portion of the foramen magnum is accomplished by making an inferior hinged mucofascial flap with the nasopharyngeal mucosa and pharyngobasilar fascia. The surgical view can be increased by drawing down the soft palate with transnasal rubber catheters. The longus capitis, rectus capitis anterior and anterior AO membrane are resected to expose the anterior ring of C1 and C2, the capsule of the AO joint and the apical ligament. The use of the diode laser to create this flap is very helpful because of its consistency and adherence to the deeper planes. In the inferior clival fenestration the inferolateral limit is represented by the occipital condyles and hypoglossal nerves. When condyle resection is needed to expose a more posterolateral portion of the posterior cranial fossa (e.g. to reach the dural entry point of the vertebral artery), an important anatomical landmark is the supracondylar groove located at the external surface of the condyle: above this groove is the bone of the jugular tubercle, and below this groove is the bone of the occipital condyle. The condyle can be removed up to the level of the hypoglossal canal.

Once the lower clivus and the anterior ring of C1 are drilled, the underlying dura mater and its basilar venous plexus superiorly, as well as the odontoid process inferiorly, are exposed. The ligamentous attachments to the odontoid process (the alar and apical ligaments) are dissected, and the odontoid process is drilled using a high-speed burr. When the dens is removed, the cruciate ligament and tectorial membrane can be incised, exposing the dura. Opening of the posterior fossa dura mater requires particular care and requires identification of the vertebral artery and the vertebrobasilar junction with the aid of surgical Doppler and the neuronavigation system.

In tumour resection, the strategy of choice is identification of the margins and circumferential dissection of the lesion; when that strategy is not feasible, internal debulking come first the centripetal dissection of the tumour margins. These debulking techniques depend on the consistency of the lesions and can be performed with suction or with cutting instruments or powered aspirators (shaver, ultrasonic surgical aspirator). Finally, decompression of the medulla at the CVJ can be achieved by removing the tumour, odontoid process and occipital condyles.

Progressively, depending on the extent of the necessary surgical dissection, it is then possible to identify the following posterior fossa structures: arteries (vertebral arteries, the basilar artery and its perforating pontine branches, anteroinferior cerebellar arteries, posteroinferior cerebellar arteries, superior cerebellar arteries and posterior cerebral arteries); the brainstem with the cerebellopontine angle and upper cervical spine; and cranial nerves II to XII.

Endoscopic Skull Base Reconstruction

Concerning posterior skull base and CVJ duraplasty, the main surgical reconstructive procedure is a combination of free grafting techniques and pedicled flaps (the 'multilayer' technique). The 'triple F' technique (fat, fascia and flap) is mostly used. Free fat grafts are used to fill dead space and form a buttress for a fascia lata inlay graft [5]. The preparatory stage of the duraplasty must include appropriate exposure of the defect, undermining of the dural margins (when possible) and smoothing of the defect edges to produce a tensioactive effect on the flap. Moreover, it is recommended to make the graft/flap larger in diameter than the dural defect, to compensate for its shrinkage during healing. Meticulous management of the tissues is required for the best integration, and it is recommended that a dedicated surgical team perform the reconstruction.

The use of underlay intracranial intradural grafts in association with overlay flaps is the so-called multilayer technique. The technique may involve application of the 'gasket-seal' closure technique, which permits fixation of the graft margins extradurally without risking damage to the neurovascular structures. To accomplish this, the graft is placed on the dural defect and its central portion is pushed inside the defect with the aid of fat or a shaped fragment of cartilage or bone, which is fixed beyond the dural border to seal the closure while still keeping the margins of the fascia outside the skull base [29].

It is worth mentioning that when postoperative radiation therapy is planned, osteocartilaginous grafts should be avoided because of the high risk of necrotic tissue sequestration. In such cases, fat can be used to perform the role and function of the osteocartilaginous grafts in the duraplasty.

Regarding the overlay vascularized flaps, the HBF is the favourite one, though not the only option. The posterior pedicle inferior turbinate flap, the Oliver pedicle palatal flap and the salpingopharyngeus myomucosal flap have all been reportedly used for closure of defects of the posterior cranial fossa and CVJ [24–27]. In patients with more extensive defects, a bilateral HBF (Janus flap) can be used for the closure or, when local flaps are not available, a transposed TPFF is the most appropriate option [23, 30].

In the EEA, reconstruction of the osteodural defect in the region of the CVJ and clivus is challenging not only because of the size of the defects but also because of the high flow of CSF, the lack of supporting structures and the effects of gravity [2]. In these latter sites, the combination of free graft techniques (especially those using abdominal fat covered by fascia lata or a gasket-seal closure) with vascularized flap techniques (above all, the HBF) have decreased the rate of postoperative CSF leakage [31].

The HBF and the TPFF are the two major flaps used in the endoscopic endonasal approach to the CVJ and the clivus. They are described in detail in the following sections.

The Hadad–Bassagasteguy Flap

The Hadad–Bassagasteguy flap (HBF), or nasoseptal flap, is a vascular pedicle flap supplied by the septal branches of the sphenopalatine artery, which is one of the terminal branches of the maxillary artery [22]. The septal branches of the sphenopalatine artery (SPAsb) supply the entire length of the nasal septum and anastomose with the ethmoidal arteries, the greater palatine artery and the anterior facial artery. The flap is planed in relation to the size and shape of the skull base defect, although it is better to overestimate the size and then trim the flap if needed.

The HBF has had a fundamental effect on the advancement and acceptance of the EEA because its use has dramatically decreased the incidence of postsurgical CSF leaks to <5% [32], thus allowing expansion of endoscopic skull base procedures [33]. Nowadays, the HBF is a cornerstone reconstructive technique in the EEA because of its versatility, wide arc of rotation, generous size and relatively ease of harvest. HBF use is less successful in patients who have undergone extensive radiation therapy in the area of the posterior choana [34].

When applied directly or placed over traditional fascia grafts, the HBF provides very strong support and rapid epithelialization, especially in critical areas. A double elevation from both sides of the septum (the so-called Janus flap) is also possible [30].

Surgical Technique

Harvesting the Flap

An HBF may be harvested if the septum and/or the artery are not involved with the tumour. The first step is identification and arrangement of the pedicle of the flap, resulting in preservation of the SPAsb: a superior mucosal incision is made above the tail of the superior turbinate, just below the natural sphenoidal ostium. This first incision is extended medially, passing through the sphenoidal rostrum, and anteriorly on the sagittal plane of the nasal septum, avoiding passing over an imaginary line joining the axillae of the turbinates, in the interests of preserving olfaction. The anterior extension of the superior incision is tailored according to the dimensions of the skull base defect: the anterior limit is constituted by the mucocutaneous junction.

Different options exist on the possibility to create a 'rescue flap' or create the HBF immediately, in relation to the different EEAs.

In the case of the *ipsilateral transpterygoid approach*, creation of the HBF has to be completed at the beginning of the surgery. The inferior incision of the pedicle is performed just above the tail of the middle turbinate, following the

choana and extending through the vomer to the maxillary crest. Laterally both the superior and inferior incisions of the pedicle of the flap have to reach the sphenopalatine foramen. On the sagittal plane of the septum, the inferior incision is extended anteriorly to the same length as that of the superior incision, then an anterior vertical incision joins the superior and inferior ones. If a wider flap is needed, the inferior incision can be placed in the lateral portion of the nasal floor, in the inferior meatus. All incisions can be modified according to the reconstructive or oncological requirements. The real dimension of the skull base defect and the distance to the sphenopalatine foramen can be evaluated by using a surgical cotton patty as a template for harvesting the HBF. Subperichondrial/subperiosteal dissection of the flap allows its entire mobilization. Therefore, the transpterygoid approach can be completed with a wide antrostomy, with removal of the orbital process of the palatine bone and the posterior wall of the maxillary sinus. The pterygopalatine fossa is opened and its contents, together with the periosteum, are lateralized to free the pedicle of the flap. The vidian artery and the descending palatine and palatovaginal arteries are transected, releasing the pedicle of the flap laterally and allowing greater length and mobility, as well as safe drilling of the medial and lateral pterygoid plates. The flap is finally placed in the maxillary sinus to be protected during surgery [35, 36].

If the HBF is *contralateral* to the lesion, a nasoseptal *rescue flap* can be arranged by dissecting the mucosa immediately below the superior incision in a subperiosteal manner [37]. The rescue flap is placed at the same level or below the floor of the sphenoidal sinus, protecting the pedicle from the instruments.

When an *ipsilateral transclival approach* is planned to reach the inferior aspect of the sphenoid sinus and clivus, creation of a modified rescue flap is suggested. It consists of performing both the inferior incision of the pedicle and dissection of the inferior aspect of the flap at the beginning of the procedure. This leads to increased mobility of the pedicle, which can be placed more inferiorly [38]. At the end of the surgical procedure the full pedicled HBF can be harvested, following the same steps as those used for the ipsilateral transpterygoid approach, extending the rescue flap and modified rescue flap incisions into standard HBF incisions.

The Temporoparietal Fascia Flap

The temporoparietal fascia flap (TPFF), or the galeal flap, is one of the most versatile flaps in skull base reconstruction. The temporoparietal fascia is located immediately deep to the skin and the subcutaneous tissue of the scalp in the temporal and parietal regions. It is vascularized by the superficial temporal artery, a terminal branch of the external carotid artery. Normally this arterial vessel has a winding course and splits into anterior (frontal) and posterior (parietal) branches 2–4 cm above the zygomatic arch. The frontal branch of the facial nerve is known to course within the innominate fascia, a plane deep to the subcutaneous musculoaponeurotic system (SMAS) and superficial temporal fascia, obliquely 1.5 cm lateral to the eyebrow and not more than 2 cm above the brow, which represents the anterior limit of the dissection to prevent iatrogenic injury to the facial nerve [39, 40].

The TPFF supplies generous healthy and well-vascularized tissue to reconstruct the skull base in the posterior cranial fossa. It has several advantages in comparison with other reconstruction techniques: predictable vascular anatomy, a good arch of rotation, the length of the vascular pedicle, and rich vascularization with subsequent quick healing even in unfavourable conditions such as in previously irradiated patients [23]. The size of the TPFF can be chosen according to the intraoperative needs: surgeons have to consider whether there is a risk of transient or persistent alopecia, correlated with the flap thickness. Moreover, its remarkable tractability permits it to be employed in extensive posterior cranial fossa reconstruction presenting problematic multiplanar surfaces (Fig. 4) [41].

Surgical Technique

Harvesting the Flap

The patient is placed in a supine position with the head rotated contralaterally to the donor side. A hemicoronal incision is made among the hair follicles, from the superior aspect of the preauricular area to the vertex of the head. The incision is made just through the dermis with caution so as not to injure the superficial temporal vessels. In the scalp area, a very thin flap is elevated just under the hair follicles: the dissection is performed superiorly toward the vertex of the scalp. When elevation of the anterior scalp is concluded, the same procedure is done for the posterior edge. At this stage of the intervention, the use of electrocautery should be avoided because of the closeness to the hair follicles. The required size of the TPFF is given by evaluating the extension of the skull base defect and its gap from the infratemporal fossa. After that, the TPFF can be harvested by first incising the fascia and staying superficial to the temporalis muscle fascia. The flap is elevated in a fan-shaped manner with the narrow end located in the preauricular area. The scalp is closed in layers over a suction drain.

Creating the Infratemporal Corridor

The deep temporal fascia is then incised along the free edge of the zygomatic arch and 1 cm from the free edge of the orbital process of the zygomatic bone to preserve the frontal branch of the facial nerve. The coronoid process is identified by sliding a finger along the anterior border of the temporalis muscle. In this way an infratemporal corridor for transposition of the flap is obtained, connecting the temporal, infratemporal, pterygopalatine and nasal fossae.

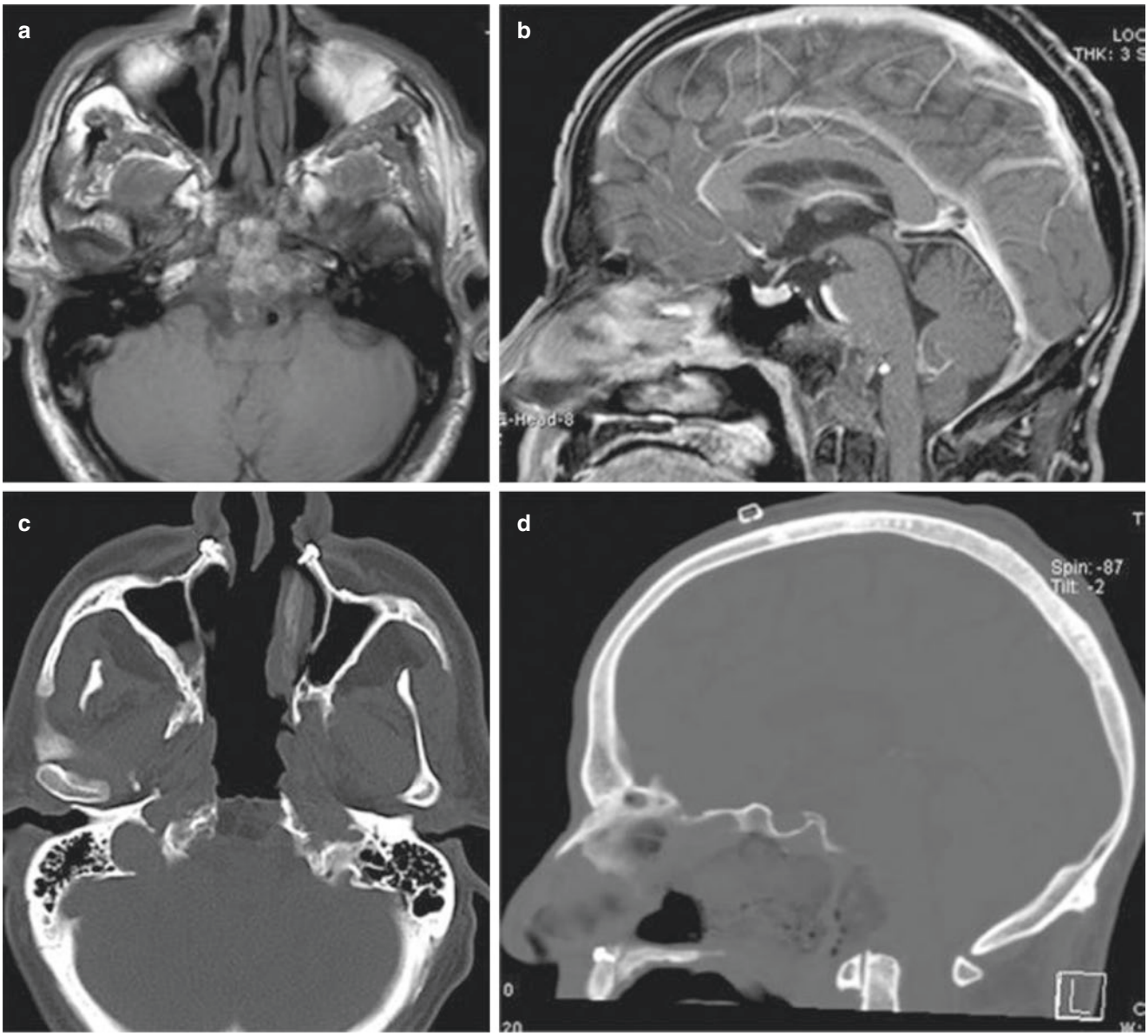

Fig. 4 (**a**) Preoperative axial T1-weighted and (**b**) preoperative sagittal T1-weighted gadolinium-enhanced magnetic resonance images (MRIs) showing a chordoma involving the middle and inferior portion of the clivus, as well as the foramen magnum and the superior portion of the odontoid process. Note that the basilar artery is dislocated but not encased by the tumour. (**c**) Postoperative axial and (**d**) postoperative sagittal computed tomography (CT) scans showing craniocaudal and lateral extension of resection after use of an exclusive endoscopic endonasal approach, with preservation of the occipitoaxial joint

Transpterygoid Transposition of the Flap

Through the transmaxillary corridor that was previously created (during the resection), the fat of the infratemporal fossa is removed. A Ciaglia percutaneous tracheostomy set (Cook percutaneous tracheostomy introducer set; Cook Critical Care, Bloomington, IN, USA) is used to create the tunnel positioning the atraumatic tube into the infratemporal fossa, which is then widened with the dilatators provided. Once a tunnel wide enough to not compress the vascular pedicle of the flap is obtained, a guide wire is fixed to the distal portion of the flap by a stitch while the proximal portion is inserted through the infratemporal corridor, and the flap is finally pulled into the nasal cavity. During this procedure, attention is paid to avoid any torsion of the vascular pedicle. The flap is positioned over the defect and fixed with Surgicel and fibrin glue.

Postoperative Care and Follow-Up

Patients receive prophylactic antibiotic therapy (to be started at the time of intubation and continued for no more than 24 h after the surgery, except in cases of documented infection) with a second-generation cephalosporin (e.g. cefuroxime

2 g) associated with metronidazole 500 mg, both in cases of exclusive intranasal surgery and in cases of intracranial surgery. Steroid therapy with deflazacort 30 mg for 5 days, reducing the dose progressively to 7.5 mg (total: 15 days) or prednisone 25 mg, reducing the dose progressively to 6.25 mg, can be added for anti-edemigenic purposes.

Nasal packing is removed on the second postoperative day and endoscopic endonasal care is performed, removing crusts, debris and clots. Irrigation with a saline solution twice daily is recommended for several weeks. Patients are discharged on approximately the fourth postoperative day and are usually advised against blowing their nose, bending their head forward or undertaking physical activity for at least 1 month.

In cases of osteodural reconstruction, a brain CT scan is performed 24 h after surgery for early detection of any signs of postoperative pneumocephalus and then repeated if considered necessary. Bed rest is maintained for 2 days and the patient is regularly monitored with complete blood tests 1, 5 and 10 days after surgery, at which time he or she is usually discharged. Lumbar drainage is generally not applied. If early CSF leakage is only suspected in the early postoperative period, a lumbar drain can be inserted to decrease intracranial pressure and facilitate healing. If it becomes evident that CSF leakage is present, surgical revision of the duraplasty should be performed immediately.

The first endoscopic evaluation is carried out 2–3 weeks after hospital discharge. Subsequently, patients are followed up by endoscopic controls at progressive intervals after 1, 3, 6, 9 and 12 months, then every 6 months for at least 4 years and once yearly thereafter. Postoperative enhanced MRI (T1, enhanced T1, T2, T2 fluid-attenuated inversion recovery [FLAIR], constructive interference in steady state [CISS]) or enhanced CT scanning is performed every 4 months for the first year and then every 6 months in the following years.

Outcomes

Surgical outcomes depend on the type of pathological condition treated and the surgical approach used. The most common condition of the clivus and CVJ treated with the EEA is a chordoma. A recent review of the literature, comprising 100 cases of endoscopic endonasal treatment of clival chordomas, found a gross total resection rate of 70.4% [42]. Chibbaro et al. reported a gross total resection rate of 65% from a series of 54 patients affected by CVJ chordomas, with a mean follow-up period of 34 months [8]. The tumour location, neurovascular structures involved and previous treatments are factors that inevitably affect the resectability of the tumour. In this regard, Koutourousiou et al. reported a discrepancy between the gross total resection rates achieved in primary tumours and those achieved when dealing with recurrent CVJ chordomas (83% versus 44%, with a mean follow-up period of 33 months) [43]. The results emerging from the major series currently available in the English-language literature describing use of the EEA for removal of clival and CVJ chordomas are detailed in Table 1.

Most of the available studies regarding the validity of the EEA in the context of CVJ tumours different from chordomas (e.g. chondrosarcomas, metastatic lesions, chondromas or meningiomas) have been small, with short follow-up periods (Fig. 5). They have also been extremely heterogeneous with respect to their patient inclusion criteria, treatment protocols, and methods of data collection and analysis. Table 1 summarizes the findings of the most relevant studies available to date. It should be noted that petroclival lesions were not considered for the present analysis, which focuses only on tumours specifically affecting the CVJ.

Complications

The main intraoperative complication is bleeding. In this regard, one potential concern with endonasal approaches is the ability to achieve haemostasis. Adapted techniques, haemostatic agents and specialized instrumentation designed for endoscopic endonasal procedures—including diamond burrs, injectable haemostatic agents, warm irrigation and bipolar forceps devices—have all made haemostasis feasible. Injury to major arteries (e.g. the carotid, vertebral and basilar arteries) is the most feared complication associated with use of the EEA in CVJ surgery. Avoidance of this complication first requires accurate preoperative imaging evaluation. Intraoperatively, accurate image guidance and use of intranasal surgical Doppler can facilitate identification of arteries. If an injury occurs, large-bore suction is used to control the surgical field and identify the source of bleeding. There are several options for haemostasis, which include bipolar cauterization to weld the defect shut, direct compression, compressive packing, suture repair, or reconstruction using clips (e.g. an aneurysm clip or Sundt clip graft). Crushed muscle, emergently obtained from the thigh, can be useful to induce thrombosis when placed over the defect under compression. Angiography should be performed in the acute and delayed phases to assess the patency of the artery and to rule out pseudoaneurysm formation. There have been anecdotal reports of carotid artery or other major vessel injuries, with a low cumulative incidence less than 0.3–0.5% at centres with extensive experience [4, 53]. Lastly, there is a risk of intraoperative damage to the lower cranial nerves and lower brainstem. Careful and gentle dissection, along with maintenance of adequate cerebral perfusion pressure, is necessary. Intraoperative neuromonitoring is useful to obtain real-time feedback on any unwanted nerve traction or manipulation. In addition, it may be prudent to leave behind a

Table 1 Results of the most relevant studies reporting the endoscopic endonasal approach to manage clival and craniovertebral junction tumours

Study authors, year	Patients [*n*]	Histology (*n*)	Resection (% or *n*)	Complications (% or *n*)	Mean follow-up period	Recurrence rate (%)
Frank et al., 2006 [44]	9	9 chordoma	33% GTR	None	24 months	11%
Carrabba et al., 2008 [45]	12	12 chordoma	59% GTR	24% CSF leakage 6% neuropathy	16 months	0%
Stippler et al., 2009 [15]	20	20 chordoma	52% GTR	25% CSF leakage 5% neuropathy	13 months	10%
Saito et al., 2012 [42]	6	6 chordoma	50% GTR	17% meningitis	NA	NA
Koutourousiou et al., 2012 [43]	60	60 chordoma	67% GTR	27% CSF leakage 7% neuropathy	18 months	33%
Tan et al., 2012 [46]	14	14 chordoma	50% GTR	21% CSF leakage	42 months	15%
Chibbaro et al., 2014 [8]	54	54 chordoma	65% GTR	8% CSF leakage 14% meningitis	34 months	11%
Mangussi-Gomes et al., 2016 [2]	32	32 chordoma	47% GTR	22% CSF leakage 12% meningitis	12 months	NA
Zhang et al., 2008 [47]	8	6 chordoma 2 chondrosarcoma	7 GTR 1 STR	None	3–39 months	16%
Vellutini et al., 2014 [5]	38	26 chordoma 2 chondrosarcoma 6 metastasis 1 fibrous dysplasia 1 meningioma 2 other	48% GTR	19% CSF leakage 9.5% mortality	6 months to 11 years	NA
Messerer et al., 2016 [48]	11	3 chordoma 3 meningioma 3 metastasis 1 chondroma 1 chondrosarcoma	4 GTR 7 STR	2 neuropathy	NA	NA
Alexander et al., 2010 [49]	1	1 meningioma	1 NTR	1 CSF leakage	14 months	0%
Prosser et al., 2012 [50]	1	1 meningioma	1 NTR	1 palsy (cranial nerve VI)	NA	NA
Simal et al., 2014 [51]	1	1 meningioma	1 GTR	None	NA	NA
Iacoangeli et al., 2014 [52]	2	2 meningioma	2 GTR	None	6 months	0%
Beer-Furlan et al., 2016 [6]	3	3 meningioma	2 NTR 1 STR	1 CSF leakage 1 septic shock 1 hypopituitarism	NA	NA

CSF cerebrospinal fluid, *GTR* gross total resection, *NA* not available, *NTR* near-total resection, *STR* subtotal resection

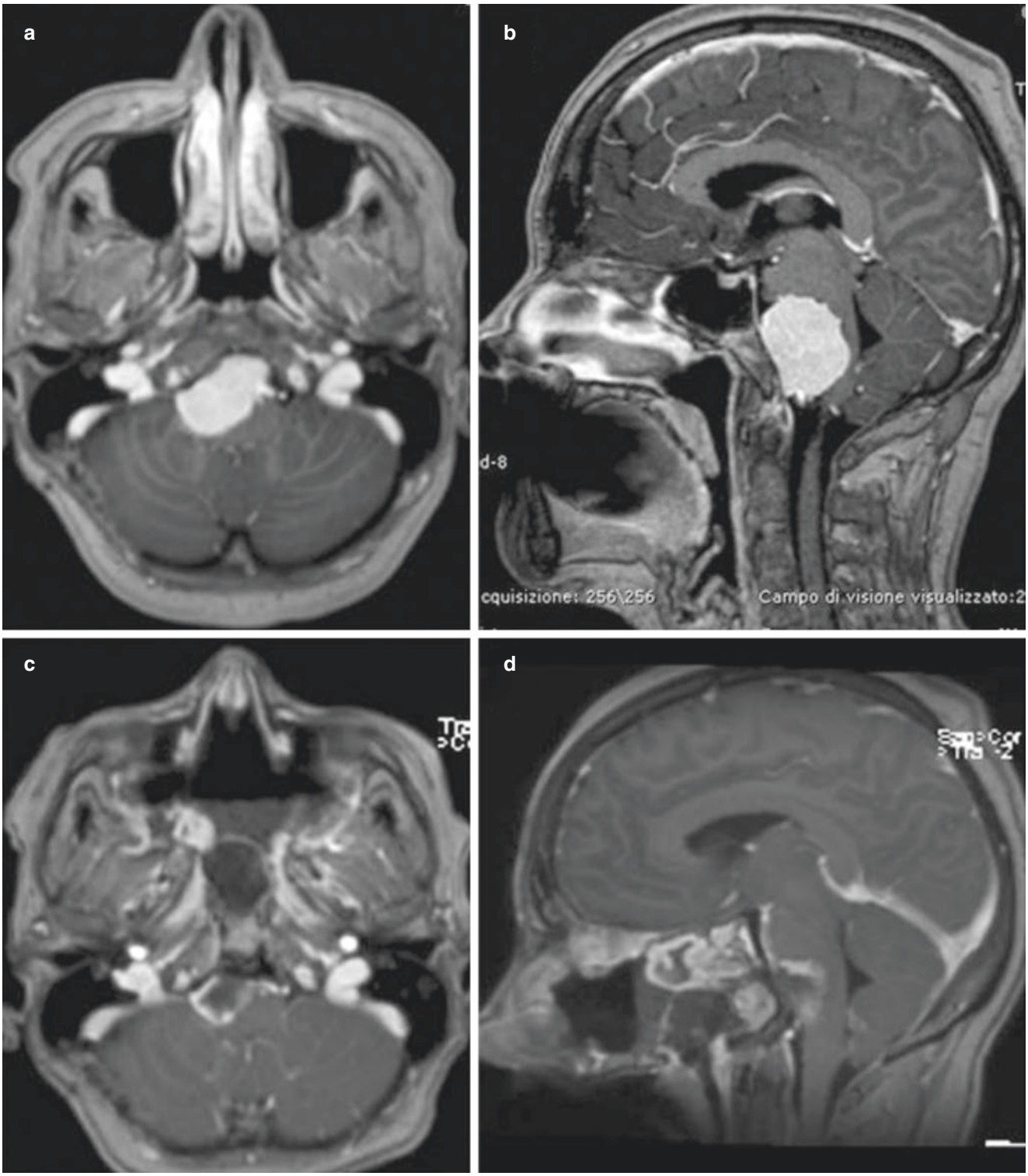

Fig. 5 (**a**) Preoperative axial and (**b**) preoperative sagittal T1-weighted gadolinium-enhanced magnetic resonance images (MRIs) of a posterior cranial fossa meningioma at the level of the middle and inferior portion of the clivus and the superior portion of the odontoid process, with compression of the brainstem and upper cervical spine. (**c**) Postoperative axial and (**d**) postoperative sagittal T1-weighted gadolinium-enhanced MRIs showing craniocaudal and lateral extension of resection after use of an exclusive endoscopic endonasal approach with decompression of the brainstem and upper cervical spine

tumour that is significantly adherent to critical neurovascular structures. At present, the incidence of a new permanent neurological complication is described as being between 5% and 7% in the largest published series [1, 6, 15].

In the postoperative period, CSF leakage represent the main concern and may occur despite meticulous reconstruction. We favour early surgical re-exploration in patients with postoperative CSF leakage to identify the site of the leak. An intrathecal fluorescein injection may be used to facilitate identification of the CSF fistula site in cases of low-flow leaks. CSF drainage through a lumbar subarachnoid drain is used only as an adjunctive measure after surgical repair, as CSF drainage with an open defect may lead to symptomatic pneumocephalus. The incidence of postoperative CSF leakage was 18.8% in a series of 80 cases treated for CVJ chordoma [42]. Other reports are consistent with this finding, describing postoperative CSF leakage rates ranging from 17.6% to 25% [4, 15].

In addition, meningitis and ascending infections (e.g. intracranial abscesses) are life-threatening complications, which are rarely described (with reported incidence rates from 0% to 17%) but must be recognized and treated early [1, 2]. One fatal case of meningitis complicated by sepsis was described recently, representing a procedure-related mortality of 1.4% [54].

Other minor early or late complications reported in the existing literature are facial numbness, periorbital swelling, oroantral fistula, V2 paraesthesia, anosmia, sinonasal scarring with sinusotomy closure and recurrent rhinosinusitis. Another reported complication is transient velopharyngeal insufficiency, which occurred in 4.7% of patients in a systematic review of 85 patients undergoing CVJ surgery via the EEA. However, this complication usually resolves within 6 months after surgery [4].

Conclusion

The endoscopic endonasal approach (EEA) to craniovertebral junction (CVJ) tumours appears to be a reasonable alternative to the traditional transoral approach in selected cases. Use of the EEA in CVJ surgery has a high success rate in achieving stable decompression and improvements in neurological status, with acceptable intraoperative and postoperative complication rates. Moreover, the EEA offers advantages such as lower rates of postoperative dysphagia and respiratory complications in comparison with the more traditional approaches. However, this approach should be considered complementary, rather than an alternative, to the traditional transoral, transcervical and far lateral approaches, which are still used for laterally extended CVJ tumours.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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The Endoscopic Endonasal Approach for Treatment of Craniovertebral Junction Pathologies: A Minimally Invasive but not Minimal-Risk Approach

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Introduction

Several pathologies can affect the craniovertebral junction (CVJ), leading to bulbomedullary compression. The microsurgical transoral approach (TOA) has long been considered the 'gold standard' for anterior decompression [1]. However, a series of complications, particularly with extended procedures [2–4], has recently led to the introduction of the endoscopic endonasal approach (EEA), which showed initial promise for overcoming previous technical challenges and surgical complications [5]. So far, however, only limited reports on this approach are available [6–18]; therefore, clear evidence of its superiority is yet to be demonstrated. In this paper we report our surgical experience with the EEA for treatment of different CVJ pathologies. The feasibility, advantages and limitations of this approach are critically assessed.

Material and Methods

Among 21 cases operated on using anterior approaches, from 2011 to 2018, a consecutive series of six patients with varied CVJ pathologies were treated using the EEA approach by a mixed team of neurosurgeons and rhinolaryngologists, who are experts in microneurosurgery and endoscopic video-assisted microsurgery (Table 1). In every patient, a subsequent short occipitocervical instrumentation and fusion procedure was completed 1 week later.

Results

The length of follow-up of our patients ranges from 1 to 48 months (Table 1 [19, 20]); all of the patients improved or remained unchanged (with a good neurological status) after surgery and during follow-up. However, in one patient (patient #1), intraoperative cerebrospinal fluid (CSF) leakage occurred, requiring immediate surgical repair with a mucosa pedicle flap. The patient improved after surgery within 48 h and was able to walk and feed herself without any assistance for 10 days, but she subsequently deteriorated rapidly and died 1 month after the surgery. Since the patient was immobilized with a halo vest, CSF nose leakage was not evident either spontaneously or with a jugular compression test (the Queckenstedt test). During the subsequent posterior fixation procedure performed in the prone position, CSF leakage was evident in the operating room, and an external CSF drain was set up and left in place for 5 days, achieving stable control of the leakage.

Postoperatively the patient developed meningoencephalitis, confirmed by positive CSF cultures for fungi (*Candida albicans*). Despite appropriate antimycotic therapy (micafungin and 5-fluorocytosine [flucytosine]), the meningoencephalitis progressively worsened, leading to fatal neurological complications.

All other patients were discharged after posterior instrumentation and fusion procedures within 2 weeks after admission.

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Table 31.1 Personal series of patients operated with EEA

Patient	Age (sex)	Primary disease	Radiology	Pre-op C1–C2 shift (X-rays)	Treatment	Post-op shift (X-rays)	Frankel scale [22] and Di Lorenzo Grade [23] changes	External orthosis	Follow-up (months)
PF #1	66 (F)	Impressio basilaris	CVJ anterior compression	Yes	(1) Transnasal decompression (0–30°) (2) C0–C3 reduction, lateral masses screws instrumentation and heterologous bone fusion	No	D/E I/I	Halovest (1 month)	6 (meningitis and death)
RS #2	47 (F)	Impressio basilaris + platybasia	CVJ anterior compression	Yes	(1) Transnasal C1 decompression and odontoidectomy (2) C0–C3 reduction, C2 pedicles and C3 lateral masses screws Instrumentation and autologous bone fusion	No	D/E I/I	Halovest (1 month)	49
LP P #3	14 (F)	Impressio basilaris + platybasia	CVJ anterior compression	No	Transnasal C1 partial decompression and partial odontoidectomy	No	D/E I/I	No	51
R N #4	22 (M)	Recurrence of clivus Chordoma	CVJ destruction and compression	Not performed	(1) Transnasal C1 decompression and odontoidectomy (2) C0–C3 reduction, lateral masses screws instrumentation and heterologous bone fusion	NO	D/E I/I	Halovest 1 month Hard collar (2 months)	34
G M #5	58 (M)	Myeloma	Odontoid infiltration	Not performed	Transnasal and odontoid biopsy		E/E I/I	No	27
O S #6	35 (F)	Chordoma	CVJ infiltration previously operated (1) transoral and (2) C0–C3 reduction, lateral masses	Not performed	Transnasal decompression (0–30°)	NO	D/D I/I	Previously performed	39
RC #7	36 (M)	Isolated plasmocytoma	CVJ anterior compression	Not performed	One stage transnasal C1 decompression, and odontoidectomy and C0–C3 reduction, lateral masses screws instrumentation and heterologous bone fusion	NO	D/D I/I	Soft collar (1 month)	3

Discussion

In this paper we present our recent institutional experience dealing with six patients affected by CVJ disease and operated on using the EEA. Of these, five had an uncomplicated postoperative course and one (patient #1) developed intraoperative CSF leakage and died 5 weeks after surgery.

A critical analysis of such a case warrants development of strategies that might decrease the risk of such complications developing in future patients. More specifically, we strongly support the following measures: (1) making every effort to avoid dural tears, as far as possible, by overcoming the learning curve; and (2) using a single-stage combined anterior and posterior approach to allow earlier identification of a possible CSF fistula—without the obstacle of a halo vest—and immediate commencement of adequate therapies.

A total of 107 patients (including our six) affected by CVJ disease and treated with the EEA have been reported in the literature so far. Among these cases, CSF leakage (intra- and/or postoperative) was reported in 13 cases (12.4%), transient velopharyngeal incompetence variably associated with nasal speech and swallowing impairment was reported in six cases (5.6%), postoperative epistaxis was reported in two cases (1.86%) and respiratory dysfunction requiring a tracheostomy was reported in two cases (1.86%). Interestingly, in our extended institutional series of more than 20 consecutive anterior decompressions for CVJ disease (including use of the transoral and transnasal microsurgical approaches), the only fatal complication was associated with use of the EEA.

According to the literature, and our personal experience, the presumed improved safety of the EEA, in comparison with the TOA, should be reassessed. In fact, a recent systematic review and meta-analysis by Shriver et al. showed that the EEA was associated with higher rates of 30-day mortality (4.4% versus 2.9%), intraoperative CSF leakage (30% versus 0.3%), postoperative CSF leakage (5.2% versus 0.8%), meningitis (4% versus 1%), need for prolonged intubation or re-intubation (6% versus 5.6%), need for reoperation (5.1% versus 2.5%), velopharyngeal insufficiency (6.4% versus 3.3%) and sepsis (7.7% versus 1.9%), while the TOA was associated with higher rates of arterial injury (1.9% versus 0%), wound infection (3.3% versus 1.9%) and need for tracheostomy (10.8% versus 3.4%). However, none of these differences was statistically significant, except for the difference in the need for postoperative tracheostomy [21].

Conclusion

Our experience and other reported experiences with the endoscopic endonasal approach (EEA) have highlighted occurrences of complications similar to those observed with the transoral approach (TOA) in craniovertebral junction surgery, including velopharyngeal insufficiency and serious infections, leading us to reconsider the presumed superiority of the EEA over the endoscopic TOA.

The most appropriate treatment should therefore be chosen on the basis of the individual anatomy of the patient and the personal experience of the surgeon.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Stability-Sparing Endoscopic Endonasal Odontoidectomy in a Malformative Craniovertebral Junction: Case Report and Biomechanical Considerations

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Abstract *Background:* The craniovertebral junction (CVJ) is often involved in a wide range of congenital, developmental and acquired pathologies that can create bony and ligamentous instability or cause direct compression on the medulla and cervical spine cord, resulting in significant impairment. Atlas assimilation is the most common malformation in the CVJ and can be frequently associated with basilar invagination (BI) and Chiari malformation (CM) type I. Posterior atlas assimilation more frequently leads to BI type II with a mass effect on neural structures but usually no signs of biomechanical instability. Operative approaches to the CVJ have undergone a remarkable evolution and can be divided into ventral, lateral and dorsal ones. In this kind of surgery, it is vital to detect and eventually treat any CVJ instability.

Case Description: We present a case of CVJ malformation comprising assimilation of the posterior arch of the atlas, BI type II and CM, treated by endoscopic endonasal odontoidectomy and partial clivus removal to spare CVJ stability.

Conclusion: Neurological and biomechanical analysis of all CVJ malformations permits stratification and selection of those cases that can be managed by simple, direct, minimally invasive decompression with no need for surgical fusion.

Keywords Cranio-vertebral junction · Cranio-cervical malformation · Atlas assimilation · Basilar invagination · Chiari I malformation · Endoscopic endonasal odontoidectomy

Abbreviations

3D	Three-dimensional
BI	Basilar invagination
CM	Chiari malformation
CSF	Cerebro-spinal fluid
CT	Computed tomography
CTA	Computed tomography angiography
CVJ	Craniovertebral junction
EEG	Electroencephalography
MRI	Magnetic resonance imaging
NPL	Nasopalatine line
PL	Palatine line
RX	Radiography

Introduction

The craniovertebral junction (CVJ) represents the top of the spinal axis, and it has a complex musculoskeletal organization: its bony anatomy and its joint configuration, shape and orientation are unique in comparison with the rest of the cervical spine [1]. This arrangement of structures is the basis of complex movements such as flexion–extension and turning of the head, and it also protects the bulbocervical junction, which contains areas critical for life. The CVJ is often involved in a multitude of congenital, developmental and acquired pathologies [2] that can create bony or ligamentous instability, direct compression and a mass effect on the medulla and cervical spine cord, resulting in significant impairment.

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Among the wide range of isolated malformations affecting the CVJ [3, 4], atlas assimilation is the most common one [5, 6]. This condition has been associated with craniocervical instability and basilar invagination (BI) in isolated cases. Goel distinguished two forms of BI, named types I and II [7]. Type I manifests itself with instability and ventral neural compression caused by the dens of the axis, while the key point for definition of BI type II is the contextual presence of a Chiari malformation (CM) with posterior cord compression, but usually without instability. In cases of BI secondary to atlas assimilation, the anterior form of assimilation leads to BI type I and atlantoaxial instability; otherwise, posterior atlas assimilation more frequently causes BI type II and usually no biomechanical stability deficits.

Operative approaches to the CVJ have undergone a remarkable evolution and advancements over the last 100 years, so that nowadays it is possible to divide them into ventral, lateral and dorsal ones [8]. Traditionally, the transoral approach has been considered the gold standard for addressing high cervical spine pathologies [9]; however, the risk of bacterial contamination, need for prolonged postoperative intubation and nasogastric tube feeding, tongue swelling and nasopharyngeal incompetence after this surgery [10] have encouraged use of the endoscopic transnasal approach to the CVJ. This latter approach was first described in 2002 [11] and in the last 10 years it has provided more options for decompression of irreducible ventral CVJ pathology [12]. Nowadays, the extended endoscopic endonasal approaches enjoy wider acceptance for improved visualization of the anatomically distant surgical field and early recognition of vital neurovascular structures, surgical borders and lesion margins [13, 14]. Furthermore, intraoperative navigation and use of modern devices (lasers and differently angled three-dimensional endoscopes) allows more accurate localization of anatomical structures and fewer complications during endoscopic CVJ surgery.

Endoscopic endonasal odontoidectomy is recommended in cases of soft tissue pannus or irreducible bony compression of the brainstem spinal cord, causing progressive myelopathy or neurological symptoms [9, 15]; after this kind of surgery, it is vital to be sure that the CVJ is stable.

Here we present a case of CVJ malformation comprising assimilation of the posterior arch of the atlas, BI and CM, which was treated by endoscopic endonasal odontoidectomy with no need for surgical posterior fusion. We aim to emphasize how correct analysis of each single malformative CVJ is mandatory to understand its unique biomechanics and stability, and to propose the best and least invasive treatment possible for the patient.

Case Description

The patient, a 63-year-old man with a history of hypertension and chronic bronchopneumopathy, was admitted to the neurological department of our hospital, complaining of general malaise, bilateral leg pain and a recent episode of epileptic crisis. Neurological examination showed gait abnormality with hypotonia and a reduction in motor power in the lower extremities.

He underwent brain magnetic resonance imaging (MRI), which demonstrated the presence of platybasia associated with BI and CM type I (Fig. 1a). Moreover, preoperative computed tomography angiography (CTA) showed assimilation of C1 with exclusion of vascular alterations (Fig. 1b). Cerebrospinal fluid analysis was normal. Electroencephalography showed irritative activity in the left brain, and electroneuromyography and somatosensory motor evoked potentials demonstrated alterations of neuronal conduction in the left leg.

After careful consideration of the patient's clinical and radiological status, the height of the odontoid, the anterior ring of C1, and the inferior edge of the clivus above the palatine line (PL) and nasopalatine line (NPL) [16], the presence of atlanto-occipital fusion and the possibility of preserving the anterior ring of C1 and the transverse ligament, we decided to perform endoscopic transnasal decompression of the CVJ with no need for secondary posterior screw fixation.

The patient was placed in a supine position with his head in a Mayfield headholder. The somatosensory evoked potentials were monitored throughout the surgical procedure, and intraoperative navigation was used. The procedure was performed by an otolaryngologist and a neurosurgeon working in tandem. An inferior septectomy was performed, removing 2 cm of the vomer bone at its junction with the hard palate. After the inferior sphenoidotomy, a nasopharyngeal flap was made; this allows better closure, facilitating the wound-healing process. The clivus with the body and arch of C1 and C2 were exposed and the caudal part of the clivus was removed using a combination of a high-speed drill, an ultrasonic aspirator with a bone tip, and standard curettes. The surgical cavity was subsequently inspected by 30° and 45° angled lens endoscopy, which confirmed the integrity of the transverse ligament. Finally, reconstruction was done with an autologous fascia lata patch [17] and reapproximation of the nasopharyngeal defect. The patient was kept intubated postoperatively and taken to the intensive care unit, where he was extubated uneventfully the next day. No major postoperative complications occurred, and oral feeding was imme-

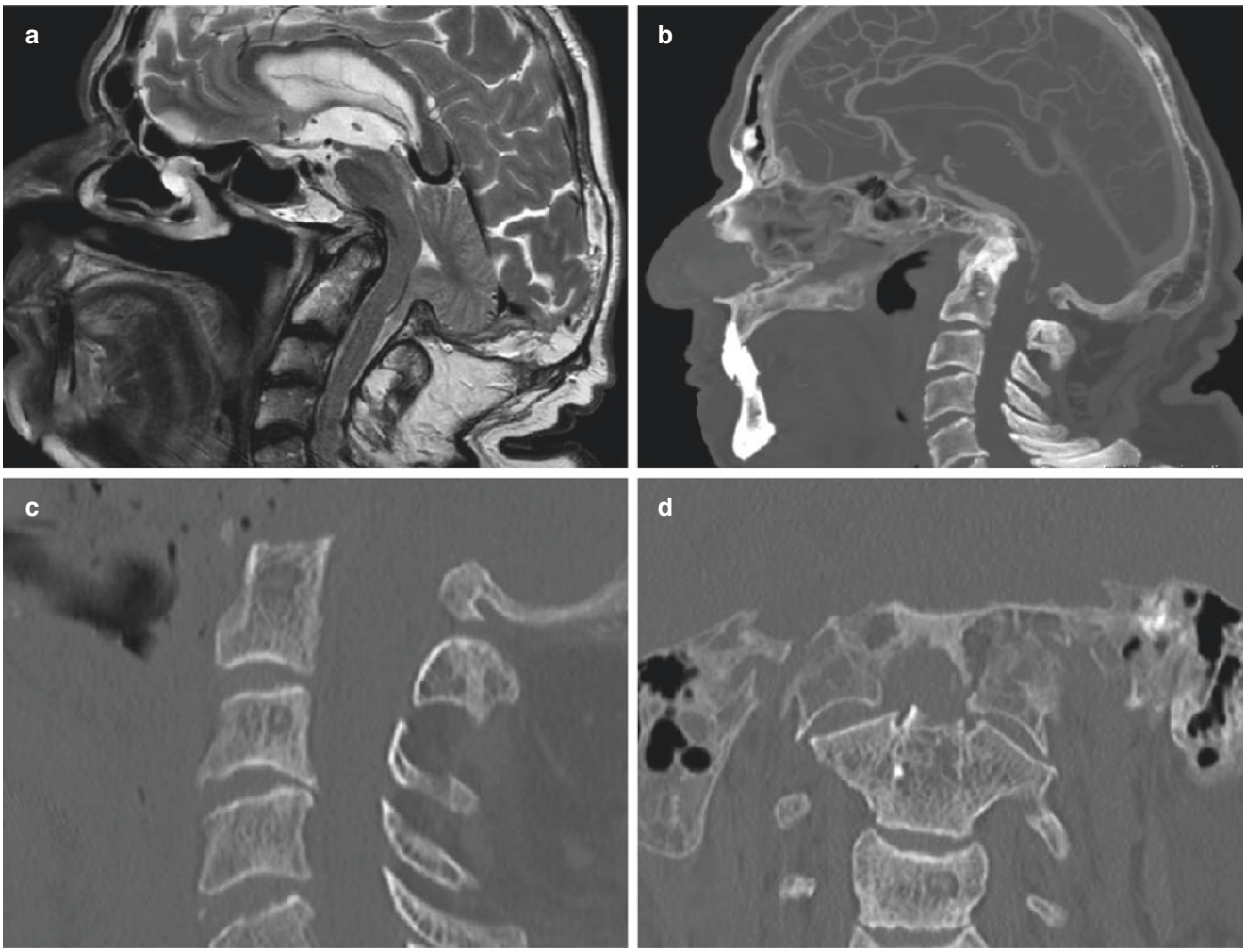

Fig. 1 (**a**) Preoperative magnetic resonance imaging (MRI) brain and cervical scan: sagittal T2-weighted sequence showing a craniovertebral junction (CVJ) malformation comprising basilar invagination (BI) and Chiari malformation (CM) type I. (**b**) Preoperative computed tomography angiography (CTA) brain and cervical scan: sagittal sequence showing assimilation of the posterior arch of the atlas and excluding intracranial vascular anomalies. (**c**) Postoperative computed tomography (CT) brain and cervical scan: sagittal bone-setting sequence showing odontoidectomy with sparing of the inferior half of the anterior arch of the atlas. (**d**) Postoperative CT brain and cervical scan: coronal bone-setting sequence showing odontoidectomy and removal of the caudal portion of the clivus

diately restarted. The neurological examination showed complete resolution of the patient's preoperative symptoms without rhinorrhoea, velopharyngeal insufficiency or Eustachian tube dysfunction. Postoperative CT scanning (Fig. 1c, d) confirmed resolution of the cervicomedullary compression without any complications, and the patient was discharged home after 10 days. A follow-up neurological examination 3 months after the surgery revealed 5/5 strength in the upper and lower extremities without signs of spinal instability, and no meningeal signs or rhinorrhoea were present. Endoscopic visualization of the surgical site showed a completely remucosalized nasopharynx without gross evidence of surgical manipulation. By this time the patient had returned to work.

The patient supplied his informed consent for publication of a case report about his clinical history.

Discussion

The surgical treatment of bulbomedullary junction compressive lesions can vary. The nature and extension of the pathology—and, above all, the reducibility or irreducibility of the

abnormality—usually dictate the type of surgical procedure to perform [16].

If the bulbomedullary junction compression is reducible, stand-alone surgical reduction simply permits indirect decompression of neural structures and posterior occipitocervical or C1–C2 fusion is required only to maintain long-term reduction [16, 18, 19]. In cases of irreducible spinal cord lesions with progressive neurological deterioration, direct decompression is mandatory and should be achieved via a ventral or posterior approach, tailored according to the site of maximum compression, with associated posterior arthrodesis when preoperative or postoperative iatrogenic instability can be demonstrated [20]. These considerations have been extensively described by Menezes et al. [21], whose treatment algorithm has confirmed its validity over the years and has subsequently been modified by Dlhouy et al. [22].

The need for posterior surgical fusion is often considered inevitable and in the decisional surgical process it is not considered as an avoidable surgery in selected cases. Actually, posterior malformative CVJ fixation often represents a surgical challenge and is threatened by frequent complications, especially in patients with many comorbidities and those already treated by a ventral approach (transoral or transnasal). If posterior fusion is performed in the context of direct decompression, it increases the surgical time and the risk of complications. Conversely, a delayed secondary posterior approach exposes patients to a period of CVJ instability until fusion is performed.

Biomechanics in a malformative CVJ do not follow the same rules as those in a normal CVJ, precisely because of anatomical abnormalities such as atlas assimilation, BI and CM [5]. In some of these cases, during preoperative treatment planning it is possible to predict potential stability of the CVJ after decompression, make a decision about the extension of decompression and possibly exclude the need for fusion [5, 22].

Preoperative analysis of the present case confirmed that the endoscopic endonasal approach was the surgical first choice, with a favourable trajectory through the nasal bone and hard palate (NPL) [16]. The malformative CVJ of our patient showed integrity of the transverse ligament, integrity of both the joints between the occipital condyles and the lateral masses of the atlas, and assimilation of the posterior arch of the atlas. In particular, the present case seemed to be a type II atlas assimilation [7] with occipitalization of the posterior arch associated with BI and CM type I. Therefore, we decided to perform anterior decompression via an endoscopic endonasal approach, performing complete odontoidectomy and removal of the superior half of the anterior arch of the atlas, with no alteration in the transverse ligament and C1 ring integrity. The bone decompression was also extended through removal of the most caudal and posterior part of the clivus. Use of a neuronavigation system, angled endoscopy and an ultrasound aspirator with a bone-specific tip permitted us to achieve adequate and precise decompression.

The patient experienced early neurological benefit, which was also highlighted by intraoperative neurophysiological monitoring, and he was stable at long-term follow-up. Postoperative CT scanning and radiography confirmed the predicted stability of the CVJ, and the patient did not complain of any symptoms of instability or pseudoarthrosis.

Conclusion

Correct neurological and biomechanical analysis of all craniovertebral junction malformations permits stratification and selection of those cases that can be managed by simple, direct, minimally invasive decompression with no need for surgical fusion. In patients with many comorbidities and advanced age, minimally invasive surgery and avoidance of secondary posterior fusion are fundamental to reduce the period of hospitalization and recovery.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Refinement of the Transoral Approach to Craniovertebral Junction Malformations

Paolo Perrini, Nicola Benedetto, Francesco Cacciola, Pasquale Gallina, and Nicola Di Lorenzo

Abstract *Background:* The transoral approach provides the most direct surgical corridor for treatment of congenital bony abnormalities that exert irreducible ventral compression of the cervicomedullary junction. In this paper, based on our experience with the transoral approach over the past three decades, we briefly describe the surgical strategies and the operative nuances that allow effective decompression of the craniovertebral junction (CVJ) while minimizing postoperative morbidity.

Methods: The surgical strategy is dictated by the type and severity of the malformation. Fibre-optic nasotracheal intubation obviates the necessity of preoperative tracheostomy, and avoidance of a soft-palate incision significantly reduces oropharyngeal morbidity. When feasible, the atlas-sparing technique minimizes postoperative instability. The transoral transatlas approach is generally required in patients with severe basilar invagination and allows wider exposure of the anterior CVJ at the price of a higher incidence of postoperative instability.

Conclusion: The transoral approach is extremely effective in providing excellent decompression of the anterior cervicomedullary junction in patients with fixed malformations. Tailoring the approach to the peculiar anatomy of each malformation reduces iatrogenic instability and minimizes postoperative complications.

Keywords Transoral approach · Craniovertebral junction · Odontoidectomy · Basilar invagination

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The transoral approach, originally described by Kanavel in 1917 [1] and subsequently refined by many contributions from different pioneers [2–7], can be regarded today as the standard approach for treatment of selected irreducible anterior malformations that compress the cervicomedullary junction.

The aim of this report is to describe the surgical nuances of the transoral approach that allow the surgeon to achieve effective decompression of the cervicomedullary junction while minimizing postoperative complications in patients with craniovertebral junction (CVJ) malformations.

Surgical Strategy

The factors dictating the surgical strategy in patients with CVJ malformations should be carefully evaluated preoperatively [5–9]. The transoral approach is indicated in selected malformations exerting irreducible ventral compression of the cervicomedullary junction. Accordingly, the reducibility of the malformation should be investigated with spinal radiography and computed tomography (CT) scanning, including dynamic flexion and extension views. The primary treatment of reducible CVJ malformations is posterior fixation and fusion [5]. In addition, the site of encroachment (anterior or posterior) should be clearly evaluated. Patients with limited mandibular excursion (i.e. an interdental space ≤30 mm) or severe basilar invagination (projection of the odontoid tip ≥20 mm above the Chamberlain line) associated with platybasia require adjuncts to the transoral approach (i.e. transmaxillary approaches) or an endoscopic endonasal approach [8, 10–12]. The surgical treatment of tonsillar herniation in the presence of irreducible ventral compression is still a matter of controversy [8, 13, 14]. According to our surgical experience [8], anterior decompression is effective in relieving obstruction of the subarachnoid space at the foramen magnum level in most patients

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with tonsillar herniation associated with fixed CVJ malformation. After generous anterior decompression, ascent of the cerebellar tonsils into the posterior fossa, with acquisition of a more rounded shape, can generally be observed in most patients. Transoral decompression is associated invariably with the risk of creating acute or delayed spinal instability [15, 16]. In our experience, single-stage transoral decompression with posterior fixation and fusion eliminates the risk of postoperative instability and allows early mobilization of patients [8, 9, 16].

Operative Technique

Anaesthetic Considerations and Positioning

Early in our experience, tracheostomy was routinely used in all transoral cases. More recently we moved to fibre-optic nasotracheal intubation, and we consider prophylactic tracheostomy in patients with vagal, hypoglossal and glossopharyngeal nerve dysfunction or in patients requiring an extended maxillotomy approach.

The patient is positioned supine with the head held in a Mayfield headholder and slightly extended. The slight head extension improves the exposure of the rostral aspect of the CVJ and is required in patients with basilar invagination. A moderate Trendelenburg position is used intraoperatively when added cranial exposure is required.

Dedicated self-retaining retractors (Crockard transoral instruments; Codman Raynham, MA, USA) are required to keep the mouth open with the tongue depressed.

To reduce postoperative morbidity we avoid soft-palate splitting and we retract the soft palate with two rubber catheters passing through the nares and stitched to the uvula. Palatal retraction substantially reduces postoperative phonation and swallowing disturbances, which may occur after palatal splitting.

In our experience, lateral fluoroscopy is effective in guiding the extent of surgical exposure in the sagittal plane and in maintaining anatomical orientation. Frameless navigation systems can improve the evaluation of the medial–lateral orientation.

Neurophysiological monitoring is utilized during positioning and throughout surgery, and permits intraoperative assessment of spinal cord function, increasing the safety of the procedure.

Incision and Soft Tissue Dissection

The tubercle on the anterior arch of C1, which is the surgical landmark for the transoral approach, is palpated and identified with lateral fluoroscopy. The posterior pharynx is infiltrated with 1% lidocaine and 1/100,000 epinephrine, and a midline incision is made, centring on the tubercle of C1. The pharyngeal mucosa and the underlying pharyngeal muscles (pharyngeal constrictor, longus colli and longus capitis muscles) are elevated in a single layer and retracted with the two blades of a pharyngeal retractor. The anterior longitudinal ligament is dissected with monopolar cautery, exposing the anterior arch of C1 and the ventral surface of the body of C2. After exposure of the ventral bony structures of the CVJ, different surgical strategies can be utilized depending on the anatomical conformation and the pathology of the patient.

Transoral Atlas-Sparing Technique

In our experience the anterior arch of C1 can be preserved in patients with fixed atlantoaxial dislocations and in selected cases of mild basilar invagination. Biomechanical investigations and clinical reports have documented that even a single break in the continuity of the C1 ring can promote lateral spreading of the lateral masses and caudal migration of the occiput toward C2 [17, 18]. Spreading of C1 and subsequent cranial settling are associated with kinking of the bulbomedullary junction and progressive neurological deterioration [8]. In addition, when the C1 ring is preserved, stability of the CVJ can be achieved with C1–C2 posterior fixation instead of occipitocervical fusion [9, 19].

Using a 3- to 4-mm diamond burr, the base of the dens and approximately 5 mm of the inferior half of the anterior arch of C1 are removed. After transection of the base of the dens, the remaining odontoid fragment is pulled down and anteriorly away from the dura with a toothed odontoid rongeur, and the alar and apical ligaments are divided with curved curettes. At this point the odontoid fragment is removed en bloc, allowing access to the retro-odontoid ligaments (Fig. 1). When the ligamentous structures are difficult to dissect, the dens can be removed piecemeal. However, in these cases, access to the apex of the dens is safer after transection of the atlas.

Transoral Transatlas Technique

In cases of severe basilar invagination, the exposure of the significantly translocated odontoid peg requires transection of the anterior ring of the atlas. The atlas is progressively removed with a high-speed drill and a Kerrison rongeur (Fig. 2). Transection of the atlas allows exposure of the odontoid peg, which is removed piecemeal. In CVJ malformations, ligaments can adhere to the attenuated dura. Accordingly, a meticulous microsurgical technique and judicious dissection are necessary to accomplish effective decompression of the cervicomedullary junction while avoiding unintentional dural tearing. Once the decompression is completed, the wound is

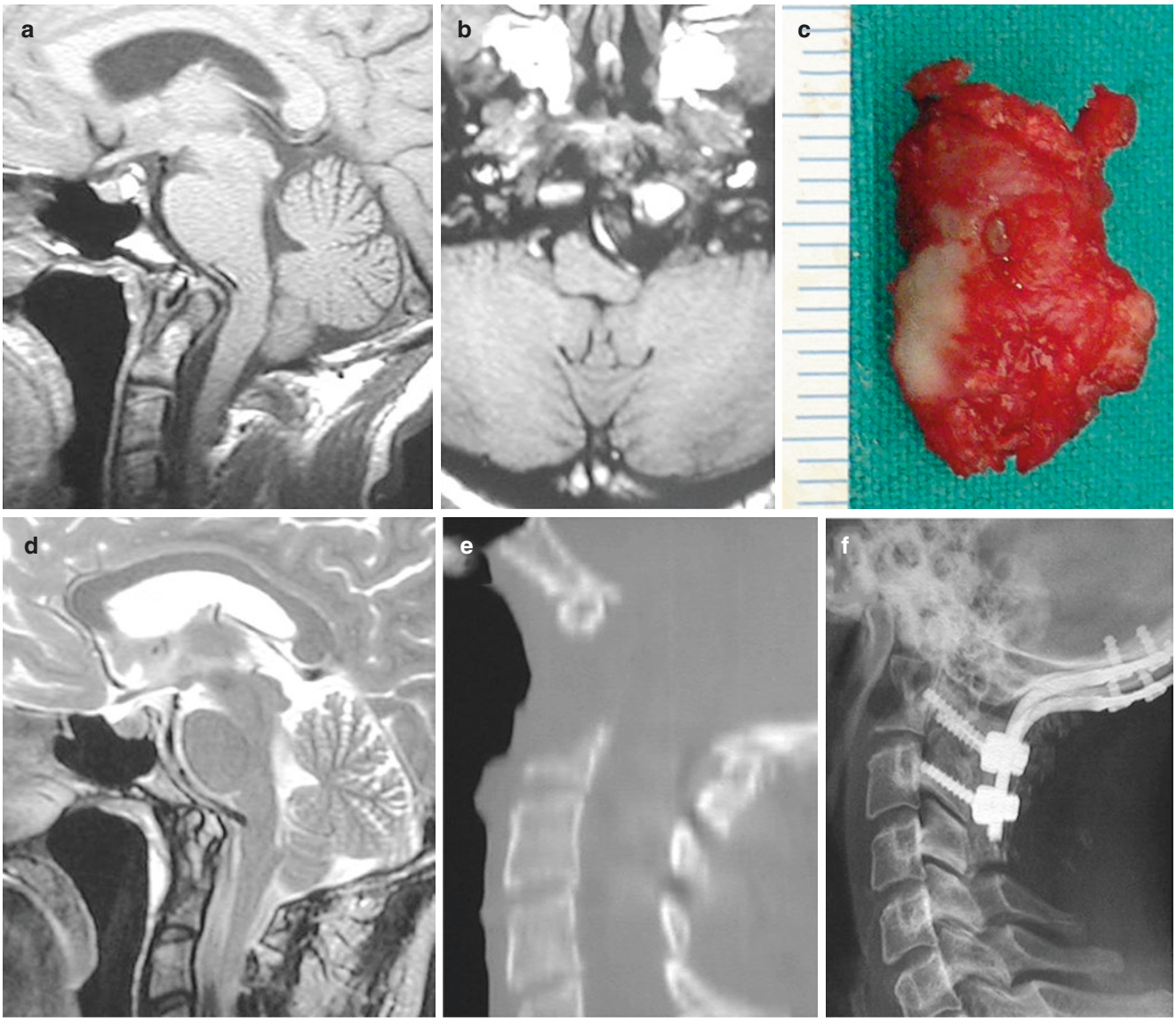

Fig. 1 Craniovertebral junction malformation suitable for an transoral atlas-sparing approach. (**a**) Preoperative sagittal and (**b**) preoperative axial T1-weighted scans demonstrating basilar invagination with severe compression of the ventral cervicomedullary junction. (**c**) After transection of the base of the dens with a high-speed drill, the odontoid process is grasped with a rongeur, and the apical and alar ligaments are sectioned. (**d**) Postoperative sagittal T2-weighted scan showing that brainstem compression is relieved after transoral surgery. (**e**) Postoperative sagittally reformatted computed tomography (CT) scan disclosing removal of the odontoid process with sparing of the atlas. (**f**) Postoperative lateral cervical X-ray demonstrating solid occipitocervical fixation

closed in a single layer with interrupted 2-0 Vicryl sutures [8, 9]. To eliminate the risk of acute postoperative instability and to mobilize patients early after decompression, we simultaneously (i.e. during the same anaesthesia session) perform posterior occipitocervical fixation and fusion.

Complications and Their Avoidance

Careful patient selection and refinement of surgical strategies, together with improvements in intraoperative monitoring techniques, have reduced postoperative complications and improved the outcomes after transoral decompression [7, 8, 19–21]. Contemporary surgical series have reported a perioperative mortality rate of 3–6% and a surgical complication rate of 7–10% following the transoral approach [8, 19]. Surgical complications include velopharyngeal dysfunction, soft tissue swelling, neurological deterioration, dural laceration, cerebrospinal fluid (CSF) leakage and meningitis, vascular injury and wound dehiscence (Table 1).

Velopharyngeal dysfunction is the result of scarring of the pharynx and the soft palate, and it causes hypernasality of the voice, nasal regurgitation and dysphagia [22]. It can be exacerbated in cases of concurrent lower cranial nerve deficits. In our experience the occurrence of velopharyngeal

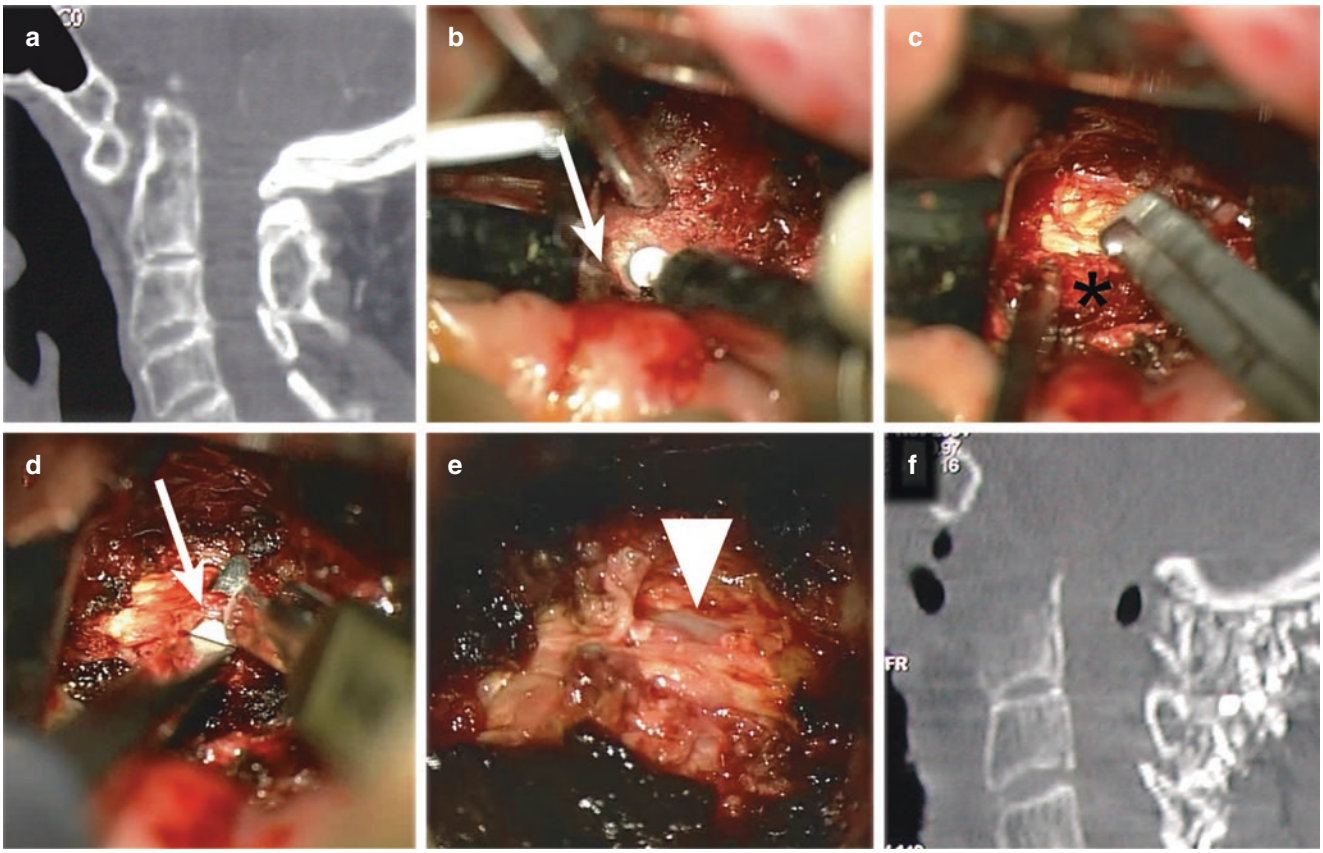

Fig. 2 Craniovertebral junction malformation suitable for a transoral transatlas approach. (**a**) Preoperative sagittally reformatted computed tomography (CT) scan revealing basilar invagination, atlas assimilation and fixed atlantoaxial dislocation. (**b**, **c**) After transection of the anterior ring of the atlas (*arrow*), the invaginated odontoid process (*asterisk*) is removed piecemeal with a Kerrison rongeur. (**d**, **e**) The retrodental ligaments (*arrow*) are incised and the dura of the craniovertebral junction is exposed (*arrowhead*). (**f**) Postoperative sagittally reformatted CT scan demonstrating the extent of C1–odontoid resection. (Acta Neurochir (Wien) 2014;156(6):1231–1236. Reprinted with permission)

Table 1 Summary of surgical complications and their avoidance

Complications	Causes	Avoidance
Velopharyngeal dysfunction	Scarring of the pharynx and soft palate	Soft palate retraction instead of soft palate division
Neurological deterioration	Direct trauma during decompression Uncontrolled movement of the neck during repositioning for posterior fixation	Dissection of the odontoid under high magnification Careful patient repositioning for posterior fixation Inside-out dissection during odontoid removal Intraoperative neurophysiologic monitoring
CSF leak and meningitis	Inadvertent dural laceration Aggressive sharp dissection	No blind dissection Avoid aggressive dissection of retro-dental tissues When cerebrospinal fluid is detected, immediately perform direct repair and insert an external lumbar drain
Injury of the vertebral artery	Direct trauma to V3 segment	Study CT and MRI for rotatory subluxation of C1 and anomalous position of vertebral artery
Pharyngeal wound breakdown	Excessive coagulation of pharyngeal mucosa	Minimize coagulation of pharyngeal mucosa Single layer closure of pharyngeal wall
Postoperative soft tissue swelling	Excessive retraction of soft tissues	Careful tongue retraction Minimize posterior pharyngeal wall retraction

dysfunction is extremely low when soft-palate incision is avoided.

Some degree of soft tissue swelling is common and generally subsides within the first 24–48 h. Careful handling of soft tissues and delicate tongue retraction during surgery minimize postoperative swelling. We maintain the endotracheal tube during the first night after surgery because early extubation can lead to respiratory distress.

Postoperative neurological deterioration is extremely unusual and is directly related to the severity of the preoperative neurological status [8, 19]. Injury of the bulbomedullary junction may be a result of technical errors during odontoid resection, loss of spinal alignment during patient repositioning between anterior decompression and posterior fusion, and a postoperative haematoma. Avoidance of aggressive retro-odontoid soft tissue dissection, along with use of intraoperative neurophysiological monitoring and careful patient repositioning, minimize the risk of postoperative neurological decline.

CSF leakage is a serious complication and originates from dural laceration during dissection of retro-odontoid ligaments. A dural breach should be treated with direct repair. When the dural breach is large and direct watertight repair is not feasible, the dura should be sealed with fat and fibrin glue, with placement of a lumbar drain for up to five postoperative days. If CSF leakage recurs after discontinuation of lumbar drainage, a CSF diversion procedure should be performed.

Injury of the vertebral artery can occur in cases of aberrant arterial anatomy or rotatory subluxation of C1. Careful preoperative evaluation of CT scans and understanding of the three-dimensional vascular anatomy of the CVJ are required to resect the odontoid process while avoiding injury of the vertebral artery.

Pharyngeal wound dehiscence is rare, occurring in 3% of patients after transoral decompression [8]. Single-layer closure and careful handling of soft tissues reduce the occurrence of wound breakdown [8, 21]. A pharyngeal wound should be periodically inspected for the first week after surgery and when dehiscence is detected, wound debridement and closure are required.

Medical complications—including urinary tract infection, pneumonia, deep venous thrombosis, myocardial infarction and pulmonary embolism—are common after the transoral approach, particularly in patients with a poor neurological status [8, 19].

Postoperative Management

After surgery, patients spend the first 24 h in the intensive care unit and are ventilated for the first 12 h. All patients receive low molecular weight heparin as prophylaxis against postoperative venous thromboembolism until they are fully ambulatory. No food is taken by mouth for 3 days, and nutrition is administered intravenously for 3–5 days. Patients initially start to sip cold fluid and usually progress to regular foods in 10 days. They are mobilized, wearing a Philadelphia collar, early in the postoperative period, with the assistance of a physiotherapist. A CT scan is obtained to assess the extent of CVJ decompression and screw placement.

Conclusion

The transoral approach allows successful decompression of the craniovertebral junction in most patients with bony malformations exerting irreducible ventral compression. Tailoring the approach to the peculiar anatomy of each malformation and meticulous exercise of the basic tenets of skull base surgery (the drilling technique and the combination of sharp and blunt dissection) minimize postoperative complications and reduce iatrogenic instability.

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CVJ Instrumentation and Fusion

Occipitocervical Fusion

Angelo Lavano, Giusy Guzzi, Carmelino Angelo Stroscio, Attilio Della Torre, Donatella Gabriele, Domenico Chirchiglia, and Giorgio Volpentesta

Abstract Occipitocervical fusion is a surgical technique in continuous evolution due to the innovation of devices, operative and instrumentation techniques. The aetiologies responsible for occipitocervical instability are trauma, neoplastic disease, metabolic disease or congenital disease. A variety of stabilization techniques are currently available depending on the type of patient and surgeon's experience. Each of these techniques requires thorough knowledge of the anatomy of the craniovertebral junction.

Introduction

The craniovertebral junction (CVJ) consists of a highly specialized anatomical complex, which ensures a wide range of motion. This biomechanical complex is provided with a relatively vulnerable junction synovial system, which should be preserved during surgical interventions. Fusion procedures of the craniovertebral region must be able to withstand compressive force, axial loading, flexion, extension, lateral rotation and lateral bending. Despite the possible alternative of a surgical procedure on the craniovertebral junction, that can promote the occipital bone fusion (as in the Brooks, Gallie and Sonntag techniques), the occipitocervical block remains the most appropriate treatment when the CVJ is unstable or if there are diffuse bone destruction, fracture, or progressive inflammatory or metabolic disease that could lead to postoperative cranial settling after atlantoaxial fixation with wires. According to the criteria of White and Panjabi, the diagnosis of craniocervical junction instability is confirmed if, on dynamic radiography, C2 is moved by ≥3 mm in adults or by ≥5 mm in children, and if there are any neurological symptoms [1].

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Overview of Techniques for Occipitocervical Fixation

Many techniques for occipitocervical fixation have been reported in the literature, using various internal fixation instruments. The technique of occipitocervical fixation was first described by Foerster in 1927, who used a simple onlay bone graft for occipitocervical fusion with fibular grafts [2–4]. It was only in 1967 that Hamblen described his experience of onlay bone grafting with or without wiring [5]. Atlantoaxial arthrodesis involves placement of a bone graft between the dorsal portion of the posterior arch of C1, the spinous process and the C2 laminae. There are two variants of the Gallie technique: the first variant involves a single loop around the arch of C1, ensuring that the bone grafts to the arc of C1 and to the upper laminar surface of C2; and the latter variant involves a double-loop technique [6]. Brooks fusion involves passing a double row of sublaminar wires above the posterior arch of C1 and the C2 lamina, ensuring in this way that the two autologous bone grafts from the iliac crest are embedded on each side between the posterior arch of C1 and the C2 laminae with four sublaminar wires [7]. Methyl methacrylate and pin fixation involves use of acrylic resins, which are polymerized in situ to provide immediate (but not as effective) stability. To adequately stabilize the craniovertebral junction, these resins should be used with pins and wires; they are indicated in patients with a short life expectancy [8]. Halifax interlaminar clamps are positioned on each side above the pedicles and make stabilization via the installation of screws. A bone graft is then placed between the spinous processes to speed up the fusion. This type of clamp provides an intermediate level of stiffness fixation in C1–C2. The mechanical effectiveness of this technique is comparable to those of fixation techniques using modified Brooks wires; they are not as stiff as transarticular screws but are more rigid than the fixation wires used by Gallie [9]. Transarticular screw fixation involves transarticular screw insertion between C2 and C1. A bicortical bone graft is fixed

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by means of wires so as to be embedded between the posterior arc of C1 and the C2 laminae. The wires on the graft facilitate melding and also provide additional stabilization. Use of transarticular screws with claw fixation consists of passing the screws through an adapted plate with an arm containing a pincer, which is fixed to the posterior arc of C1 [10]. The fixation of C1–C2 with polyaxial screws and rods is specially chosen to stabilize C1–C2 in all patients presenting with position abnormalities of the vertebral artery that would preclude passage and use of screws. They are inserted inside the lateral masses of C1–C2 and are then fastened bilaterally by bars [11]. In 2002, Matsumoto described a fixation technique of the craniovertebral junction that used mesh cages positioned between the posterior arc of C1 and the upper laminar surface of C2 bilaterally, in addition to C1–C2 transarticular screws [12].

Discussion

Occipitocervical instability may be caused by trauma, degeneration, neoplastic disease, inflammatory disease or congenital anomalies. The solution to this problem is occipitocervical fixation with bone fusion [2, 3]. Whichever technique is used, the objectives of CVJ fixation are to restore and preserve the anatomical alignment of the spine, to decompress nervous elements, to resist forces along all axes of motion (rotation, translation, lateral bending, flexion, extension and axial loading) and to try to provide the best biological environment to ensure good bone fusion, because failure to meet any one of these objectives makes all techniques ineffective in the long term, compromising the stability of the CVJ. According to recent biochemical studies, the screw-based construct is more rigid than the wire–rod construct [13]. The best location for occipital fixation is the suboccipital midline ridge, just below the external occipital protuberance (EOP), which has a thickness of 11.5–15.1 mm in males and 9–12 mm in females [14]. Mullett observed that the ridge was located on the anatomical midline in just 52% of patients and was deviated to the right or left by 2–5 mm in 28% of patients and by 5–10 mm in 20% of patients [15]. The complications that may arise with skull penetration if the midline suboccipital bone is not thick enough are injuries of the dural sinus and epidural haemorrhage of the posterior fossa [16]. There is the potential for vertebral artery or cervical root injury to occur with a screw-based approach because of the cervical screw purchase [17].

Conclusion

Instrumentation and fusion procedures are particularly effective in metabolic conditions, inflammatory diseases or neoplastic diseases of the spinal cord with involvement of the craniovertebral junction. The benefits of an approach using wires include simplicity, safety and efficacy. The use of titanium bars and/or more compact and resistant occipital wires is not recommended when metal screws are used, to better ensure stability of the scaffold. Preoperative radiological reducibility or irreducibility in an awake patient is not predictive of the operating reducibility under general anaesthesia. There is no ‘gold standard’ surgical technique for occipitocervical fixation. The choice of technique, with its advantages and disadvantages, is based on the type of instability, the integrity of the posterior cervical elements, the extension of decompression, comorbidities, individual anatomical variations and the surgeon’s practical experience.

Competing Interests The authors declare that they have no competing interests.

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Occipitocervical Fusion: An Updated Review

Nabeel S. Ashafai, Massimiliano Visocchi, and Norbert Wąsik

Abstract Occipitocervical fusion (OCF) is indicated for instability at the craniocervical junction (CCJ). Numerous surgical techniques, which evolved over 90 years, as well as unique anatomic and kinematic relationships of this region present a challenge to the neurosurgeon. The current standard involves internal rigid fixation by polyaxial screws in cervical spine, contoured rods and occipital plate. Such approach precludes the need of postoperative external stabilization, lesser number of involved spinal segments, and provides 95–100% fusion rates. New surgical techniques such as occipital condyle screw or transarticular occipito-condylar screws address limitations of occipital fixation such as variable lateral occipital bone thickness and dural sinus anatomy. As the C0–C1–C2 complex is the most mobile portion of the cervical spine (40% of flexion-extension, 60% of rotation and 10% of lateral bending) stabilization leads to substantial reduction of neck movements. Preoperative assessment of vertebral artery anatomical variations and feasibility of screw insertion as well as visualization with intraoperative fluoroscopy are necessary. Placement of structural and supplemental bone graft around the decorticated bony elements is an essential step of every OCF procedure as the ultimate goal of stabilization with implants is to provide immobilization until bony fusion can develop.

Keywords Occipitocervical fusion · Occipitocervical fixation · Surgical techniques

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Anatomy and Biomechanics

The occiput–C1–C2 complex is the most mobile portion of the cervical spine. The occiput–C1 motion segment makes the largest contribution to flexion (21°) and extension (3.5°), while the primary movement of the C1–C2 motion segment is axial rotation (23.3–38.8° per side). Patients need to be informed before occipitocervical fusion (OCF) about the substantial restrictions it will cause in the neck's range of motion (40% of total cervical flexion–extension, 60% of total cervical rotation and 10% of total cervical lateral bending if the occiput–C1 and C2 are involved) [1]. The occipital bone and the vertebral artery are key structures, which demand meticulous preoperative assessment. The variable thickness of the squamous part of the occipital bone determines the lengths of the screws. The external occipital protuberance is the thickest part of the occiput (9.7–15.8 mm), and the thickness decreases in a radial fashion laterally and inferiorly (Fig. 1) [2]. The occipital bone is built up of two layers of cortical compact bone, and located in between them is spongy diploe. Because of the marginal contribution of the inner cortical layer to the overall occipital thickness, unicortical screws provide fixation strength comparable to that of bicortical screws of the same length, but only in the area near the external occipital protuberance [3]. Aberrant vertebral artery loop recognition on preoperative imaging studies is also critical. Use of some types of cervical screw might not be feasible, because of a high-riding vertebral artery or narrow C2 pedicles [4].

Indications

The main indication for OCF is instability of the craniocervical junction (CCJ). The most common causes are listed in Table 1. Posterior internal stabilization prevents compres-

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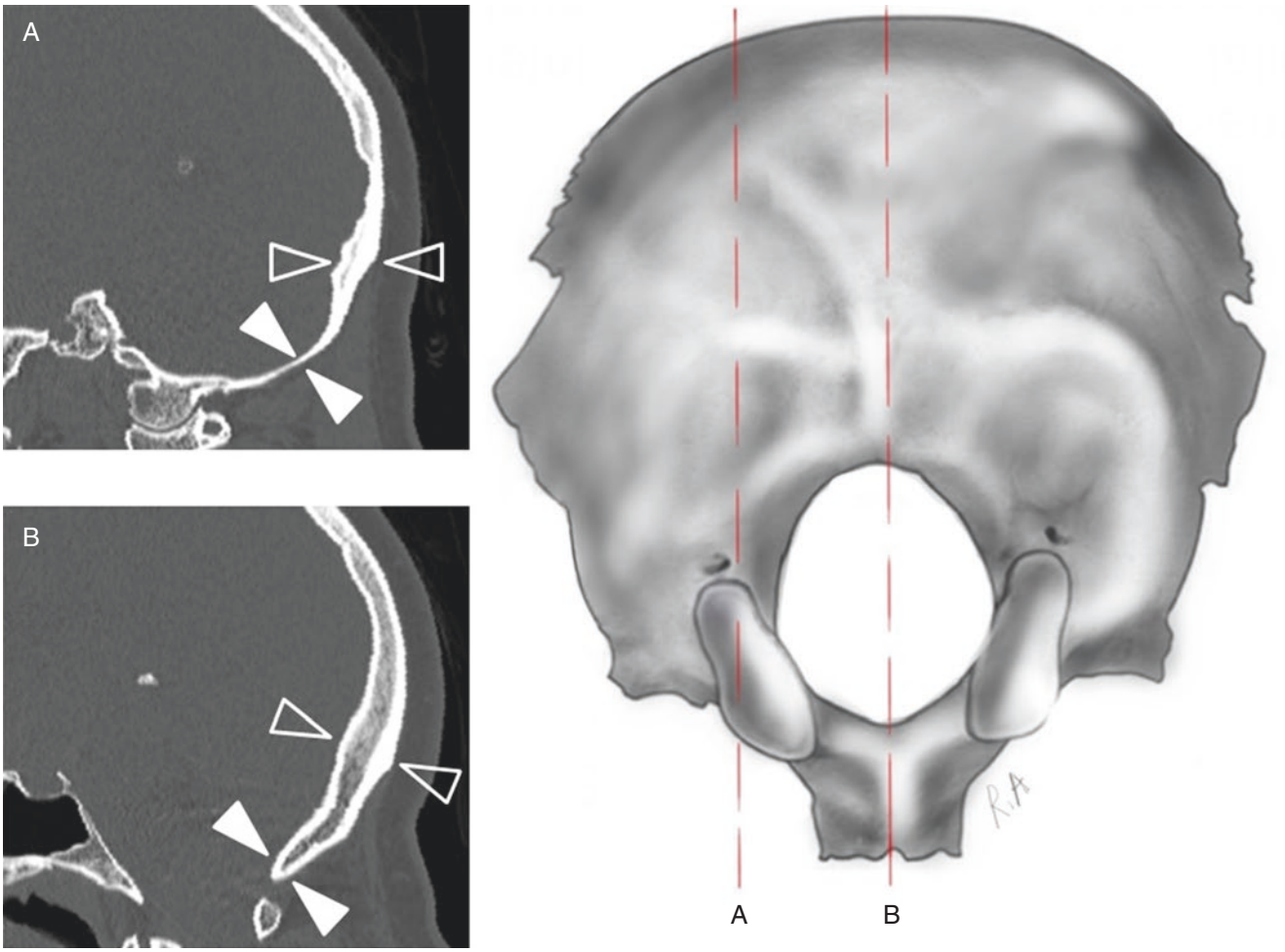

Fig. 1 Occipital bone thickness. The maximum thickness of the occipital bone on sagittal sections A and B is 8.3 mm and 13.8 mm respectively (blank triangles). The minimum thickness of the occipital bone on sagittal sections A and B is 2.1 mm and 5.7 mm respectively (filled triangles). The left side is usually 1 mm thinner than the right side

sion of the neural structures, enables correction of cervical deformity and reduces pain (Fig. 2).

Surgical Technique

Patients who qualify for OCF have their head and cervical spine stabilized by an external fixation device (a halo device, Gardner-Wells tongs or a hard collar), which must be maintained until rigid head fixation to the surgical bed is used—for example, a Mayfield skull clamp.

Remember that cervical traction is contraindicated in the case of an CCJ dislocation or a significant ligamentous injury on magnetic resonance imaging [5]. In the initial supine position, fibre-optic intubation is performed, an arterial line is placed and baseline somatosensory evoked potentials (SSEPs) and motor evoked potentials (MEPs) are obtained if available. Anaesthetic agents interfering with SSEPs (nitrous oxide) and MEPs (long-acting paralytic agents) must be avoided [6]. The patient is rotated to the final prone position on chest rolls or a Jackson frame, and the SSEPs and MEPs are checked immediately. Persistent changes in SSEPs or MEPs mandate returning the patient to the supine position and undertaking an awake

Table 1 Causes of craniocervical junction instability requiring occipitocervical fusion

Cause of instability	Example
Trauma	Atlanto-occipital dislocation, occipital condyle fracture, atlas and axis fractures
Inflammation/autoimmune disease	Rheumatoid arthritis, psoriatic arthritis, ankylosing spondylitis, inflammatory bowel disease–associated arthropathy, tuberculosis, osteomyelitis
Neoplasm	Metastasis, chordoma, Ewing tumour, osteoblastoma, osteochondroma, haemangioma, aneurysmal bone cyst
Congenital	Chiari malformation with basilar invagination, Down's syndrome, Klippel–Feil syndrome, Morquio's syndrome, os odontoideum
Iatrogenic	After odontoidectomy, failed previous attempts at C1–C2 fusion, after the far lateral approach with occipital condyle resection

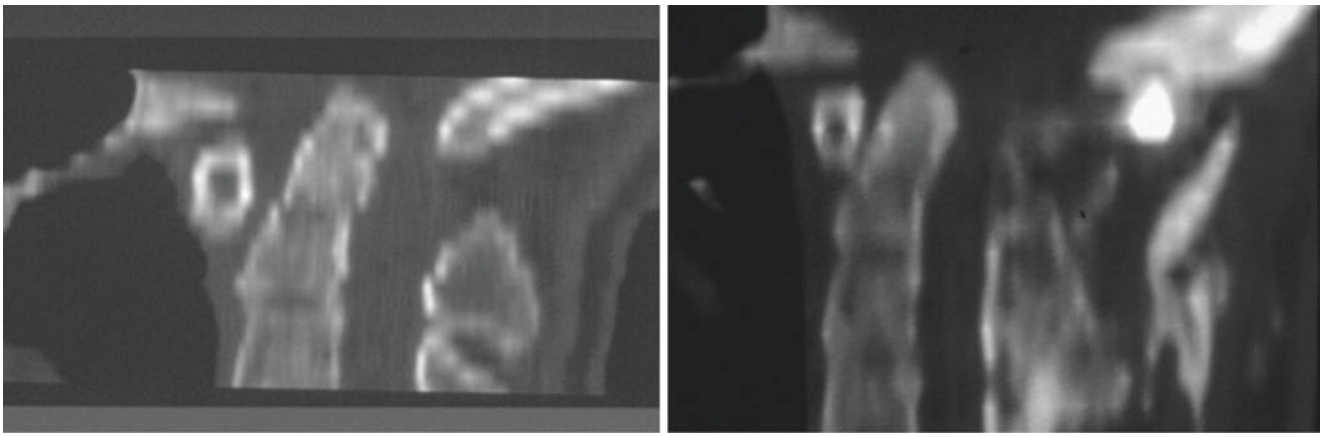

Fig. 2 Basilar invagination before (*left*) and after (*right*) occipitocervical fusion in light extension with 'functional decompression' of the craniocervical junction

neurological examination. The proper alignment of the head in a neutral position or slight flexion is confirmed by fluoroscopy. The bed is placed in a reverse Trendelenburg position to decrease venous bleeding. The midline skin incision should extend from the inion to the spinal segment below the last segment that will be incorporated into the construct. Sharp dissection is used to split the paraspinal muscles via the midline avascular plane, then subperiosteal dissection of the occiput and the posterior spine elements is performed.

If necessary, decompression of the neural elements is performed. The surgeon retains discretion as to whether the occipital or cervical part of the construct should be placed first. The occipital plate is placed in position and its apertures are marked. As bicortical purchase is desirable (but not essential) and plunging through the bone is possible, a manual drill with a drill guide is used. The depth is set on the basis of the neuronavigation indications. The holes are tapped and the blunt occipital screw is placed to secure the plate to the occiput. In the event of cerebrospinal fluid leakage or dural sinus violation, a screw should be inserted immediately. Our preferences for cervical fixation are C1 lateral mass screws and C2 pars or pedicle screws [7]. Care is taken not to injure the vertebral artery while exposing the posterior ring and lateral masses of C1. The lateral dissection should not exceed 1.5 cm from the midline. The screw heads and occipital plate are connected by a 3.5 mm or 4 mm rod. Contouring and notching of the rod (involuntary cutting of an indentation on the surface, i.e. during nut locking) decreases the fatigue life of the construct [8]. To minimize the risk of rod weakening, pre-bent or hinged rods have been introduced. A flexible endotracheal tube stylet may be used as a template before the rod is bent. Lateral offset connectors are useful in the event of difficulties in connecting the laterally directed lateral mass screws and the medially directed pedicle screws with the rod [9]. Extension of the construct caudally beyond the C2 level is indicated when an irreducible lesion is treated, posterior C1/C2 elements are damaged or basilar invagination is present [10]. Visocchi et al. first described the possibility of achieving 'functional decompression' of basilar invagination by performing OCF in light extension, thereby avoiding the more challenging transmucosal anterior transnasal and/or transoral decompression (Visocchi M, Di Rocco F, Meglio M: Craniocervical junction instability: instrumentation and fusion with titanium rods and sublaminar wires. Effectiveness and failures in personal experience. Acta Neurochir (Wien Austria) (2003) 145:265–272).

Types of Occipitocervical Fusion

Occipitocervical fusion has advanced significantly since 1927, when Foerster was the first surgeon to reported successful stabilization of the CCJ by in situ onlay application of a fibular strut graft [11]. Currently, screw-based constructs are the most popular option (Fig. 3). The advantages are superior rigidity of fixation, resistance to fatigue and nearly a 100% rate of fusion, while incorporating in the construct fewer motion segments than any wire/cable fixation [12, 13]. The first constructs of this generation (lateral mass plates, Y-shaped plates, Ohio Medical Instruments loops) have predesigned slots, which limit screw entry points and provide non-rigid screw fixation. Those issues have been addressed by the later screw-based constructs, which are currently most popular with neurosurgeons. Occipital plate/eyelet connectors and cervical screws (C1 lateral mass, C2 pedicle/pars/laminar, C1–C2 transarticular, C3–C6 lateral mass) are placed independently and linked by the two 3.5- to 4.0-mm rods (bent during surgery, pre-bent or hinged). Cervical rods with an integrated occipital plate end provide more lateral fixation (shorter screws) but allow placement of more screws than a midline plate. In addition to the consequence of significantly less motion in lateral bending with lateral fixation, the surgeon's personal preference may determine the choice

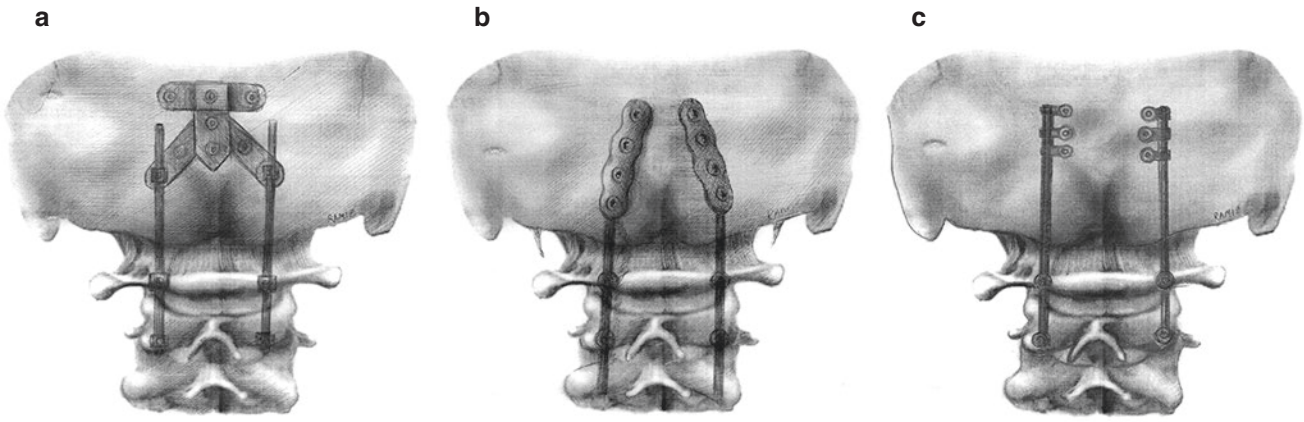

Fig. 3 The most common screw-based constructs. (**a**) Occipital plate. (**b**) Hinged rods with an integrated occipital plate end. (**c**) Eyelet connectors directed medially

between lateral or midline occipital fixation [14]. Occipital fixation may be also achieved by inside–outside screws. The occipital bone is secured between an inside button (inserted through an adjacent burr hole) and an outside nut. The large surface of both buttons provides a satisfactory area of occipital bone fixation in cases of osteoporosis or a thin occipital bone [15, 16]. The most recently introduced surgical techniques are occipital condylar screws and transarticular occipitocondylar screws [17, 18]. They enable avoidance of occipital screw complications (e.g. dural sinus perforation). These new techniques may be used in patients after large suboccipital craniectomies. The first case series, as well as biomechanical studies, have proved that OCF using occipital condylar screws is a feasible alternative to the current occipital plate fixation [19, 20].

Arthrodesis

Iliac crest and rib autografts remain the gold standard for use in posterior cervical procedures. Although both sites are considered safe for graft harvest, the use of rib grafts provides higher fusion rates and less morbidity for patients [21]. Long bone grafts are secured with cables fastened to the rod construct. If decompression has been performed, acquired bone should be marsupialized and used as an autograft. At the same time, care must be taken to avoid graft placement into the defect and on the dura [7]. Bone allografts are frequently used in combination with autografts (e.g. in 69% of patients in a case series reported by Nockels et al. [13]). Use of bone morphogenetic protein (BMP)-2 is controversial, as the reported complication rate is high (10.3% in a paediatric population; no study in adults has been published) [22]. Nevertheless, some authors advocate it in cases of severe osteoporosis or reoperation in chronic smokers [23]. The bony surface of the occiput, and the posterior elements of the cervical spine (including the facet joints) should be decorticated to bleeding cancellous bone with a high-speed drill prior to graft placement.

Postoperative Immobilization

The use and duration of external immobilization depends strongly on the OCF technique used and the neurosurgeon's personal experience. No external stabilization is used in cases of a technically satisfactory screw and rod OCF procedure in patients with good bone quality [24]. Alternatively, a soft cervical collar, a Philadelphia collar or a Miami J collar may be maintained for 6–12 weeks after OCF with the same technique [25]. If the patient's bone quality is poor, immobilization in a halo ring is required. If a non-rigid OCF method has been used, halo immobilization or use of a Minerva vest for at least 12 weeks is mandatory [26].

Complications

The general complication rate varies from 10% to 33% [25, 27]. Minor complications that are encountered are wound infection or dehiscence, dural tearing and cerebrospinal fluid leakage. OCF-specific complications concern proper head alignment. Excessive flexion results in the patient having an impaired line of sight and swallowing difficulties. Fixation in exaggerated extension results in poor visualization of the ground. Potential complications of major significance include meningitis, posterior fossa haematoma, and direct injury to neural structures and the vertebral artery by misplaced screws. Concerning hardware failure, although wiring

techniques are old fashioned, they still deserve consideration, mainly in redo craniovertebral junction surgery after screwing technique failure. Nevertheless, also in the wiring technique, which is a safe and effective procedure, care must be exercised in the preparation of the cranial holes to avoid sliding complications with the U-shaped rods, as has occurred in our experience. To understand and analyse this event, it is important to underline that the distance between the two burr holes in the occipital bone needs to be shorter than the distance between the ends of the U-shaped rod to secure and preventing caudocranial sliding of the hardware and pulling out of the construct (Fig. 4) [28].

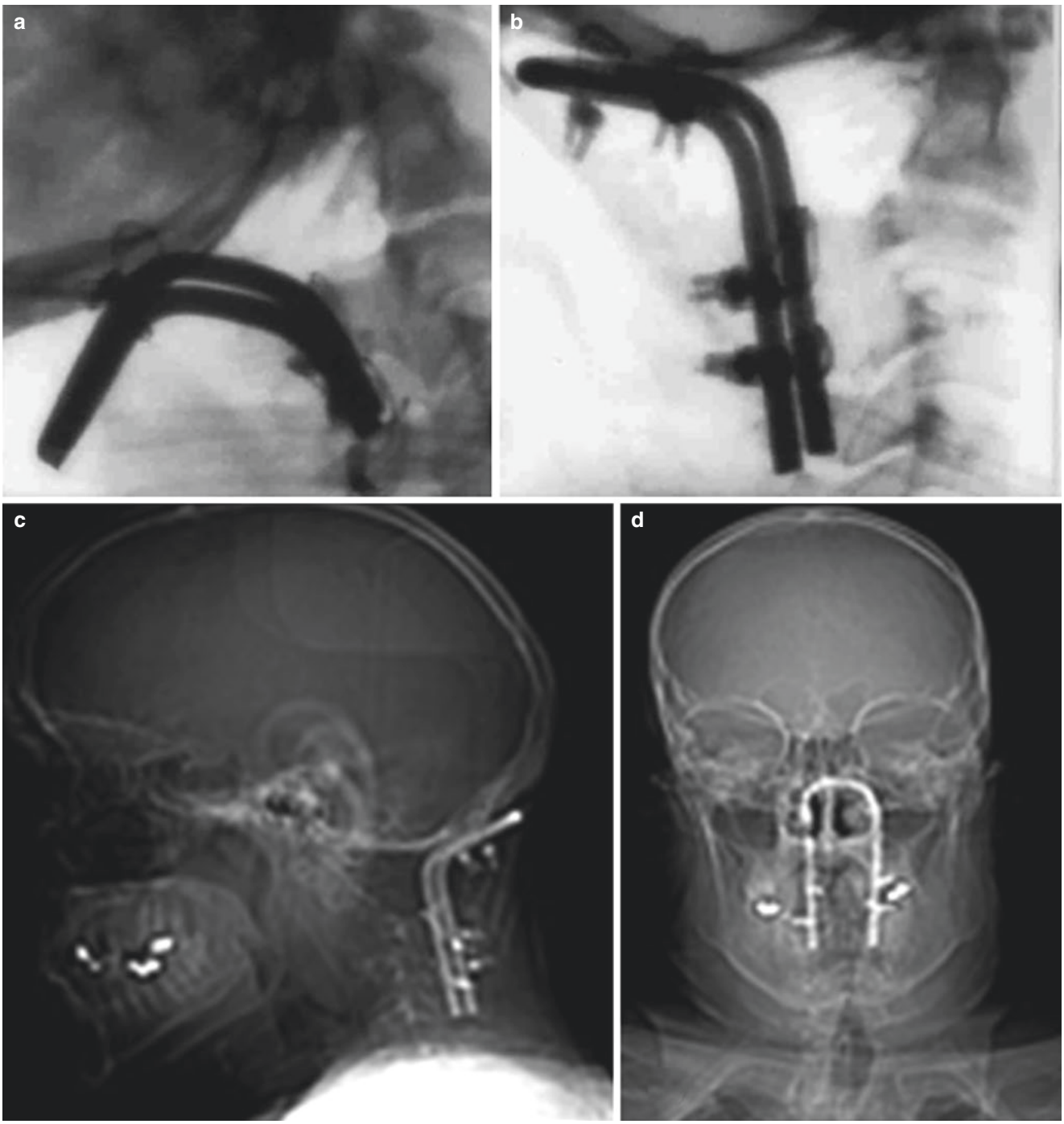

Fig. 4 Early postoperative lateral X-rays demonstrating sliding of the titanium construct associated with pulling out of the hardware. (**a**). Postoperative X-rays (CT scout view). (**b**–**d**) Control X-rays after reoperation

Competing Interests The authors declare that they have no competing interests.

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Wiring or Screwing at the Craniovertebral Junction in Childhood: Past and Present Personal Experience

Massimiliano Visocchi, Francesco Signorelli, Alessandro Olivi, and Massimo Caldarelli

Abstract *Background:* Craniovertebral junction (CVJ) instrumentation and fusion in childhood are frequently performed with either sublaminar wires or screws in lateral masses, and both are considered quite safe procedures.

Methods: Our experience deals with 12 children: six (mean age 9.5 years) harbouring a congenital instability associated with Down's or Morquio's syndromes and primary os odontoideum; and six (mean age 11.5 years) with acquired iatrogenic instability due to transoral anterior decompression for different reasons (inferior clivectomy, anterior arch removal and odontoidectomy). All patients in the 'congenital group', except for one, had preoperative dynamic x-rays and underwent surgical correction by means of posterior wiring, fusion and an external orthosis. All patients in the 'iatrogenic group' had no preoperative dynamic x-rays and underwent a screwing technique with fusion and an external orthosis.

Results: The postoperative clinical picture had improved in all patients at the latest follow-up (observation range 63–202 months [mean 118.5 months]), with neuroradiological confirmation of satisfactory bony fusion and with neural decompression in all patients.

Conclusion: Although it requires a more accurate preoperative neuroradiological setting, the screwing technique takes less time and is characterized by less blood loss and less postoperative discomfort than the wiring technique. The latter features confirm the simplicity, safety (continuous fluoroscopic assistance is not necessary, and there is no risk of neurovascular injuries) and lower expense (neither complex hardware devices nor neuronavigation systems are required) of the screwing technique.

Keywords Craniocervical junction instability · Sublaminar wiring · Screwing technique · Mucopolysaccaridosis · Down's syndrome · Os odontoideum · Transoral approach

Introduction

Anterior transnasal or transoral decompression—which is used in treatment of irreducible neoplastic, dysembryogenetic, inflammatory and chronic traumatic diseases of the anterior craniovertebral junction (CVJ)—has been reported in the literature for many years [1–4].

Surgical treatment of CVJ compression by the transoral or transnasal route is still strongly suggested in cases of irreducible dislocation. Surgical management of CVJ compression aims to achieve neural decompression and to stabilize the CVJ in order to relieve neurological manifestations arising from bulbospinal compression both at rest and during motion, secondary to CVJ instability [5]. *Functional decompression* is a concept in our therapeutic strategy, aiming to achieve neural decompression by performing simple reduction, instrumentation and fusion of the CVJ dislocation when it is reducible [6]. In cases in which accurate preoperative x-ray examinations demonstrate CVJ irreducibility and associated neural compression, the goal of surgery is to maintain anatomical alignment while preserving the motion of normal adjacent elements, with the aim of protecting the neural elements [7, 8]. In this paper we present an update of our personal experience of instrumentation and fusion in children, using titanium rods, sublaminar wires and screws [6].

Materials and Methods

From 1998 to 2018, 12 children were operated on in the Section of Paediatric Neurosurgery at Policlinic Gemelli, Catholic University School of Medicine, in Rome. Six female patients aged 6–14 years (mean age 9.5 years) were treated for os odontoideum (group 1). Five of these patients

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were affected by Down's syndrome, one had a metabolic disease (mucopolysaccharidosis type IV, i.e. Morquio's syndrome) and one had an isolated os odontoideum. A second group (group 2), consisting of six male patients aged 8–15 years (mean age 11.5 years), underwent transoral anterior decompression and staged posterior instrumentation and fusion with screws for different diseases. One of these patients had impressio basilaris, one had basilar invagination, two had os odontoideum, one had a C0–C1 developmental anomaly and one had a C2 fracture and dislocation. All patients underwent computed tomography (CT) scans and static and dynamic magnetic resonance imaging (MRI) of the brain and CVJ. Further preoperative static and dynamic x-rays were performed in patients in group 1, and CVJ instability was shown by atlantoaxial displacement greater than 4.5 mm in all but one patient (Table 1) [9]. One group 1 patient (patient #3) with a hyperintense signal at the medulla and at the bulbospinal junction had gait disturbances and dyspnoea, which led to an emergency tracheostomy [10]. In group 1, the CVJ shift was reducible in five patients and irreducible in one patient (patient #4). Preoperative fixation was accomplished by use of a hard collar.

Surgical Techniques: Posterior Instrumentation and Fusion

Group 1

Patients were placed in a prone position. Intraoperative traction and reduction of the C2 shift were obtained using a Mayfield headholder under fluoroscopic control. After preparation of the occiput and the cervical spine, occipitocervical instrumentation was carried out. Two burr holes into the occiput, 3 cm cranially to the rim of the foramen magnum, represented the proximal point for passing titanium wires. To facilitate the passage of the wires, notching (with Kerrison rongeurs) of the rim of the foramen magnum and of the cervical laminae to be fused, and removal of the atlanto-occipital membrane and ligamentum flavum, were carried out. In patients with C0–C1 assimilation (patients #1, #2 and #3), C1 laminectomy was performed. A wide-diameter (5 mm) nonthreaded titanium rod was bent into a U shape, cut in a way that the ends extended a few millimetres beyond the fused segments to prevent them from slipping out during flexion and extension movements of the neck, and to adapt them to the bony contours of the CVJ. The assimilated and bifid posterior arch of the atlas was excised during posterior decompression of the posterior foramen magnum margin prior to passage of sublaminar wires. With use of the Sonntag method, Songer titanium wires were passed under the involved bone segments and over the titanium rod and bone graft, being stretched up to approximately 10 pounds (Table 1).

Group 2

After transoral decompression a second staged procedure was performed. Under fluoroscopy, C0–C2–C3 screws 3.5 mm in diameter and 12 mm in length (Vertex System [Medtronic, Minneapolis, MN, USA]; Summit SI OCT Spinal Fixation System and Mountaineer OCT Spinal System [DePuy Synthes Spine, Warsaw, IN, USA]; and VuePoint OCT [NuVasive, San Diego, CA, USA]) were inserted in the C2 isthmus bilaterally in the centre of the lateral masses (taking care to spare the vertebral notch) and in the C3 lateral masses from medial to lateral and from caudal to cranial.

Both Groups

Bone fusion was performed by decortication of the occiput and the posterior arches of the cervical facet joints by a high-speed drill and curettes to facilitate bone fusion.

Autologous bone was harvested from the right posterior iliac crest, cut in a double-wing shape and fixed over the construct, using a silk suture along with antigen-free synthetic bone graft substitute fusion (beta-tricalcium phosphate [Vitoss Synthetic Cancellous Bone Void Filler; Stryker, Kalamazoo, MI, USA]). Moreover, cancellous bone was placed upon the levels to be fused when available after further posterior CVJ decompression.

Postoperative Care (Table 1)

Group 1

After completion of the surgical treatment, a halo or SOMI vest was utilized for 4 months in all patients except for patient #3, for whom it was necessary to prolong the application of the external orthosis.

Group 2

After completion of the surgical treatment, a halo or SOMI vest was utilized for no more than 3 months in all patients.

Bone fusion was evaluated on CVJ radiological studies and bone window CT scan examinations. Radiological and CT scans plus MRI and neurological examinations were performed 1 week after surgery, then every 4 months up to 1 year and finally at the last follow-up [11]. The Frankel scale and the Di Lorenzo disability grade were used to evaluate the neurological condition.

Table 36.1 Patients with *congenital* (1–6, Group 1) and *iatrogenic* (7–12, Group 2) instability

Pt initials Case N°.	Age (sex)	Primary disease	Radiology	Pre-op C1C2 shift (X-rays)	Treatment	Post-op shift (X-rays)	Frankel scale and Di Lorenzo Grade changes	External orthosis (months)	Follow-up (months)
CV 1	8 (F)	MPS (Morquio)	– C0-C1 assimilation – C1-C2 instability – C2-C3 partial fusion – Os odontoideum reducible	>4.5 mm	– Suboccipital craniectomy and C1 laminectomy – C0–C3 reduction, instrumentation and fusion	No	E/E I/I	SOMI (4 months)	202
BS 2	6 (F)	Down syndrome	– C0-C1 assimilation – C1-C2 instability – Os odontoideum, reducible	5 mm	– Suboccipital craniectomy and C1 laminectomy – C0–C3 reduction, instrumentation and fusion	No	E/E I/I	Halovest (1 month) SOMI (3 months)	175
TP 3	8 (F)	Down syndrome	– C0-C1 assimilation – C1-C2 instability – Os odontoideum, reducible	10 mm	– Suboccipital craniectomy and C1 laminectomy – C0–C3 reduction, instrumentation and fusion	No	D/E III/I	SOMI (4 months) Halovest (4 months)	172
EAK 4	(7) (F)	Down syndrome	– C1 bifid – C1–C2 instability, – Os odontoideum irreducible	>8 mm	– C0–C3 reduction, instrumentation and fusion;	No	E/E I/I	SOMI (4 months)	143
FI 5	14 (F)	Down syndrome	– C1–C2 instability – Os odontoideum reducible	>4.5 mm	– C0–C3 reduction, instrumentation and fusion	No	E/E I/I	Halovest (4 months)	130
RRS 6	14 (F)	Down syndrome	– C1–C2 instability, – Os odontoideum reducible	>4.5 mm	– C0–C3 reduction, instrumentation and fusion	No	E/E I/I	Halovest (4 months)	124
CL 7	15 (M)	Developmental anomaly C0–Cl	– C0–C1 anterior compression – C1–C2 instability	>5 mm	1. Transoral decompression 2. CI laminectomy, C0 double vertical screws, C2 pedicles and C3 lateral masses screws instrument.	No	D/E II/I	Soft collar (1 month)	74
EA 8	11 (M)	Impressio basilaris Os odontoideum (Down s.)	C1–C2 anterior compression	Virtual, previously documented	Transoral C1 odontoidectomy and clivectomy in C0–C2–C3 screwing instrumentation and heterologous bone fusion (previously implanted)	No	D/E II/I	Soft collar (1 month)	70
RR 9	14 (M)	C2 fracture and dislocation	C2 fracture and C1–C2 dislocation with cervico medullary contusion	>7 mm	1. Transoral C1–C2 decompression 2. C0–C3 C5 screwing instrumentation and heterologous bone fusion	No	D/E II/I	Soft collar (1 month)	67
MP 10	8 (M)	Impressio basilaris	C0–C1–C2 compression	No	1. Transoral C1–C2 decompression 2. Staged C0–C2–C3 screwing instrumentation and fusion	No	D/E II/I	Halo + Philadelphia (2 months)	63
SS 11	9 (M)	Os odontoideum	C2 compression	Virtual	1. Transoral C1–C2 decompression 2. C0–C2–C3 screwing instrumentation and fusion	No	D/E II/I	Halo + Philadelphia (2 months)	118
AE 12	12 (M)	Basilar invagination	C2 compression	Virtual	1. Transoral C0–C1–C2 decompression 2. C0–C2–C3 screwing instrumentation and fusion	No	D/E II/I	Halo + Philadelphia (3 months)	84

Results

The follow-up period ranged from 63 to 202 months (mean 118.5 months). All patients improved soon after surgery independently of the type of surgery they underwent, but an immediate clinical improvement in gait disturbance occurred in patient #3 in group 1, who had Down's syndrome; her Frankel grade changed from D to E and her Di Lorenzo grade changed from III to I. In this patient the improvement in respiratory dysfunction allowed closure of the tracheostomy 24 months after surgery. Nuchal pain disappeared in all of the children postoperatively.

No arterial injuries, bleeding, haematomas or systemic complications occurred. At the site of bone harvesting, no infections, cosmetic problems, pain or complaints were reported.

Blood loss ranged from 17 to 23 mL (mean 20 mL) in group 1 and from 11 to 19 mL (mean 15 mL) in group 2.

Concerning the duration of the posterior approach procedures, it ranged from 3.2 to 4.0 h for wiring and from 2.3 to 3 h for screwing.

Diagnostic imaging, immediately after surgery, showed restoration of bone alignment with decompression of the brainstem in all patients. Neuroradiological signs of bone fusion were already evident 4 months after surgery in all but two patients.

Bone Fusion

Group 1

In patient #3, failure of bone fusion occurred 9 months after surgery, as a consequence of a cerebrospinal fluid (CSF) fistula and wound infection; further bone grafting from the iliac crest was successful after resolution of the CSF leakage. A significant reduction in the cervicomedullary junction contusion was evident at late follow-up.

Group 2

In patient #10, the hardware was revised and the synthetic bone graft substitute was removed 2 weeks after the staged instrumentation and fusion procedure, because of infective dehiscence of the surgical wound. After 2 months of polyantibiotic therapy (intravenous daptomycin 350 mg/day and oral rifampicin 600 mg/day), and 1 month after collar removal, a dynamic cervical MRI examination confirmed CVJ stability.

In both groups, despite the cranial fixation, no limitations in social life due to impaired head motion were observed in any patient.

Discussion

Sublaminar Instrumented Wiring Versus Lateral Mass Screw Implants

Wiring Technique

Sublaminar instrumented wiring remains an excellent and simple procedure for stabilizing the CVJ and upper cervical spine, resulting in a reasonably good mechanical outcome with a low incidence of complications [22]. The stiffness provided by the wiring, determined by the number of vertebrae enclosed by the instrumentation and augmented with external immobilization, is associated with bone fusion in 100% of cases [6, 22, 23]. This observation may help to overcome the early biomechanical drawbacks of sublaminar instrumented wiring with respect to lateral mass screw implants.

Screwing Technique

After early reports on small series of patients treated with this approach, several clinical studies reported that the results obtained with use of lateral mass screw implants were better than those obtained with sublaminar instrumented wiring [12–18]. However, complications reported at the very beginning of the experience (such as 30% of screws pulling out in the suboccipital area and a mortality rate of up to 9% after complex spine decompression and fixation) discouraged some paediatric spine surgeons from using lateral mass screw implants [14, 19]. Lateral mass screw implants in a paediatric population achieved bone fusion in 100% of cases, with a 10.4% complication rate, including vertebral artery injuries [7, 9, 20, 21].

Wiring Versus Screwing

More recently published experiences have seemed to report the same 100% incidence of fusion with both lateral mass screw implants and sublaminar instrumented wiring [6, 8, 14, 22, 23]. Despite a clear advantage of the screwing technique in terms of blood loss, surgery duration and postoperative immobilization, the infectious complication rate appears comparable.

Complications in Our Series

Wiring Technique

The difficulties encountered in patient #3, who had Down's syndrome, were ascribed to the patient's immunocompromised state (impaired monocyte and neutrophil chemotaxis, decreased phagocytosis and qualitative T-lymphocyte defi-

ciency), which may have predisposed her to respiratory infections and postoperative complications. In similar cases, the rate of bone fusion may also be lower than that in other patients, probably as a result of deficient collagen synthesis, which contributes to bone graft pseudoarthrosis [24]. In accordance with the literature, to prevent frequent neck movements in the postoperative period—especially in children with delayed mental milestones and those with spasticity—halo immobilization was instituted early and until there was objective evidence of bony fusion, which could take as long as 6 months [5, 25].

Screwing Technique

We can hypothesize that a long-lasting infection, which occurred in one of our patients, played a role in the ossification process implied in CVJ fusion, since the ossification occurred 33 months after the onset of the infection. Finally (and very surprisingly), in our case the postinfective bone fusion not only produced good fixation but also resulted in a kind of odontoid regeneration that has not been reported so far. In fact, although our group has recently published a description of 'true' odontoid process regeneration (along with clival regeneration and recurrence of Chiari malformation) after transoral decompression, in the present case we observed union of the remaining C2 and C3 bodies, strongly mimicking concomitant, rather complete axis and clival regeneration [26, 27].

Conclusion

The wiring technique is simple, safe (continuous fluoroscopic assistance is not necessary and there is no risk of neurovascular injuries) and less expensive than the screwing technique, as no complex hardware devices and no neuronavigation systems are required.

The screwing technique requires a more accurate preoperative neuroradiological setting than the wiring technique but seems faster and is characterized by less blood loss and less postoperative discomfort.

Caution is needed to avoid postoperative complications (namely, a cerebrospinal fluid fistula) that might lead to secondary infections, bone graft pseudoarthrosis or external contamination.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Retro-odontoid Degenerative Pseudotumour Causing Spinal Cord Compression and Myelopathy: Current Evidence on the Role of Posterior C1–C2 Fixation in Treatment

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Abstract *Background:* A retro-odontoid pseudotumour compressing the spinal cord and causing myelopathy is often associated with an inflammatory condition such as rheumatoid arthritis. A degenerative non-inflammatory retro-odontoid pseudotumour responsible for clinically relevant spinal cord compression is a rare condition described in small clinical series and is likely associated with craniovertebral junction hypermobility or instability–like conditions. For several years, direct removal of the lesion through an anterior or lateral approach has been advocated as the best surgical option. However, in the last decade the posterior approach to the craniovertebral junction, to perform C1–C2 fixation and C1 laminectomy without removal of the retro-odontoid tissue, has demonstrated its efficacy in reducing retro-odontoid pannus as well as in obtaining improvement of myelopathy.

Methods: In this paper we analyse the clinical and radiological outcomes of seven patients (five males and two females) treated with posterior C1–C2 fixation and C1 laminectomy for a degenerative non-inflammatory retro-odontoid pseudotumour responsible for spinal cord compression. C1 laminectomy provided immediate spinal cord decompression. We also review the relevant literature focusing on associated cervical degenerative conditions that may contribute to triggering or acceleration of atlantoaxial hypermobility or 'instability', causing formation of the retro-odontoid tissue.

Results: The mean follow-up period (of six followed-up patients) was 55.8 months (range 10–96 months). In all cases the Nurick score at the latest follow-up visit demonstrated clinical improvement; magnetic resonance imaging during follow-up demonstrated progressive reduction of the retro-odontoid pseudotumour in all but one patient, who died of surgery-unrelated disease in the early postoperative period. No vascular or neural damage secondary to C1–C2 fixation was observed.

Conclusion: C1–C2 fixation associated with C1 laminectomy is an effective surgical option to treat myelopathy secondary to a degenerative retro-odontoid pseudotumour. In these cases, direct removal of intracanalar tissue compressing the spinal cord is not required, as C1–C2 fixation is sufficient to cause its disappearance.

Keywords Atlantoaxial fixation · Craniovertebral junction · Cervical myelopathy · Odontoid process · Retro-odontoid pseudotumour

Introduction

A retro-odontoid pseudotumour [1], also known as pannus or a phantom tumour [2], is non-neoplastic, fibroreactive tissue involving the odontoid process and the surrounding structures of the craniovertebral junction [3, 4], causing different degrees of spinal cord compression.

Although the aetiology and pathophysiology of retro-odontoid pseudotumour formation and growth are unclear, various predisposing factors associated with its development have been identified, including inflammatory diseases (rheumatoid arthritis [5] and psoriatic arthritis [6]) and, less frequently, non-inflammatory pathologies (including post-traumatic pseudoarthrosis of the odontoid process, unstable odontoid fractures [7], os odontoideum [3, 8–10], postlaminoplasty kyphotic cervical instability [11], long-term haemodialysis [12], craniocervical junction malformations [10] and chronic atlantoaxial subluxation/instability [1, 13]). In cases related to inflammatory conditions, chronic damage of the C1–C2 ligamentous complex has been postulated as a likely factor inducing the development of retro-odontoid pannus [14]. Conversely, the pathogenesis is not clear in non-inflammatory cases, although atlantoaxial instability is

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often identified [13, 15]. Barbagallo et al. hypothesized that extensive subaxial cervical spine spondylotic changes, with reduced segmental hypomobility, might be coupled with compensatory hypermobility in the C1–C2 area and that such a phenomenon would eventually induce formation of retro-odontoid pannus [16].

Surgical treatment of a retro-odontoid pseudotumour can be performed by direct removal of the mass through a transsphenoidal/transoral/transpharyngeal approach [5, 17], a high cervical lateral approach [18] or a posterior extradural approach via laminectomy. Recently, posterior approaches to the craniovertebral junction, such as occipitocervical [10, 11, 19–22] or atlantoaxial [7, 9, 19, 23–28] fixation or C1 laminoplasty [2], have been reported by several authors, as they can induce a progressive, spontaneous reduction in the volume of the pseudotumour, avoiding the need for its direct resection.

We report our institutional experience with a cohort of patients presenting with compressive myelopathy related to a degenerative, non-inflammatory retro-odontoid pseudotumour, surgically managed with C1–C2 fixation combined with laminectomy of the C1 posterior arch. The timing of the pseudotumour reduction over medium-term follow-up and its relation to each patient's clinical and radiological features are analysed.

Materials and Methods

Patients

Between July 2009 and August 2016, seven patients (five male and two female), with a mean age of 63.7 years (range 55–76 years), were treated. Clinical and radiological data were retrieved from the patients' charts and/or the institutional electronic database. Clinical and radiological follow-up evaluations were performed at 3, 6 and 12 months after surgical treatment, and then every year.

Clinical and Radiological Assessments

All patients underwent neurological evaluation before surgery, postoperatively and at each follow-up visit. The Nurick score was used to assess the severity of myelopathy. The preoperative imaging protocol was the following: cervical x-rays with flexion–extension views, magnetic resonance imaging (MRI) and computed tomography (CT) angiography of the craniovertebral junction with multiplanar reconstruction. Retro-odontoid pannus features were classified according to MRI findings, as proposed by Yonezawa in 2013 [29]: type 1 pannus (hyperintense on T2 and hypointense on T1) was observed in two cases; type 2 pannus (hypointense on T2 and iso- or hyperintense on T1) was identified in one case; and type 3 pannus (a combination of hypo- and hyperintense on T2 and T1) was depicted in four cases. Interestingly, a patient with type 2 pannus had eggshell calcification surrounding the retro-odontoid pannus, clearly visible on the CT scan. Preoperative radiological evaluation also documented the presence of atlas assimilation in two patients, platybasia associated with C1 assimilation and consequent Chiari malformation type I in one patient, and diffuse idiopathic skeletal hyperostosis (DISH) in two other patients (Fig. 1). Both MRI and CT scanning clearly documented signs of subaxial spondylosis in all patients. Two patients had already undergone surgery for cervical spondylotic myelopathy at the C5–C6/C6–C7 and C5–C6 levels, respectively (Table 1). The preoperative radiological evaluation documented signs of advanced subaxial cervical spondylosis in all patients.

Cervical x-rays, CT scanning and MRI were also performed immediately after surgery. Moreover, the patients underwent MRI over the follow-up period to document the progressive reabsorption of retro-odontoid tissue.

Surgical Treatment

Depending on the patients' individual associated conditions (i.e. the presence of atlantoaxial assimilation), different surgical approaches were performed. Three patients underwent C1–C2 fixation according to the Goel–Harms technique. Occipitocervical fixation was performed in three patients: two with atlantoaxial assimilation and one with radiological signs of pre-existing C0–C1 fusion (in one patient, screws were inserted into the occipital condyles; in the remaining two cases, an occipital plate was implanted). In two other patients, C1–C2 fixation with C2 translaminar screws was performed (Fig. 1). Resection of the C1 posterior arch was performed in four of the seven patients, and C1–C3 laminectomy and posterior decompression of the foramen magnum were performed in two other cases (Table 1). All surgical procedures were performed by the senior author (GMVB).

Results

The mean follow-up period (in the six followed-up patients) was 55.8 months (range 10–96 months).

All surgical procedures were performed without perioperative complications. Postoperative x-rays and CT scans demonstrated correct device positioning in all cases. One patient died 10 days after the surgery because of acute kidney disease and multiorgan failure. The mean Nurick score of the other six patients decreased from 4 (range 3–5) to 2.3 (range 1–3) by the time of the latest follow-up. The neurological status

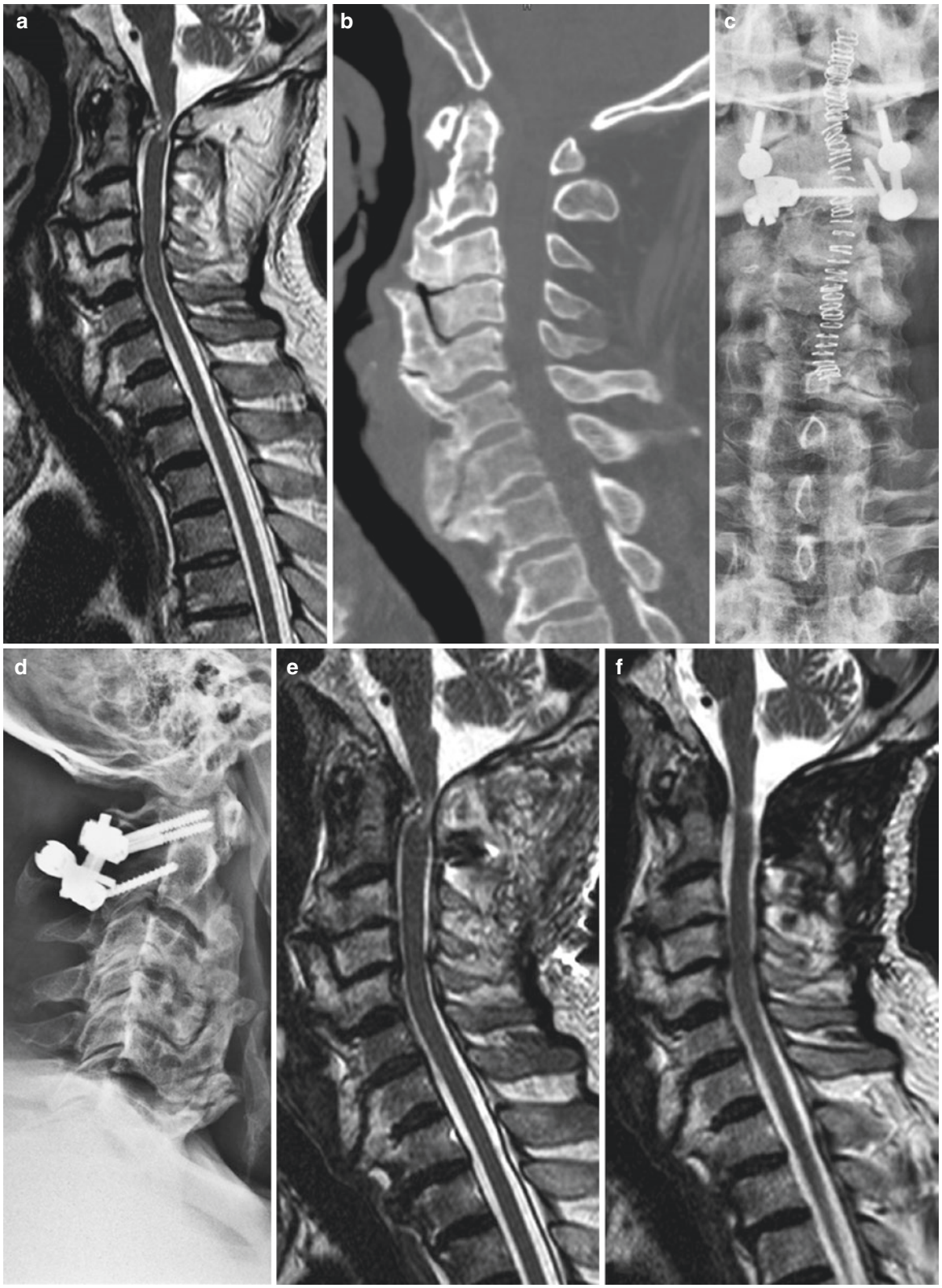

Fig. 1 Images of patient #7. (**a**) Preoperative sagittal T2-weighted magnetic resonance imaging (MRI) showing the presence of retro-odontoid pannus compressing and dislocating the spinal cord. (**b**) Sagittal reconstructed computed tomography (CT) scan showing multiple anterior osteophytes limiting the physiological motion of the cervical spine. (**c**) Postoperative anteroposterior x-rays and (**d**) postoperative lateral X-rays demonstrating C1–C2 fixation; because the right C2 pedicle was too narrow, a translaminar screw was used (a hybrid construct). Postoperative 1-month and 6-month spine MRI documented (**e**) the progressive reduction and (**f**) the disappearance of the retro-odontoid pseudotumour

Table 1 Review of institutional series of degenerative retro-odontoid pannus treated by posterior fixation

Patient (age/sex)	AAI or AAS (Y/N)	Subaxial spondylosis (level/s)	Associated conditions	Surgical procedure	Time to pannus reduction/ disappearance	Follow-up
55/M	N	C2–C3	–	C1–C2 fixation with C1 laminectomy (C1 lateral mass and C2 pedicle screws)	8 months	96
63/M	N	C5–C6 (fusion)	DISH	C1–C2 fixation with C1 laminectomy (C1 lateral mass and C2 pedicle screws)	12 months	83
76/F	N	C4–C5/C5–C6	Atlas assimilation	C0–C3 (C2 translaminar screws and C3 lateral mass)	NA	NA
58/F	N	C4–C5	Atlas assimilation	Occipito-C5 fixation (C2 translaminar screws and C3 lateral mass, C4 and C5 pedicle screws on one side and lateral mass screw on the other side)	6 months	78
64M	N	C4–C5/C5–C6	Atlas assimilation, platybasia and Chiari malformation	Occipito-C3 fixation in 1 pt with C1 assimilation (C2 translaminar screws and C3 lateral mass)	14 months reduction (pannus still present at 24 months FU)	46
59/M	N	C5–C6	Previous ACDF	C1–C2 fixation with C1 laminectomy (C1 lateral mass and C2 pedicle screws)	12 months	22
71/F	N	C3–C4/C4–C5/ C5–C6	DISH	C1–C2 fixation with C1 laminectomy (C1 lateral mass and C2 one translaminar screw and one pedicle screw)	6 months	10

AAI atlanto-axial instability, *AAS* atlanto-axial subluxation, *DISH* diffuse idiopathic skeletal hyperostosis, *ACDF* anterior cervical discectomy and fusion

remained unchanged immediately after surgery in all but one patient (the one showing calcified pannus). This patient reported transient worsening of pre-existing right upper limb weakness, which progressively recovered within 6 months after the surgery. A clinical improvement over the follow-up period was documented in all patients.

Radiological follow-up was performed in six of the seven patients. A significant and progressive decrease in retro-odontoid tissue was documented in all cases during the follow-up period. In the patient with calcified retro-odontoid tissue, incomplete reabsorption of the pseudotumour was documented 2 years after the surgery, albeit that the patient had experienced a slight clinical improvement (the Nurick score varied from 4 to 3). A direct anterior transoral approach to remove the pseudotumour was advised, but the patient did not consent to further surgery.

Discussion

Compressive myelopathy due to a retro-odontoid pseudotumour has been described as being related to chronic inflammatory conditions involving the skeletal system and joints, such as rheumatoid or psoriatic arthritis [5, 6, 24, 30]. Rare cases of retro-odontoid pannus associated not with an inflammatory condition but, rather, with a purely degenerative aetiology have been described [3, 7–12, 25]. The formation and growth of degenerative retro-odontoid pannus has been correlated with underlying instability of the atlantoaxial joint, which is responsible for continuous mechanical stress on the ligamentous and cartilaginous complex of the C1–C2 joint. This condition may lead to altered tissue turnover, causing formation of retro-odontoid pannus. However, even more rare cases of retro-odontoid pannus without overt C1–C2 instability have been also described [26, 31].

Symptomatic patients usually present with symptoms and signs of pyramidal tract involvement as a result of direct compression of the anterior columns of the spinal cord. Direct decompression can be achieved by resection of the abnormal retro-odontoid tissue through an anterior (transoral, transsphenoidal or transpharyngeal) [5, 17] or anterolateral route [18]. Posterior approaches to the craniovertebral junction have been more recently proposed as an alternative to anterior or anterolateral approaches, with the rationale of managing the retro-odontoid pseudotumour and treating the C1–C2 instability, which may have caused its formation. Different surgical techniques have been proposed to stabilize the craniovertebral junction, according to the different anatomical features of patients [32–34]. In general, when the anatomy of the C0–C1–C2 joints is normal and there are no signs of instability at the C2–C3 level, C1–C2 posterior fixation using a Magerl or Goel–Harms technique is sufficient to ensure solid fixation of the craniovertebral junction [32, 33, 35–40]. Conversely, in cases of atlas assimilation, platybasia, etc., occipitocervical fixation appears to be the most effective and safe surgical option [41]. Controversies exist regarding the best surgical strategy for patients without overt C1–C2 instability [26, 31]. In such cases a different pathogenesis, not related to the presence of C1–C2 instability, has

been postulated. Indeed, some authors have suggested that simple posterior neural decompression may be effective [2, 31]. Conversely, a case of a patient affected by a retro-odontoid pseudotumour not associated with C1–C2 subluxation, which was successfully managed by C1–C2 fixation, was reported in 2009 by Tanaka et al. [26], who observed a clinically and radiological significant reduction of the retro-odontoid pseudotumour 6 months after the surgery.

Atlantoaxial instability was not present in any patient included in the present series. In all cases we observed 'delayed' indirect neural decompression after C1–C2 fixation, related to the progressive reduction and disappearance of the retro-odontoid pannus. Moreover, we found a constant presence of subaxial spondylosis in all cases. This findings has not been well investigated in the literature. Chikuda et al. published findings from a series of ten patients who had retro-odontoid pannus without associated inflammatory diseases [4]. Eight patients in that series had no signs of C1–C2 instability, and four of those eight also had subaxial spondylosis. We have already hypothesized in a previous paper that the genesis of degenerative retro-odontoid pannus in cases not associated with an inflammatory condition or C1–C2 instability may be explained by abnormal mechanical stress on the craniovertebral junction, caused by reduced motion at subaxial levels [16]. In our series, the hypothesis of subaxial reduced motion as a cause of retro-odontoid pannus development is supported by the fact that two of our patients had previously undergone anterior cervical discectomy and fusion at subaxial levels and two other patients suffered from DISH.

In our previous publication on this topic, we also performed a systematic literature review of studies reporting retro-odontoid pannus treated by a posterior approach [16]. We found only a few reports on retro-odontoid pannus that was not associated with inflammatory disease or with atlantoaxial subluxation or instability.

The timing of the reduction/disappearance of the retro-odontoid tissue was variable in our series, as in the other published series [7, 10, 11, 13]. However, a retrospective classification of preoperative MRI scans according to the classification proposed by Yonezawa et al. [29] for rheumatoid retro-odontoid pannus showed that the timing of its disappearance in two patients with type 1 pannus and in three of four patients (one patient died 10 days after surgery) with type 2 pannus ranged from 6 to 12 months. One patient with type 2 pannus had a reduction but not disappearance of the pannus at follow-up. In this patient the presence of eggshell calcification of the retro-odontoid tissue probably limited the reabsorption process. The timing of pannus reduction/disappearance reported in our series is similar to those described in other recent series. We did not observe immediate or early postoperative disappearance of the pseudotumour, as reported by Shah et al. [42] and Ito et al. [43].

Posterior decompression including the C1 posterior arch was performed in four of the seven cases in the present series. We believe that this further surgical adjunct may ensure early neural decompression in patients with a more severe clinical status [44].

A further interesting finding in the present series is related to the medium-term follow-up results. Indeed, we observed that the progressive reduction of the retro-odontoid pseudotumour demonstrated by sequential follow-up MRI was accompanied by consequent improvement in the clinical status, documented by the reduction in the Nurick score. In a subsequent phase, both the clinical status and MRI findings remained stable. In two patients, worsening of subaxial spondylosis required further surgery 13 months and 38 months after the previous surgery, respectively. The clinical history of such cases supports the hypothesis that a retro-odontoid pseudotumour represents a clinical and anatomopathological manifestation of a larger and dynamic degenerative phenomenon, involving—at an earlier stage—the subaxial spine and then extending also to the craniovertebral junction.

Conclusion

A degenerative retro-odontoid pseudotumour not associated with inflammatory disease is a rare condition requiring proper management. Posterior craniovertebral junction fixation is a reliable and effective option leading to 'delayed' neural decompression related to progressive pannus reabsorption. This condition remains stable at medium-term follow-up, albeit that associated subaxial spondylosis, which is often observed in cases without atlantoaxial instability, may require further surgical procedures. Multicentre studies and larger clinical series are encouraged in order to clarify the optimal management of these complex conditions.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Insights into the Past and Future of Atlantoaxial Stabilization Techniques

Nabeel S. Alshafai, Agnieszka Kramarz, and Minou Behboudi

Abstract Over the past century, atlantoaxial stabilization techniques have improved considerably. To our knowledge there has been a scarcity of articles published that focus specifically on the history of atlantoaxial stabilization. Examining the history of instrumentation allows us to evaluate the impact of early influences on current modern stabilization techniques. It also provides inspiration to further develop the techniques and prevents repetition of mistakes. This paper reviews the evolution of C1–C2 instrumentation techniques over time and provides insights into the future of these practices.

We did an extensive literature search in PubMed, Embase and Google Scholar, using the following search terms: 'medical history', 'atlantoaxial', 'C1/C2', 'stabilization', 'instrumentation', 'fusion', 'arthrodesis', 'grafting', 'neuroimaging', 'biomechanical testing', 'anatomical considerations' and 'future'.

Many different entry zones have been tested, as well as different constructs, from initial attempts with use of silk threads to use of hooks and rod–wire techniques, and handling of bone grafts, which eventually led to the development of the advanced screw–rod constructs that are currently in use. Much of this evolution is attributable to advancements in neuroimaging, a wide range of new materials available and an improvement in biomechanical understanding in relation to anatomical structures.

Keywords Medical history · Atlantoaxial · Stabilization Fixation · History · C1/C2 · Instrumentation · History of surgery

Introduction

Stabilization of the spine started at the turn of the twentieth century, primarily for treating Pott's disease. The atlantoaxial joint is a treacherous surgical zone because of the proximity of vital neurovascular structures, such as the vertebral artery with all of its associated anatomical variations [1]. Added to that, it is a highly mobile joint and has a relatively frail osseous structure. Consequently, surgical approaches to stabilize the atlantoaxial joint are usually attempted later than approaches to stabilize the subaxial spine.

This historical review aims to identify the successes and failures of C1–C2 fusion—knowledge of which, we believe, provides a basis for continued innovation. Advances in neuroimaging, smart material availability and minimally invasive technologies have brought us to the dawn of a new way of stabilizing the atlantoaxial joint. Consequently, this evolution is expected to continue, and this paper touches on the ongoing improvements and what is on the horizon.

Methods

A retrospective literature review was conducted to identify historical landmarks in the evolution of atlantoaxial stabilization. Numerous surgeons have attempted to improve this operation, and this review does not provide a comprehensive list of all of them. It is merely an attempt to highlight the milestones that have led this operation to become what it is now.

Search Strategy and Study Selection

A non-systematic review with no year-of-publication limit was performed using the following databases: PubMed, Embase and Google Scholar. Moreover, a traditional Google

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search was performed and major book chapters were used. The keywords used were 'medical history', 'atlantoaxial', 'C1/C2', 'stabilization', 'instrumentation', 'fusion', 'arthrodesis', 'grafting', 'neuroimaging', 'biomechanical testing', 'anatomical considerations', 'innovations' and 'future'.

In addition, screening of the reference lists and bibliographies of the chosen publications extended the full list. Because of our linguistic capabilities, we were able to include papers written in English, Polish, French and Turkish. These searches yielded a total of 53 relevant articles.

Results

The Beginning of C1–C2 Stabilization: Wiring and Bone Grafting

The cause of cervical instability at the level of C1–C2 can be attributed to congenital deformity or acquired abnormalities such as those secondary to infection, trauma or a tumour [2]. The first attempt at stabilizing the atlantoaxial complex dates back to 1910, when Mixter and Osgood used heavy silk thread and laid the ground for more sophisticated wiring techniques (Fig. 1). This is consistent with the fact that this region is so complex that the techniques need to prove viable for less technically demanding regions [3]. By the early 1900s, it was known that spinal instability could be corrected using instrumentation but that fusion needed to be achieved, as instrumentation alone resulted in breakage.

In 1939, Gallie reported his method of laminar wiring (Fig. 2). Bone grafts from the patient's iliac crest were placed over the C2 spinous process and leaned against the C1 posterior arch. Then a steel wire was wrapped around the arch of the C1 and C2 processes, which gave good stability on flexion and extension movements, but not rotation. In 1978, to address this rotational instability problem, Brooks and Jenkins offered an alternative in which two iliac crest grafts were placed between the C1 and C2 arches and stabilized with two wires: one on each side [4]. However, the increased number of wires and the process of passing them under the C2 lamina posed a risk of greater neural injury.

Dickman and Sonntag published a modification of the wiring techniques in 1991 as an attempt to solve the two disadvantages of the two previously described techniques [5]. They passed a sublaminar wire from inferior to superior around the C1 posterior arch, then an iliac crest graft was placed as in the Gallie technique, with the difference that it was not just laid against the C1 arch but wedged underneath it and the surfaces underneath the bone graft were decorticated [6]. Next, a notch was made in the C2 spinous process; the cable was wrapped around the bone graft and secured in the C2 notch. Along with wiring techniques, a halo vest was required for good stabilization.

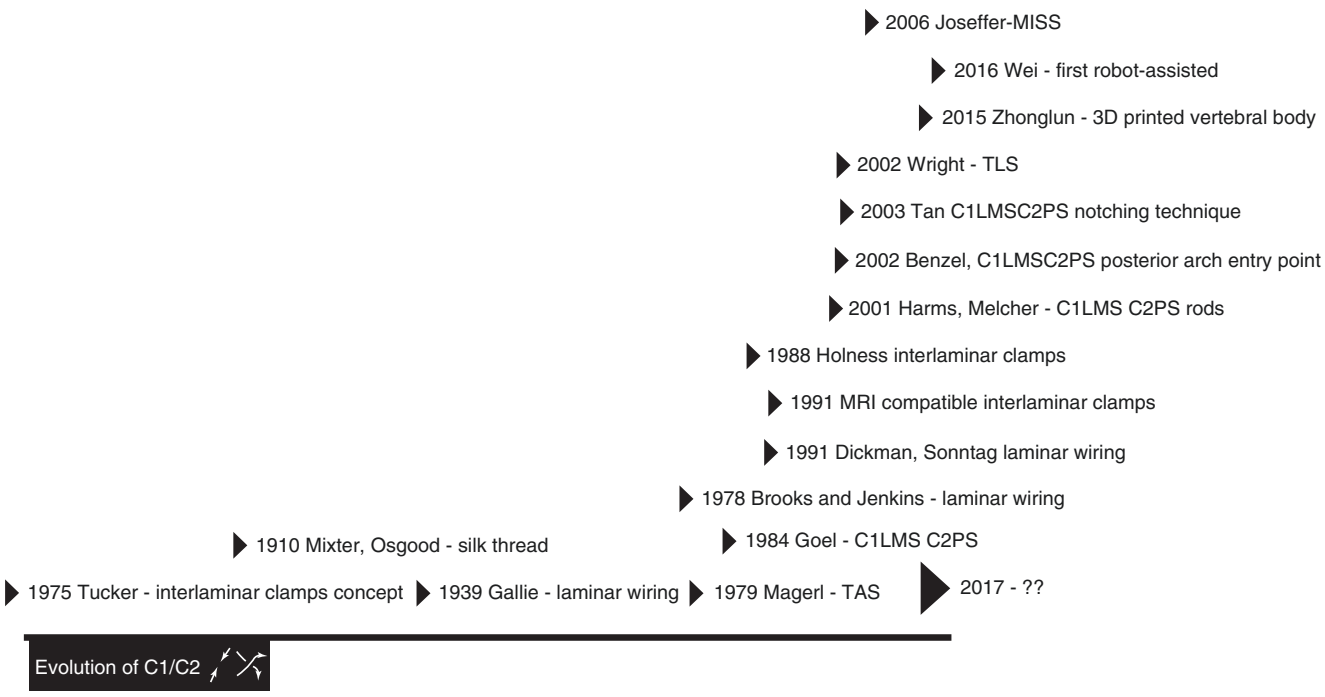

Fig. 1 Timeline of the evolution of C1–C2 stabilization techniques

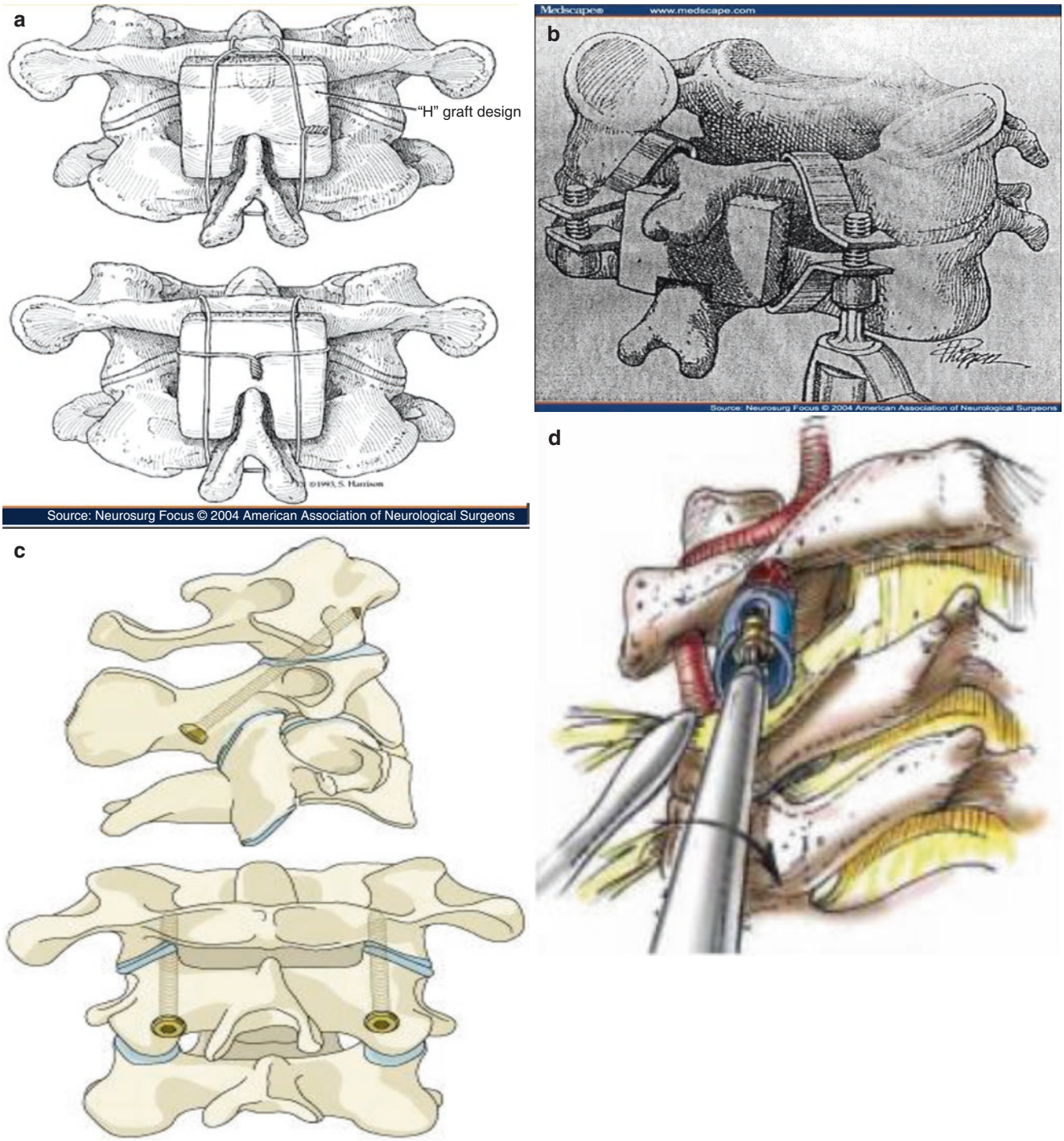

Fig. 2 (**a**) A Gallie type of fusion with two variations: (*upper*) (**a**) single loop of wire around the posterior arch, securing the only bone graft; and (*lower*) a double-loop technique with a wire around the arch of C1 [24]. (**b**) Demonstration of the use of interlaminar clamps. The clamps are placed on each side over the lamina and are secured by tightening of the screws. A bone graft is placed between the spinous processes to enhance fusion [17]. (**c**) Magerl first described the use of transarticular screws. This is a relatively simple and inexpensive way to fixate the C1–C2 joint [25]. (**d**) Placement of a C1 lateral mass screw [10]. (**e**) A completed C1 lateral mass screw–C2 pedicle screw construct (Goel's technique) [10]. (**f**) Anterior view of the atlas, depicting the fixation and the final position of the screw, and lateral view showing bone graft insertion in the facet joint [9]

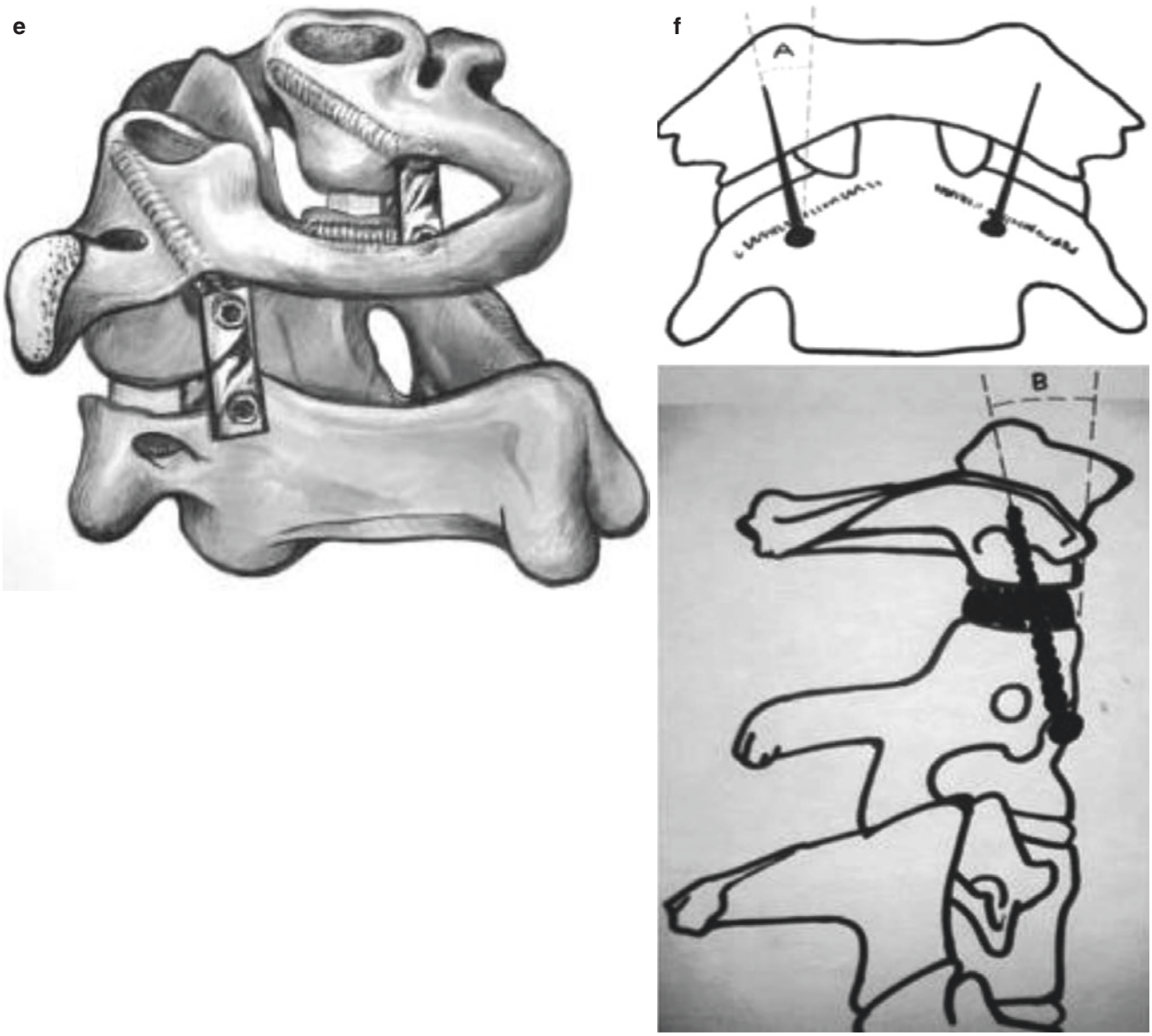

Fig. 2 (continued)

Wiring techniques are still used, with the improvement of using more modern materials, such as titanium, which are more flexible and do not pose as great a risk of neural damage.

Interlaminar Clamps, Hooks and Magnetic Resonance Imaging

Interlaminar clamps, just like wiring techniques, require an intact lamina and are not solutions for fractures or when decompression is needed [7]. They also require use of a halo vest for satisfactory stabilization. They were introduced to avoid the need for passing sublaminar wires and to reduce the risk of neural injury. They were successful in the subaxial spine [8]. Therefore, as an expected progression, surgeons tried to apply them to C1–C2 [8].

Tucker introduced the concept in 1975, and Holness described the first case of application of interlaminar clamps to the atlantoaxial joint in 1988. In the meantime, magnetic resonance imaging (MRI) started becoming a primary diagnostic modality. To avoid ferromagnetic artefacts, surgeons started using alternatives to stainless steel, such as titanium alloy clamps, which had an impact on all stabilization techniques. In contrast to the wiring techniques that are still being used, interlaminar clamps have fallen out of favour for use in C1–C2, mostly because of failure of the hardware, which tends to slip before fusion is achieved [8–10].

The Transarticular Screw

In 1979, Magerl introduced a technique that, for the first time, caused complete obliteration of rotational movement and could be used when the posterior axis was damaged: the transarticular screw (TAS) technique. However, this technique has a steep learning curve and poses a great risk to the vertebral artery. This risk was decreased by applying the concept of stereotaxis, which was developed as early as 1906 and used for the brain for the first time in 1947 [7]. Nevertheless, this technique is still not applicable to cases with a high-riding vertebral artery. Other drawbacks of the TAS technique are the need to align the vertebrae before screwing and the fact that the screws cannot be adjusted afterward.

Screw–Rod Constructs and Biomechanical Testing

To eliminate the drawbacks of the TAS, use of C1 lateral mass screws and C2 pedicle screws (C1LMS–C2PS) was introduced. This technique is just as stable as the TAS technique, but it is easier to perform and poses a smaller risk to the vertebral artery [11]. In 2001, Harms and Melcher substituted the plates used by Goel with rods and contributed to popularization of the method.

In 2002, Benzel et al. reported a different entry point: through the posterior arch. In 2003, Tan et al. described a notching technique in which they made a different entry point by notching the posterior arch. In 2002, Wright developed the translaminar screw (TLS) for C2, which is less biomechanically stable but poses only a very small risk to the vertebral artery.

To sum up the surgeon's armamentarium, the C1LMS technique done by any of the above three variants—Harms, Benzel or Tan—can be combined with one of the following C2 screws: the pedicle, the pars (the same as the pedicle but shorter) and the translaminar screw. The screws are connected with rods to create stabilization. This allows a wider range of possible angles for adjustment to individual anatomy (including a high-riding vertebral artery) and for reduction and adjustment of the final fixation position after the screws are put in.

This presents a wide range of options available for the modern neurosurgeon to choose from, using different innovative techniques with their practical advantages. The choice of technique needs to be carefully balanced between the individual anatomy of the patient and the stability that each technique provides, as well as the surgeon's capabilities.

Biomechanical testing has its origin in antiquity and was attempted a long time ago by Hippocrates, Leonardo da Vinci and, more comprehensively, by Borelli, who is considered the father of spinal biomechanics. The introduction of biomechanical testing laboratories resulted in deeper understanding of the load-bearing structures, described by the Denis thoracolumbar three-column model, which laid the grounds for modern spinal instrumentation [12]. After screw–rod systems were introduced, Richter et al. released a paper comparing six C1–C2 stabilization techniques, with use of biomechanical studies [11].

Minimally Invasive Techniques and Computed Tomography–Guided Neuronavigation

The continuous improvement in the quality of neuroimaging, and surgeons' increasing familiarity with it, are causing its benefits (such as lowering morbidity) to outweigh those of using fluoroscopy [13–15]. Given that we are entering an era of simulation, this also allows more effective preoperative planning, which is key for the atlantoaxial region, given its high rate of anatomical variability. The introduction of this technique has opened new doors for previously implausible surgical techniques because unlike fluoroscopy, it offers three-dimensional (3D) images. However, the drawback of increased patient and doctor exposure to radiation (up to 40 mGy with CT-guided neuronavigation versus up to 6 mGy with fluoroscopy) still needs to be addressed [16].

Minimally invasive spine surgery (MISS) is gaining more recognition for use in the subaxial spine, because of its main advantages in decreasing tissue dissection, postoperative pain, intraoperative blood loss, infection rates and hospitalization time. Nevertheless, these improvements still need to be proved to be safer than open techniques specifically for the atlantoaxial region. In April 2006, Joseffer published the first application of MISS in atlantoaxial instability. Since then, there have been reports on more than 14 patients and 17 cadavers (by Holly, Joseffer [March 2010], Srikhanta, Taghva and Bodon). In December 2013, Sonntag expressed an opinion in a commentary that, theoretically, MISS stabilization of C1–C2 has advantages that need to be investigated further.

Anterior Approaches

The anterior approach has been neglected in the literature, as it is a challenging procedure and carries a risk of retropharyngeal wound breakdown [7]. It has been speculated that the anterior approach is possible but has limited application. Earlier techniques using wiring have been replaced by more

advanced techniques and instruments because of drawbacks and high rates of failure [17]. In 1994, Goel et al. described a technique for treatment of an unstable craniovertebral junction, called transoral instrumentation. Harms and colleagues have been using this technique for anterior cervical spine fusion [7]. An advantage of this technique is that it avoids future posterior neck incision for C1–C2 fixation after anterior decompression.

Barbour first introduced anterior TAS fixation of C1–C2 in 1971, and it was used to stabilize the lateral atlantoaxial joints in odontoid fractures [18]. In 1987, Leson et al. introduced a modified technique of Barbour's in which the screws were entered laterally on the midsagittal line at the small of the groove in the lateral mass [18]. This technique was introduced again in 2007 by Schaeren et al. [19]. The stability of this technique has been proved in human cervical spines on biomechanical testing and in an anatomical review [17, 20].

Minimally Invasive Anterior Transarticular Screw Fixation

Wang et al., using intraoperative fluoroscopic guidance and micro-endoscopy, introduced a new minimally invasive technique for anterior atlantoaxial fixation and fusion. Some scholars believe that this approach could replace transoral surgery, allowing a direct anterior approach to C1–C2 [19]. This technique is functional for achieving C1–C2 stabilization with less blood loss. However, if decompression is needed, it is not possible through this approach to the cervical spine. Despite being a useful approach, it has its limitations. It is a challenging technique for less experienced surgeons [19].

Minimally Invasive Techniques and CT-Guided Neuro-Navigation

The continuous improvement in quality of neuroimaging has contributed to less need to use fluoroscopy leading to lower surgical morbidity [13–15]. This is key, allowing more effective pre-operative planning for the atlantoaxial region which is associated with a high rate of anatomical variability.

Controversies still exist regarding the increased patient and doctor radiation exposure (CT-guided Neuro-navigation provides up to 40 mGy compared with fluoroscopy up to 6 mGy radiation). [16] This technique opened new doors for previously implausible surgical techniques, because as opposed to fluoroscopy it offers 3D images.

Minimally invasive spine surgery (MISS) is gaining more recognition for the sub-axial spine due to their main advantages in decreasing tissue dissection, postoperative pain, intraoperative blood loss, decreased infection rate and shorter hospitalization time. Nevertheless, these improvements still need to be proved scientifically to be safer than open technique specifically for the atlantoaxial region. In April 2006, Joseffer published the first application of MISS in atlantoaxial instability. Since then, there has been literature on more than 14 patients and 17 cadavers (Holly, Joseffer (March 2010), Srikhanta, Taghva and Bodon). In December 2013, V. Sonntag expressed that theoretically MISS stabilization of C1/C2 has advantages that need to be investigated further.

Discussion

Posterior approaches to the spine were the first to be described, since the posterior aspects of the spine are more superficial and the spinous processes are easy to palpate [21]. Anterior approaches were developed independently of spinal surgery. Smith and Robinson described anterolateral approaches in 1958. In general, a trend away from posterior toward anterior techniques was observed in spinal surgery in the 1950s, mainly because it allowed surgeons the ability to directly address pathologies that occurred in the anterior spine [21]. Nowadays, however, posterior TAS and C1LMS–C2PS fixation are the most widely used and acceptable techniques [17]. Over the years, the evolution of instrumentation techniques for C1–C2 fusion, from the use of posterior wiring methods to the use of TAS and C1LMS–C2PS, have advanced the efficiency and effectiveness of fusion to almost 100% [2]. Nevertheless, although the modern surgeon is better equipped, old concerns such as blood loss, pseudoarthrosis and neurological injury are still a concern. We argue that surgeons have laid stable groundwork for this to become a mainstream neurosurgical operation with specific guidelines that still need to be developed. One can notice that as new techniques are developed, they build on previous developments—for instance, MISS, which uses the standard C1LMS–C2PS or TAS but just in an innovative way. Also, techniques that were being abandoned are now being rediscovered as innovative technology becomes available, such as the revival of the TAS in stereotactic surgery. In the words of Shakespeare, 'What's past is prologue.'

The Future

In 2015, Zhongjun published a paper on the first successful reconstruction of the upper cervical spine (UCS) with a personalized 3D-printed vertebral body in an adolescent with Ewing sarcoma. These kinds of implants give hope for patients with severe deformities, resulting in better biomechanical sta-

bility and enhanced bone healing. In April 2016 the first robot-assisted posterior C1–C2 TAS fixation was reported by Wei [22]. Improvement of existing screw placement techniques with application of innovative technology is the expected trend in this century [23]. The future of atlantoaxial stabilization surgery may include a few or all of the following entering mainstream neurosurgery: frameless stereotactic surgery, micro-robotic dexterity enhancement, personalized 3D-printed substitutes for natural bones and robotic surgery.

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Realignment of Basilar Invagination by C1–C2 Joint Distraction: A Modified Approach to a Paradigm Shift

Francesco Cacciola, Bronek Boszczyk, Paolo Perrini, Pasquale Gallina, and Nicola Di Lorenzo

Abstract *Background:* Distraction of the C1–C2 joint and maintenance thereof by introduction of spacers into the articular cavity can successfully and durably reduce basilar invagination (BI). Thus, with the adjunct of instrumented fusion and decompression, BI-induced myelopathy can be efficiently treated with a one-stage posterior approach. This intervention is technically challenging, and in this paper we describe a procedural variation to facilitate the approach.

Methods and Results: Through a description of a case of BI, the main anatomopathological alteration underlying and perpetrating the condition of BI is elucidated. A technique of realignment of BI is then described in which this alteration is specifically targeted and neutralized. The result is a single-stage posterior-only approach with decompression, C1–C2 distraction and introduction of poly(methyl methacrylate) (PMMA) into the joint cavity. Instrumented occipitocervical fusion completes the procedure.

Conclusion: C1–C2 joint distraction is a technically demanding procedure. By providing a modification of the original technique and a detailed description of the crucial steps necessary to successfully and safely carry it out, we hope to make this excellent procedure more approachable.

Keywords Basilar invagination · Surgical treatment · Joint distraction · Posterior fusion

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Introduction

Atlantoaxial joint distraction and direct lateral mass fixation to treat basilar invagination (BI), as described by Goel in 2004 [1], has represented a true paradigm shift in the approach to a complex structural problem. In symptomatic BI, a condition in which myelopathy is essentially caused by an odontoid process that encroaches upward and posteriorly on the spinal cord, that same odontoid process has classically—and quite rightly—been seen as the main culprit [2, 3]. If the odontoid process was extending too far into the foramen magnum and impinging on the spinal cord, it was in a place where it was not supposed to be, and it was therefore 'at fault', so to speak.

As a consequence the transoral approach was adopted and subsequently refined to efficiently eliminate the culprit by excising the odontoid process and thus relieving pressure on the cord [4–10].

While this technique definitely remains valid and is still the procedure most widely used for symptomatic BI today, Goel's discovery was truly revolutionary, as it 'saved the messenger'. According to this vision of things, the odontoid process was no longer the culprit, even though it effectively impinged on the cord; rather, it was 'the messenger carrying the bad news' of an aberrant process that resided somewhere else. As a matter of fact, in BI it is not the dens that is malformed; rather, it is its adjoining structures that put it in the wrong place.

BI generally entails a varying combination of platybasia, assimilation of the atlas, atlantoaxial dislocation (AAD) and/or anterocaudal inclination of the C1–C2 articular surfaces, leading to the atlanto-occipital complex 'sliding' anteroinferiorly with respect to the axis and thus, in turn, causing the axis and dens to migrate upward and backward.

It is the identification and definition of these corollary—or, rather, causative—features that has led over the last two decades to a refined classification of BI that draws attention away from the odontoid impinging on the cord as the end

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product of the pathological process and, instead, elucidates the fundamental role of C1–C2 facet conformation and disposition as the root cause of the problem and therefore also the essential aim of reparative intervention [11].

While a detailed explanation of the classification of C1–C2 facetal anomalies clearly transcends the purpose of this paper, we attempt to outline the basic principles of the anomaly and its direct repair with the help of the following case description.

Case Report and Technical Note

A 56-year-old woman with an uneventful past medical history was referred to our department for worsening myelopathy and pain in the nape of the neck. On neurological examination she scored IIIA on the Ranawat scale for myelopathy [5].

She underwent cervical magnetic resonance imaging (MRI), which showed that the spinal cord was severely compressed at the level of the craniovertebral junction. Furthermore, the cord showed signs of hyperintensity on T2-weighted images and initial cavitation (Fig. 1). To better identify the nature of the compression, a computed tomography (CT) scan was done, which showed assimilation of the atlas (AA) with BI and AAD. As a result, the odontoid peg was protruding into the foramen magnum (Fig. 2a, c).

The patient was thus scheduled for posterior-only surgery in the prone position. Transcranial traction of 16 pounds was applied with the head supported by a horseshoe headrest and the table inclined to roughly 30° anti-Trendelenburg (Fig. 3a).

After a vertical incision was made, centring on C1–C2, the posterior arch of the atlas and the laminae and spinous process of C2 were exposed. Subsequent dissection was carried out laterally to identify and section both C2 ganglions to

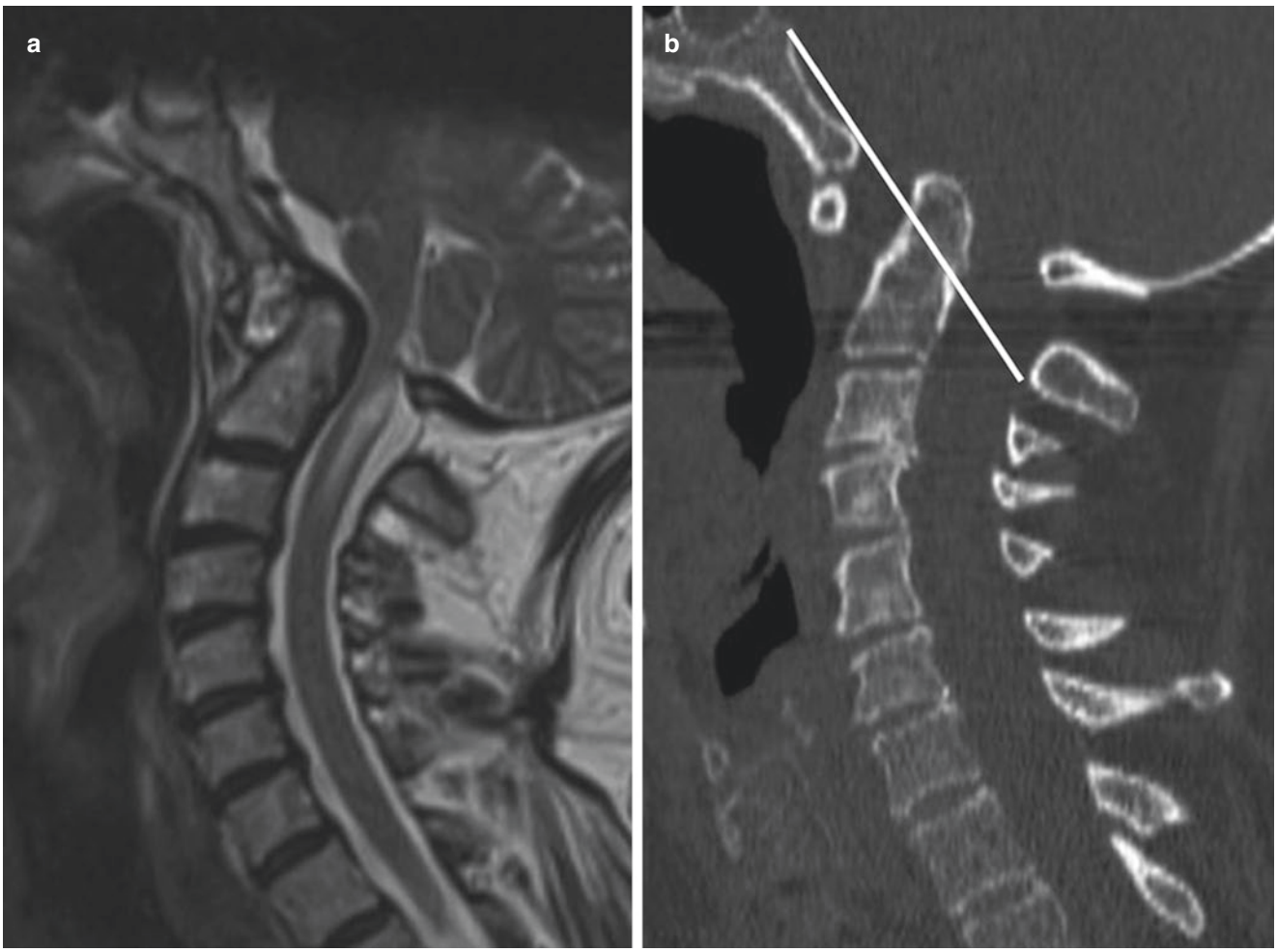

Fig. 1 (**a**) Sagittal T2 magnetic resonance imaging (MRI) scan showing the craniocervical region with basilar invagination (BI), severe compression of the spinal cord, intramedullary changes of oedema and initial syrinx formation. (**b**) Sagittal computed tomography (CT) scan showing BI with atlantoaxial dislocation (AAD) and clear upward migration of the odontoid process. Note the position of the dens traversing Chamberlain's line (*white*). (Reprinted from Cacciola et al. [12], with permission)

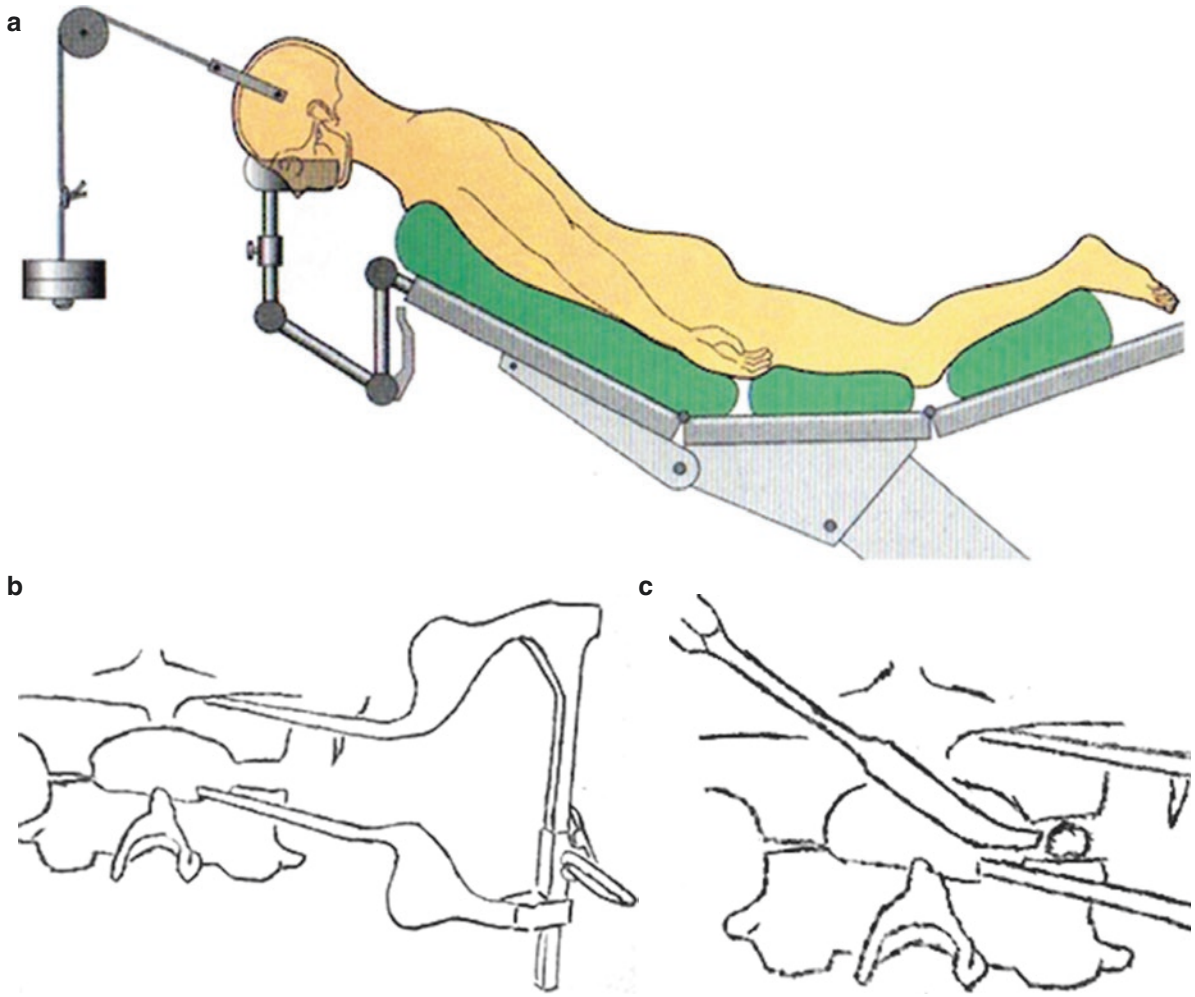

Fig. 2 (**a**) Prone position of the patient with the head in traction and resting on a horseshoe headholder. (Reprinted from Goel and Laheri, with permission.) (**b**) Positioning of a laminar spreader between the occiput and the C2 lamina with ensuing opening of the C1–C2 joint space. (**c**) Introduction of small poly(methyl methacrylate) (PMMA) spheres into the opened joint space with a dissector. (Reprinted from Cacciola et al. [12], with permission)

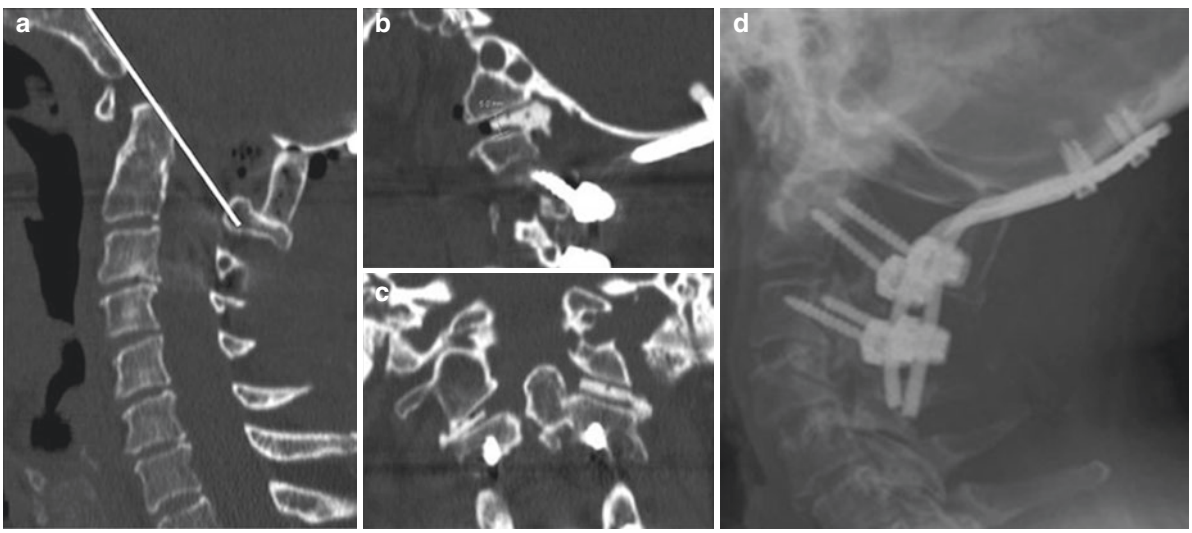

Fig. 3 (**a**) Sagittal computed tomography (CT) scan centred on the C1–C2 joint postoperatively. Note the oblique inclination of the joint and the filling of the distracted joint with hyperdense material [poly(methyl methacrylate) (PMMA)] to an opening of 5 mm. (**b**) Coronal CT scan through the C1–C2 joints bilaterally. Note the distracted joints and PMMA in the cavities. (**c**) Lateral cervical radiograph showing the instrumented fusion. (Reprinted from Cacciola et al. [12], with permission)

provide good exposure of both C1–C2 joints. During this stage the perivertebral venous plexus can often bleed significantly, and this can be made even worse by small emissaries of the vertebral artery. The surgeon should be prepared for this and, through sound knowledge of the anatomy, not be withheld by such bleeding, as this can sometimes be so copious as to lead to the misbelief that the vertebral artery itself may have been injured.

Once the joints were exposed, a small osteotome was introduced into the joint spaces and rotated to achieve some distraction and mobilization of the joint complex. Subsequently, a cervical laminar spreader was inserted between the occiput and the superior rim of the C2 laminae to distract the joint space in a controlled fashion and keep it distracted at first on the right side (Fig. 3b).

Once the joint was distracted, poly(methyl methacrylate) (PMMA) was made in a standard fashion and stirred until it reached a semi-solid state, just enough to allow for the making of small pea-sized spheres. These were then introduced into the joint with a dissector until the cavity was felt to be sufficiently filled with cement (Fig. 3c). Once the cement had turned solid, the laminar spreader was taken off and put into place on the contralateral joint, and the operation was performed in the same fashion on the left side. Following that, the foramen magnum was decompressed in a standard fashion, and instrumented C0–C3 fusion was performed with integration of an autologous tricortical bone graft harvested from the posterior iliac crest, which was placed under compression between the occiput and the superior border of the C2 spinolaminar complex.

The patient was discharged with a hard collar and, at 1-month follow-up, showed a neurological improvement of one grade (to II) on the Ranawat scale.

At that stage a CT scan was done, which showed a clear realignment of the odontoid process moving below Wackenheim's line, thus normalizing and essentially reversing the condition of BI (Fig. 2b). The AAD showed marked reduction as well.

Follow-up at 6 months confirmed maintenance of the clinical improvement, and imaging studies showed persistence of the realignment.

Discussion

Goel's posterior-only C1–C2 distraction and fusion technique has revolutionized the treatment of BI, with the potential to relegate the need for a transoral approach to a more restricted selection of cases. The rationale for this technique lies in the fact that the origin of the problem, or the 'culprit', is the conformation of the atlantoaxial joints. Being generally inclined in an oblique direction, mostly in the anteroinferior direction (Fig. 3b), the occipitoatlantal complex slides forward, thus causing AAD and BI. Therefore, careful attention should always be given to the joint complex when facing a case of BI as, if these findings are present, the whole pathology can be corrected with a posterior-only distraction operation, as depicted in our case.

In the original technical description, intraoperative distraction of the joint (besides the head traction) is achieved by insertion of small osteotomes inside the joint and rotation of the same so as to open the joint. Titanium spacers are then impacted into the joint. The spacers have a bullet-shaped nose, and that edge is abutted at the entrance of the joint cavity. Then, with the help of a mallet and an impactor, they are made to advance into the joint cavity, similarly to the insertion of a cage into the lumbar disc space during posterior lateral interbody fusion (PLIF). As these manoeuvres might appear challenging to the first-time user of this technique, given both unfamiliarity with the anatomical region and the proximity of the vertebral artery on one side and the spinal cord on the other, we have introduced a variation into these two steps. With the use of a laminar spreader, the joint spaces are distracted gradually and in a controlled fashion. Then, once the joint cavity is open, the small spheres of semi-cured PMMA can be inserted into the cavity with the help of a dissector, without the need to exert any force on the structures. Once the PMMA has reached the solid state inside the joint cavity, the spreaders are taken off and the distraction is thus maintained.

Conclusion

C1–C2 distraction has proven to be a reliable technique in the treatment of basilar invagination. Considering the relatively low overall incidence of this pathology, many surgeons lack sufficient exposure to gain experience in the transoral approach, which is classically the mainstay of treatment for this condition. Even though it is still technically demanding, the joint distraction technique could prove to be more approachable by a spinal surgeon who is mainly confident with posterior approaches, and our technical modification might further facilitate the endeavour.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Grisel's Syndrome: Non-traumatic Atlantoaxial Rotatory Subluxation—Report of Five Cases and Review of the Literature

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Abstract *Background:* In children, when unresponsive neck rigidity and distress are observed after ear, nose and throat (ENT) surgical treatment or nasopharyngeal inflammation, Grisel's syndrome should be suspected. This is a rare syndrome involving non-traumatic rotatory subluxation of the atlantoaxial joint. Conservative management with external cervical orthoses and empirical antibiotic, muscle relaxant and analgesic therapy should be the first choice of treatment. Surgical stabilization is indicated when high-grade instability or failure of stable reduction are observed. The instability is graded according to the classification system devised by Fielding and Hawkins. Several recommendations for treatment are available in the literature, but there are no common guidelines. In this paper, the authors discuss the need for prompt diagnosis and treatment considerations.

Case Description: Five children with Fielding type I–III rotatory subluxation are reported. Three patients were treated with a cervical collar, and one patient was treated with skull traction and sternal–occipital–mandibular immobilizer (SOMI) brace application. Surgical treatment was necessary for one patient after failure of initial conservative management. The intervals between the onset of torticollis and radiological diagnosis ranged from 12 to 90 days. A relationship between an increased grade of instability and delayed diagnosis was observed.

Conclusion: In children with painful torticollis following ENT procedures or nasopharyngeal inflammation, Grisel's syndrome should always be suspected. Cervical magnetic resonance imaging (MRI) allows prompt and safe diagnosis, and a three-dimensional computed tomography (CT) scan provides better classification of the instability. Surgery, which is indicated in cases of high-grade instability or failure of conservative treatment, may be avoided with prompt diagnosis.

Keywords Grisel's syndrome · Fielding classification · Atlantoaxial instability · Atlantoaxial rotatory subluxation

Abbreviations

3D	Three-dimensional
ADI	Atlantodental interval
CT	Computed tomography
ENT	Ear, nose and throat
MIP	Maximum-intensity projection
MRI	Magnetic resonance imaging
SOMI	Sternal–occipital–mandibular immobilizer

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Highlights

- Grisel's syndrome is a non-traumatic rotatory subluxation of the atlantoaxial joint, following nasopharyngeal inflammation or ear, nose and throat (ENT) procedures.
- The primary treatment of Grisel's syndrome is conservative.
- In cases of failure of conservative treatment, surgery is indicated.
- Delayed diagnosis can result in a need for surgery.
- Rigid C1–C2 or C1–C2–C3 fixation is the best surgical option because it provides immediate spinal stability in all planes of the atlantoaxial complex.

Introduction

In 1930 Grisel reported a spontaneous rotatory subluxation of the atlantoaxial joint after pharyngitis not associated with trauma or bone disease [1]. The first description of an association between inflammation of vertebrae caused by a retropharyngeal abscess and vertebral luxation was reported by Aëtius of Amida, who lived in the fifth to sixth centuries CE. Later the syndrome was described by Rhazes (865–925 CE), quoting from Hippocrates in the *Epidemics* [2, 3].

This is a rare syndrome with a predominant occurrence in young patients, mainly younger than 12 years [4–12]; only 14 cases in patients aged 18 years and older have been reported since 1830 [13]. Grisel's syndrome may occur after nasopharyngeal inflammation [14–17] or after ear, nose and throat (ENT) procedures such as tonsillectomy, adenoidectomy, mastoidectomy, cochlear implantation, uvulectomy or tympanoplasty [4, 18–25].

A neuroradiological workup, with x-rays and computed tomography (CT) scans of the cervical spine, discloses the diagnosis. Magnetic resonance imaging (MRI) provides information about the grade of lymphadenopathy and additional data about the ligament status of the C1–C2 joints. Early detection is the key to prompt treatment because the degree of instability of the rotatory subluxation can vary significantly. Depending on the Fielding and Hawkins classification [26] (Fig. 1), the choice of treatment will involve initial conservative management, with the patient wearing an external orthoses such as a cervical collar, halo vest or sternal–occipital–mandibular immobilizer (SOMI) brace, depending on the grade of instability. Empirical or targeted antibiotic, muscle relaxant and analgesic medications constitute the pharmacological treatments for this condition, depending on the clinical status. Surgical upper cervical internal fixation and fusion are options for a greater degree of

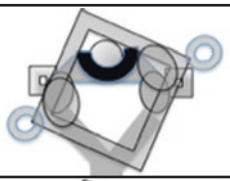
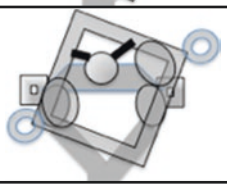
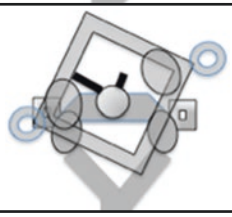
Schematic illustration of Fielding and Hawkins classification of the Grisel's syndrome		
Type I		No subluxation Pure rotation of the Atlas in relation to the Axis with no or less than 3 mm of anterior displacement of the atlas
Type II		Unilateral subluxation of one atlanto-axial joint. Atlas anterior displacement of 3–5 mm, associated with deficiency of the transverse ligament.
Type III		Ventral subluxation of Atlas in both Joints. Atlas rotated with anterior displacement > 5 mm, associated with deficiency of the transverse and alar ligaments
Type IV		Dorsal subluxation of Atlas. The posterior displacement of the atlas is associated with deficient odontoid process with possible fractured Dens axis or congenital Dens-aplasia

Fig. 1 Fielding and Hawkins classification, reported in 1977

Table 1 Patient demographic and diagnostic details

Case	Age (years)	Sex	Initial diagnosis	Time between torticollis and radiological diagnosis (days)	Fielding and Hawkins classification, and ADI	Treatment
#1	5	Male	Pharyngotonsillitis	12	Type I ADI <3 mm	Antibiotic therapy (oral and intravenous) and analgesic therapy (oral) Cervical collar for 4 weeks
#2	7	Male	Adenotonsillectomy	7	Type I ADI <3 mm	Antibiotic, anti-inflammatory and steroid therapy (oral) Cervical collar for 4 weeks
#3	4	Male	Tonsillitis	60	Type II ADI 4 mm	Antibiotic therapy (intravenous) Cervical collar for 8 weeks
#4	12	Female	Tonsillitis	90	Type II ADI 4 mm	Skull traction for 4 days SOMI brace for 12 weeks
#5	9	Male	Adenoidectomy	56	Type III ADI 6.7 mm	Collar for the first 6 weeks after C1–C2–C3 internal fixation

ADI atlantodental interval, *SOMI* sternal–occipital–mandibular immobilizer

instability or after conservative treatment failure [24, 25, 27–32]. In this paper, the authors present five cases of this condition (summarized in Table 1), which demonstrate the importance of prompt diagnosis in relation to the choice of treatment.

Case Series

Case #1

A 5-year-old boy developed painful torticollis after 2 days of fever due to pharyngotonsillitis. Laboratory examinations showed leucocytosis and an increased C-reactive protein (CRP) level (51.15 mg/L). He was treated with 7 days of oral antibiotic therapy (amoxicillin clavulanate), and prolonged oral analgesic therapy (ibuprofen). Twelve days later his CRP level had decreased to 7.98 mg/L, but the torticollis was unchanged on clinical examination. A cervical ultrasound evaluation showed bilateral submandibular lymphadenopathy extending along the jugular chain and in the posterior triangle. After hospital admission to check the lymphadenopathy status, MRI was performed. A lymph node consolidated and packed in the right parapharyngeal space (Fig. 2a), rotatory subluxation at C1–C2 with intact ligaments compatible with Fielding type I atlantoaxial subluxation (Fig. 2b) and no critical involvement of the spinal cord were detected.

A cervical collar was applied, intravenous antibiotic therapy was administered (ceftriaxone plus metronidazole plus vancomycin) for 7 days and the patient was discharged home. Follow-up with MRI after the patient had worn the cervical collar for 4 weeks revealed almost complete resolution of the inflammation and resolution of the subluxation (Fig. 2c).

Case #2

A 7-year-old boy, receiving treatment with ceftibuten and tranexamic acid, presented with a 3-day history of painful torticollis and a headache following adenotonsillectomy. Bilateral laterocervical lymphadenopathy with tympanic membrane and external auditory canal hyperaemia were revealed on an ENT examination and otoscopy. The cervical x-ray findings were a right lateral flexion of the neck and loss of cervical lordosis. Neck ultrasound scans showed reactive lymph nodes along the jugular chain. No improvement was obtained with ibuprofen add-on therapy and rest.

At 7 days postoperatively a CT scan (Fig. 2d) and MRI (Fig. 2e) showed Fielding type I rotatory atlantoaxial subluxation. The patient continued to receive antibiotic therapy with ceftibuten, paracetamol with codeine, prednisone, pridinol mesylate, and chlorhexidine, and wore a cervical collar. At discharge the patient's symptoms had resolved and at 1-month follow-up, MRI confirmed his recovery from the instability (Fig. 2f).

Case #3

A 4-year-old boy presented to the emergency department with torticollis and his head tilted to the left. Three months prior to presentation he had suffered from tonsillitis. His initial complaints included non-specific sensory symptoms involving the upper limbs and pain in the neck with stiffness. The tonsillitis had settled uneventfully with antibiotics.

On examination there was fixed rotation of the neck to the left, with a normal neurological assessment. Active manipulation of the head and neck was painful; therefore, reduction was impossible. A CT scan with maximum-intensity projec-

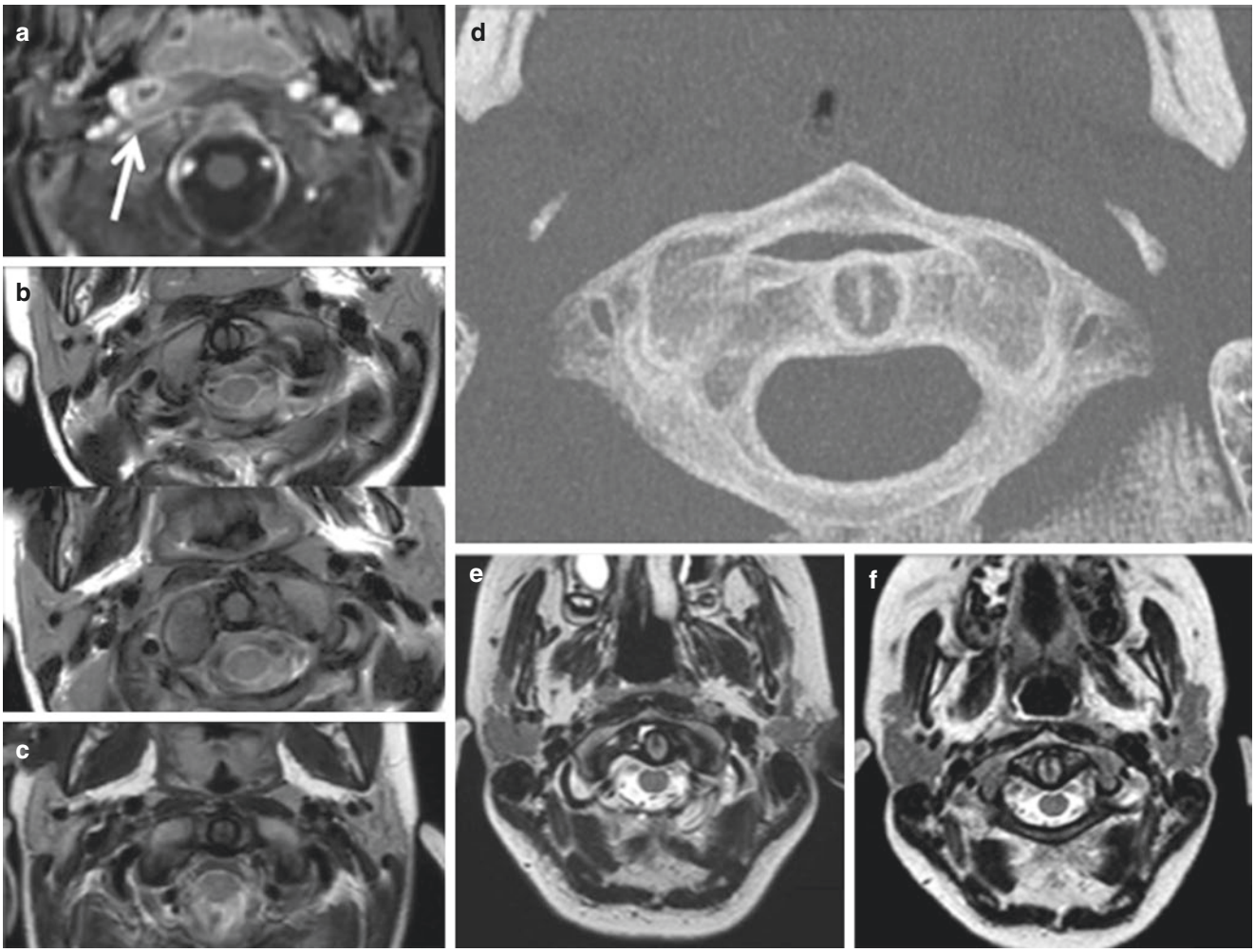


Fig. 2 Cases #1 and #2, with Fielding type I rotatory atlantoaxial subluxation. Case #1: Axial contrast-enhanced T1-weighted magnetic resonance imaging (MRI) reveals a right lymph node consolidated and packed in the parapharyngeal space with peripheral rim enhancement like a 'developing abscess' (**a**, *white arrow*). (**b**) Initial axial T2-weighted MRI shows enhancement of the alantodental interval on the left and superior articular facet of the axis. Repeated MRI, 30 days later, shows disappearance of the signal change in the atlantodental joint (**e**) and superior facet articulation of the axis (**c**). Case #2: Initial computed tomography (CT) scan with maximum-intensity projection (MIP) reconstruction shows anterior subluxation of the right C1 facet (**d**); on the same day, axial T2-weighted MRI demonstrates hyperintensity involving the C1–C2 articular capsules with intact ligaments (**e**). At 1-month follow-up, axial T2-weighted MRI shows a normal signal in the articular space and normal alignment (**f**)

tion (MIP) demonstrated Fielding type II rotatory subluxation (Fig. 3a); a three-dimensional (3D) CT reconstruction ventral view showed anterior subluxation of the right C1 facet (Fig. 3b). MRI confirmed cervical lymphadenopathy and rotatory subluxation (Fig. 3c).

The initial treatment consisted of cervical orthoses and intravenous antibiotic therapy (ceftriaxone plus metronidazole) for 7 days. After 2 months, resolution of the torticollis was observed, with disappearance of subluxation detected on a follow-up MIP 3D CT scan at 1 month (Fig. 3d, e), and the patient had complete restoration of the cervical range of motion. Follow-up MRI 10 days after collar removal revealed substantial resolution of the rotatory subluxation (Fig. 3f).

Case #4

A 12-year-old girl presented with painful torticollis, in a posture of complete cervical rotation to the left, accompanied by subtle flexion without neurological signs, 4 days after scarlet fever and tonsillitis. After 3 months her neck posture was unchanged despite initial analgesic, muscle relaxant and empirical antibiotic treatment.

Finally, the patient was admitted to hospital and MRI disclosed the presence of Fielding type II rotatory subluxation (Fig. 4a, b). An attempt at manual reduction under sedation was unsuccessful, so Gardner–Wells tongs with a gradual increase in traction weight from 4 to 6 kg were applied with

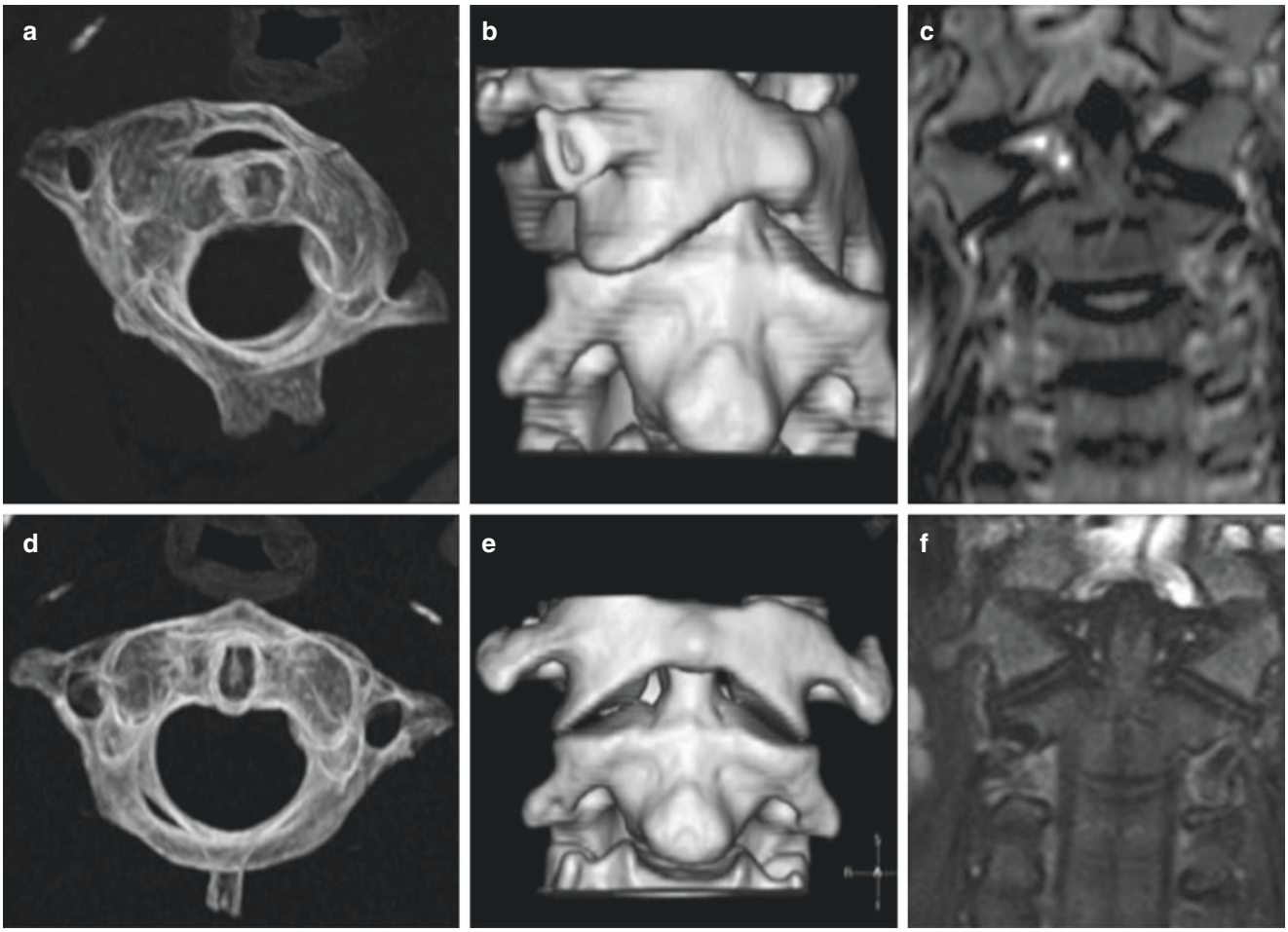


Fig. 3 Case #3, with Fielding type II rotatory atlantoaxial subluxation treated with a cervical collar. Computed tomography (CT) scan with maximum-intensity projection (MIP) (**a**) and three-dimensional reconstruction ventral view (**b**) and coronal T1-weighted magnetic resonance imaging (MRI) (**c**) demonstrate rotatory subluxation at C1–C2 with intact ligaments. Ten days after collar removal, follow-up imaging performed with identical scans and reconstruction reveals substantial resolution of the rotatory subluxation (**d**–**f**)

regular oral diazepam during the intensive care unit (ICU) stay. After 4 days a CT scan disclosed a reduction of the subluxation (Fig. 4c, d). The Gardner–Wells tongs were removed and a SOMI brace was applied for 3 months. The patient remained neurologically intact, and a CT scan of her neck performed 1 month later confirmed near-normal alignment (Fig. 4e).

Case #5

A 9-year-old boy was discharged home 1 day after uneventful adenoidectomy and bilateral tonsillar reduction. Three days after discharge, painful torticollis was observed. The patient was treated with medical therapy and a cervical collar was applied; unfortunately, no imaging was performed. Two months postoperatively, because of persistent symptomatology, a CT scan disclosed atlantoaxial rotatory subluxation (Fig. 5a).

At hospital admission the patient's neurological status was intact, with painful torticollis. Paediatric and ENT consultations showed no signs of inflammation/infection of the upper airways. Haematological and biochemical test findings were within the normal range. X-rays revealed the typical head tilt of atlantoaxial subluxation, a reduced range of motion for flexion and no mobility during extension. On CT scanning, 3D reconstructions confirmed atlantoaxial rotatory subluxation with anterolateral dislocation of C1 and a widened (6.7 mm) distance between the atlas and dens. The atlas was right rotated by 45° with respect to the axis. CT angiography identified the relationship of the vertebral artery to the posterior elements of the upper cervical vertebrae, with no anatomical anomalies (Fig. 5b).

The initial management consisted of a cervical collar and anti-inflammatory and muscle relaxant medication for

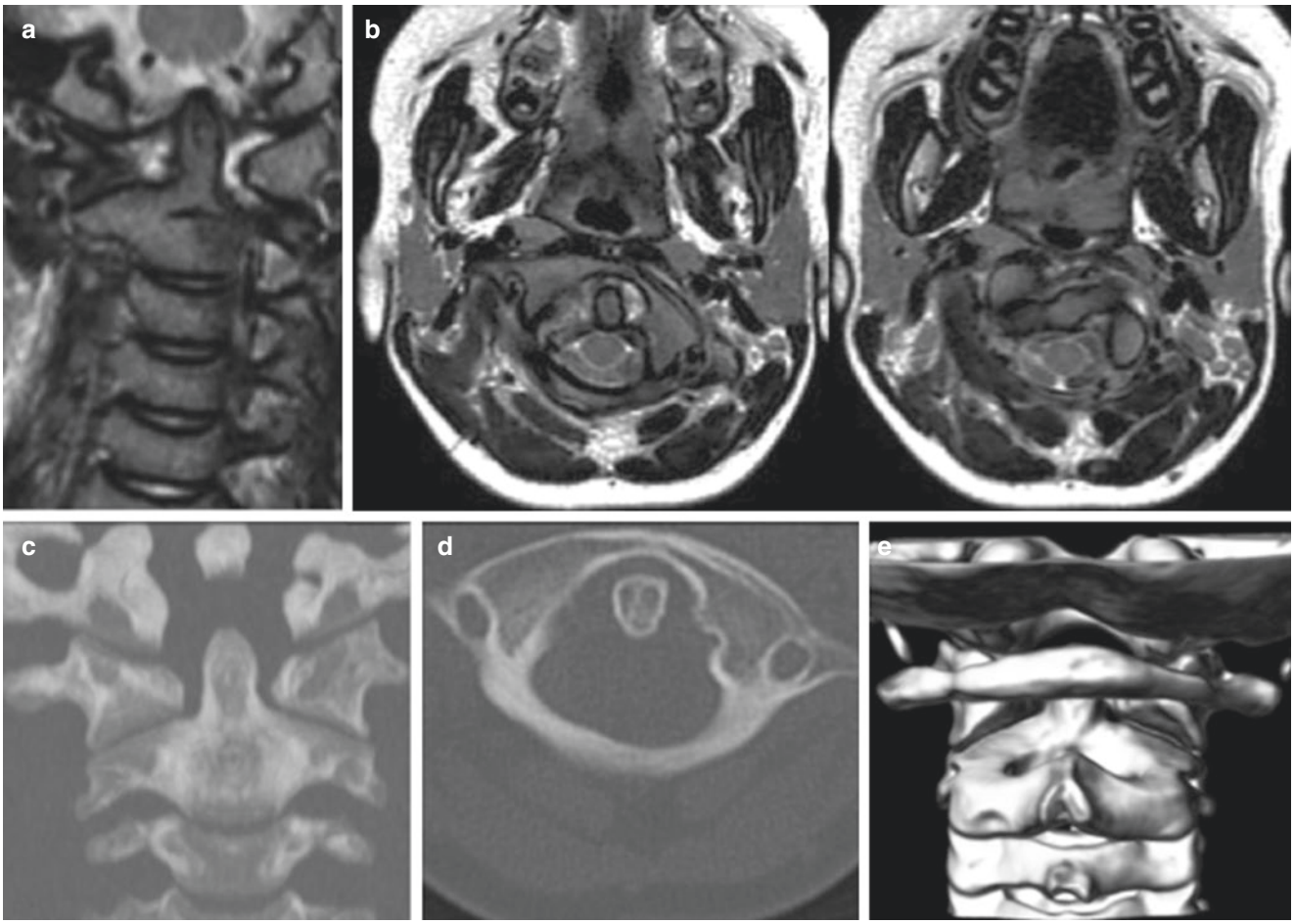


Fig. 4 Case #4, with Fielding type II rotatory atlantoaxial subluxation treated with skull traction and a sternal–occipital–mandibular immobilizer (SOMI) brace. Coronal (**a**) and axial (**b**) T2-weighted magnetic resonance imaging (MRI) shows enhancement of the alantodental interval on the right and superior articular facet of the axis. After 4 days of skull traction, a computed tomography (CT) scan with maximum-intensity projection (MIP) (**c**, **d**) shows reduction of the subluxation. At 1-month follow-up a three-dimensional reconstruction ventral view (**e**) confirms stable reduction of the rotatory subluxation at C1–C2 during SOMI brace treatment

2 weeks, with unchanged clinical and radiological features. Therefore, under general anaesthesia, reduction of the rotatory subluxation was accomplished using fluoroscopy. The patient subsequently underwent surgery with C1 lateral mass screws (Fig. 5c) and C3 pars interarticularis screws (Fig. 5e); one intralaminar screw was also placed in the left lamina of C2 (Fig. 5d). The system was assembled with lordotic-shaped titanium bars. Throughout the positioning of the patient and the surgical procedure, somatosensory evoked potential and motor evoked potential neuromonitoring was used. The postoperative period was uneventful. All preoperative symptoms resolved. The patient was mobilized after 3 days with a rigid collar. The collar was maintained for 6 weeks, during which the patient received physiotherapy. Postoperative CT scanning disclosed proper positioning of the instrumentation. At 6-month follow-up the patient had returned to his normal daily activities, except that he had a 30° limitation in neck rotation.

Discussion

Grisel's syndrome is a rare and not completely clarified pathological non-traumatic subluxation of the atlantoaxial joint, which may occur in children after nasopharyngeal inflammation or otolaryngological procedures. This syndrome is more common during childhood, usually not seen in patients aged >12 years and only rarely observed in adults.

The most accepted pathogenetic hypotheses involve spreading of inflammation to the atlantoaxial ligaments through anastomoses between lymphatic vessels and pharyngovertebral veins. A haematogenous pathway seems to conduct peripharyngeal infection to the atlantoaxial ligaments through the connections between the pharyngobasilar fascia and the pharyngovertebral veins and between the atlanto-occipital membrane and the periodontoid venous plexuses. The rotatory atlantoaxial hypermobility could be caused by laxity of transverse and alar ligaments and articular capsules

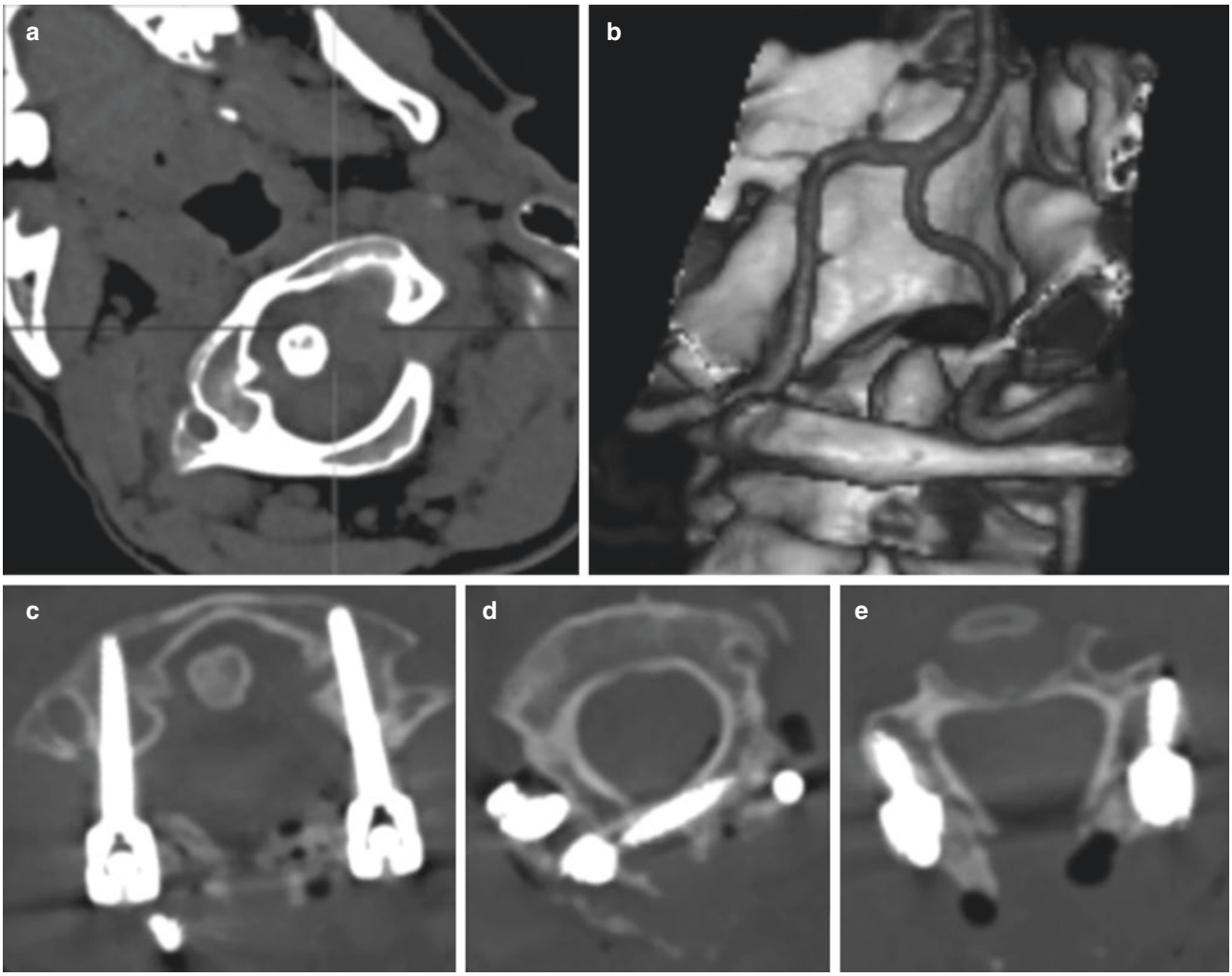


Fig. 5 Case #5, with Fielding type III rotatory atlantoaxial subluxation treated with surgical C1–C2–C3 internal fixation. An axial computed tomography (CT) scan shows atlantoaxial rotatory subluxation with anterolateral dislocation of C1 (**a**). A contrast-enhanced three-dimensional reconstruction posterior view confirms a widened atlanto-dental space and excludes associated vessel anomalies (**b**). A postoperative CT scan confirms the internal rigid fixation achieved with C1 lateral mass screws (**c**); one intralaminar screw has been placed in the left lamina of C2 (**d**), starting from the right, in order to push the spinous process of C2 to the left to counteract the rotation of the axis and C3 pars interarticularis screws (**e**)

due to inflammatory oedema [9, 31, 33–35]. Further weakening of the insertions of the transverse ligament could be caused by decalcification of C1 and C2 due to inflammation [25]. After ENT procedures an increased incidence of Grisel's syndrome has been reported. The introduction of monopolar suction electrocautery in adenoidectomy, as opposed to the use of bipolar coagulation, has been reported as an hypothetical explanation [4, 5, 36, 37].

Grisel's syndrome usually manifests with torticollis, cervical pain, head tilting, and restricted and painful neck movements. A strong suspicion of this condition could be raised by an evident dislocation of the spinous process of the axis following the side of neck rotation on manual examination. A spasm of the sterno-cleidomastoid muscle ipsilateral to the rotation and inability to turn the head beyond the midline in the direction of the side opposite the lesion are observed [4, 38].

Suspected C1–C2 subluxation can be detected simply by standard x-rays of the cervical spine [36]. Diagnosis of atlanto-axial subluxation requires a thorough neuroradiological examination including plain x-rays, CT scanning and MRI. Plain lateral x-rays of the upper cervical spine may show an increase in the atlanto-odontoid distance. The normal atlanto-odontoid distance is ≤3 mm in adults and ≤5 mm in children, while the findings at an antero-posterior trans-oral X-Ray examination include asymmetry and deletion of the C1–C2 articular surfaces, an increase in one lateral C1 mass (the one anteriorly dislocated) and a reduction in the contralateral one (the one posteriorly dislocated ipsilateral to the cervical rotation). In the

event, clear x-rays may be difficult to obtain, because of the rotation in the cervical spine and head [4].

A 3D CT scan of the craniocervical transition is the gold standard in establishing the diagnosis of atlantoaxial subluxation. The patient must keep the head in a neutral position to avoid misdiagnosis. On the axial images a measurement of the atlantodental interval (ADI) is made, with the normal measurement being 2–3 mm in adults and up to 5 mm in children. An injury of the transverse ligament results in an increase in this distance.

Usually MRI is useful to disclose abnormalities of soft tissues, lymph nodes and neural structures, such as laxity of the transverse and alar ligaments [6]. Nevertheless, in the presented series, MRI was the first radiological examination performed in cases #1 and #3. Therefore, in cases of painful torticollis, a previous ENT procedure or an ultrasound-disclosed lymphadenopathy could be a reasonable indication to perform MRI, to reduce the risk of radiation exposure in a paediatric patient. 3D CT scanning remains necessary in cases where there is doubt about the Fielding and Hawkins classification grade after atlantoaxial findings on MRI.

Several recommendations for diagnosis and treatment have been reported in the literature, but no clear guidelines are available although about 90 scientific articles have been published on this topic since 1950. Usually the Fielding and Hawkins classification of non-traumatic subluxation of the atlantoaxial joint drives the choice between conservative and surgical treatment, assuming that the degree of subluxation and instability increase from types I to IV, according to the displacement of the dens and the asymmetry of the atlantoaxial joint [9, 26].

Types I and II are the most common subluxations, usually with intact neurological status; rotatory dislocation in the absence of anterior displacement of the atlas (type I) or minimal (<5 mm) anterior displacement of the atlas (type II) are observed. The first choice of treatment may include antibiotic, muscle relaxant and anti-inflammatory medications for 1 week and application of a cervical collar for 4 weeks [6, 31, 39]. In some patients, a first manual reduction manoeuvre under deep sedation has been reported. The patient rests in a supine position, the operator's left hand fixes the occiput–occipital protuberance while the right hand fixes the patient's chin, and gentle stretching of the patient's neck along with a gentle rotary manoeuvre is applied to the side opposite the rotatory deformity. During this manoeuvre, an assistant should fix the patient's shoulders to prevent dangerous movements [36, 40]. After reduction these cases are usually treated with oral administration of antibiotic, muscle relaxant and anti-inflammatory medications for 1 week, in combination with immobilization of the cervical spine with a SOMI brace for 4–6 weeks. Cases #1 and #2 in the current series were treated with 4 weeks of neck immobilization with a cervical collar because prompt diagnosis allowed detection of Fielding type I rotatory subluxation <15 days after the onset of painful torticollis. In case #3 the delayed diagnosis (after 60 days) could have been the cause of the Fielding type II rotatory subluxation found on MRI, with a need for more prolonged application of a cervical collar for 8 weeks. In case #4, manual reduction of the subluxation under sedation was unsuccessful; therefore, treatment with skull traction for 4 days and a SOMI brace were applied. In this case, too, there was a delay in radiological diagnosis of Grisel's syndrome, which was not confirmed until 3 months after the onset of torticollis.

Type III presents with a rotatory dislocation with >5 mm anterior displacement of the atlas, while type IV is associated with posterior displacement of the atlas. Patients with Fielding type III and IV subluxations generally need bed rest with cervical traction, followed by a period of neck immobilization in a halo brace for up to 12 weeks. Types III and IV, although rare, are often associated with spinal cord compression, are usually associated with neurological deficits—which also occur in up to 15% of cases treated with surgical fusion [41]—and can have fatal consequences. Because of the rotatory dislocation with posterior displacement of the atlas, type IV is primarily treated with surgical C1–C2 fusion [7, 9, 18, 26, 42, 43]. Case #5 in this series, which was classified as type III, had a 56-day delay in diagnosis despite persistent painful torticollis. The ADI was 6.7 mm, so to avoid the need for prolonged use of a postoperative halo vest or SOMI brace, C1–C2 or C1–C2–C3 fixation using lateral masses and pars interarticularis screws was recommended [27]. As Spennato et al. reported in their previous publication on this case, this technique is associated with high fusion rates and immediate spinal stability in all planes of the atlantoaxial complex; therefore, it is preferred to semi-rigid constructs using interlaminar or interspinous wires and cables [25]. In this case, the authors adopted C3 inclusion in the construct as there was no safe and effective option for placing bilateral translaminar screws, because of the C2 anatomical features. Pedicular or pars interarticularis screws were considered more risky and technically more demanding than the translaminar screw technique, as suggested by Wright [44].

In cases in the literature where there was a delay in diagnosis of >3 weeks after the onset of symptoms, failure of conservative treatment and a high risk of recurrence have been reported [4, 33, 38, 45, 46]. Prompt diagnosis is essential for safe and effective treatment. More recently a concept of management has been proposed where surgical intervention could be avoided in cases of Fielding type III subluxation, by use of manual repositioning and application of a Minerva cast under general anaesthesia. This approach was used in two cases reported by Pilge et al. and in one case reported by Viscone et al. [9, 39]. Those authors suggest that data from a more extensive patient collection are needed to confirm the safety and efficacy of this treatment, and that the technique needs to be performed by an experienced specialist at a specialized centre.

Conclusion

In paediatric patients with painful torticollis following ear, nose and throat procedures or nasopharyngeal inflammation, Grisel's syndrome should always be suspected. A prompt diagnosis can be confirmed simply with an initial standard cervical x-ray. Magnetic resonance imaging is indicated to detect the deep and superficial lymph node status and to define the atlantoaxial rotatory subluxation. In cases with higher-grade instability, a three-dimensional computed tomography scan of the craniocervical transition will establish the patient's Fielding and Hawkins classification and appropriate management. Early treatment of Grisel's syndrome can avoid neurological complications and surgical intervention.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Odontoid Fusion

Corey T. Walker and Volker K. H. Sonntag

Abstract Anterior odontoid screw fixation allows for the internal fixation of unstable odontoid fractures with low morbidity, good fusion rates, and preservation of the atlantoaxial range of motion when applied in appropriate clinical cases. Advances in surgical techniques have allowed for safer, more minimally invasive approaches that reduce the risk of injury to vital prevertebral structures and minimize soft tissue retraction. Moreover, improvements in surgical image guidance technology for spinal surgery that have been applied to odontoid screw placement have helped improve surgeon confidence about exact screw trajectories. In this chapter, we review traditional screw placement techniques and highlight the trends in technical improvements that improve the safety and efficacy of these procedures.

Keywords Anterior screw fixation · Fusion · Odontoid fracture · Odontoid fusion · Rostral fractures

Introduction

Anterior odontoid screw fixation represents a method for the surgical fusion of specific odontoid fractures. Although many stable type fractures can be treated with cervical orthosis, anterior screw fixation promotes improved fusion and outcomes in acute, unstable Type II and rostral Type III fractures, as classified originally in 1974 by Anderson and D'Alonzo [1]. In comparison to posterior fusion techniques, anterior fixation allows for the preservation of the large range of motion that occurs at C1–C2. It also decreases patient morbidity because it requires less muscular dissection than required by posterior approaches. Over the past few decades, numerous reports in the neurosurgical literature have helped to establish the indications and contraindications for this procedure, resulting in improved selection of patients. Thus, anterior odontoid fusion has become a safe and well-accepted treatment when it is applied appropriately and with expert surgical skill. The latest techniques build upon the traditional techniques, which will be discussed in what follows, and they also improve the safety of the procedure and reduce morbidity in patients undergoing it.

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Materials and Methods

Traditional Surgical Technique

The traditional surgical technique for anterior odontoid fusion requires the use of simultaneous intraoperative anterior-posterior (AP) open-mouth and lateral fluoroscopy to appropriately guide the screw trajectory. The patient is placed in the supine position with the neck extended. Appropriate extension performed under fluoroscopy allows the trajectory of the screw to be toward the tip of the dens (Fig. 1a). The use of radiolucent materials in the mouth and around the head prevents obscuration of the working field.

Surgical exposure is completed by making a transverse incision at the level of C4–5 to allow for a cranially directed screw trajectory (Fig. 1b). After subplatysmal dissection down to the prevertebral fascial plane is complete, cephalad dissection is performed along the anterior longitudinal ligament up to the level of C2. Retractors are placed within the path, and the longus colli muscles are coagulated at their medial insertions and elevated laterally to expose the underlying C2–3 vertebral space (Fig. 1c). In some cases, a curette can be used to rasp the bony surfaces at the fracture site to help remove soft tissue and instigate bony fusion.

Although some groups have suggested that two screws may be placed, biomechanical research has shown that one screw is sufficient to provide the necessary fixation [2].

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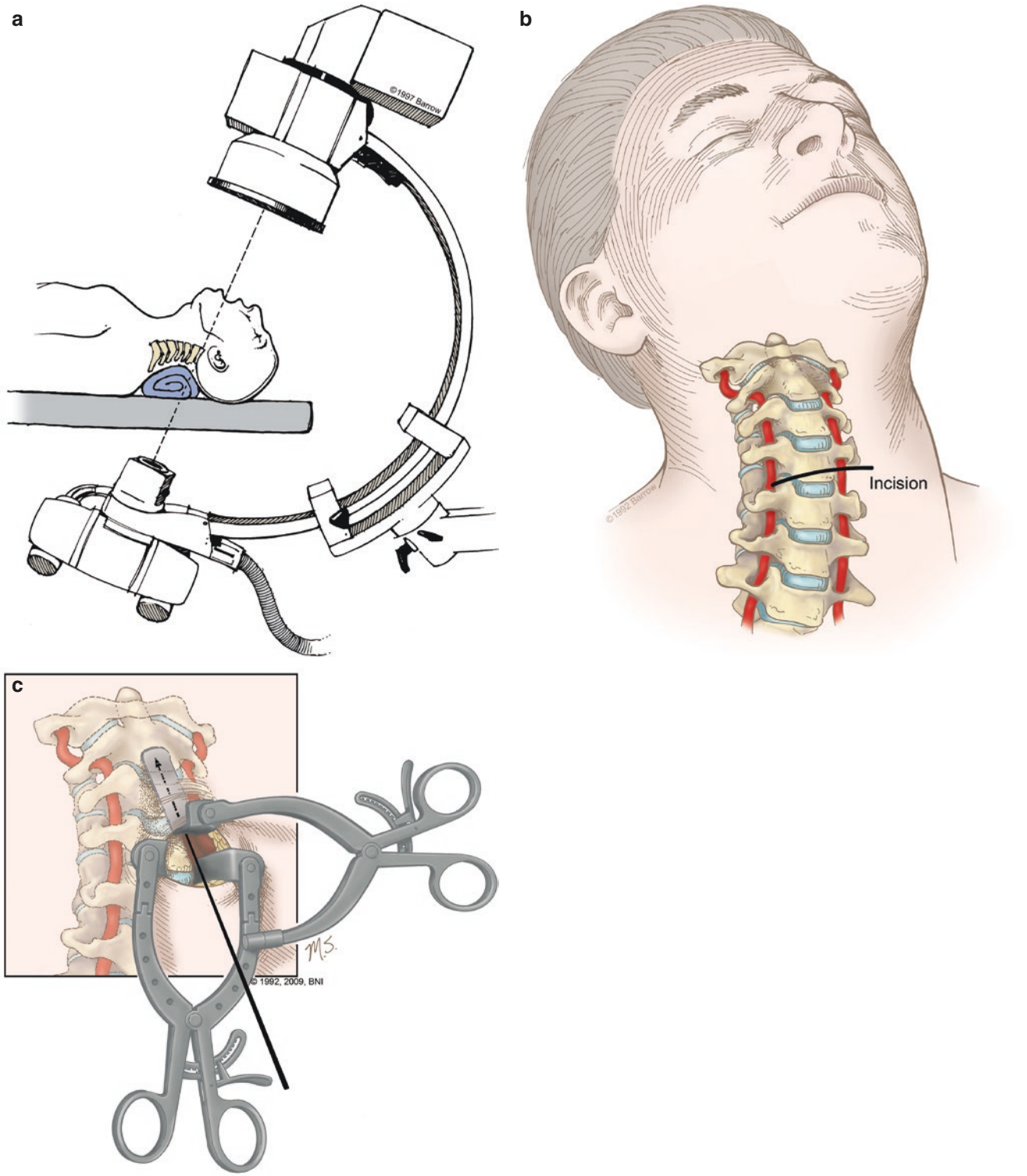

Fig. 1 Traditional surgical technique for odontoid fusion. (**a**) The anterior-posterior (A-P) orientation of the C-arm creates an open-mouth view of the odontoid process. (**b**) Incision over the C5 level with dissection angled toward the C2–3 disc space entry point, with (**c**) the horizontal trajectory aimed at the tip of the dens. In this image, a traditional Apfelbaum retractor is used to hold back the soft tissue. Used with permission from Barrow Neurological Institute, Phoenix, Arizona

In the single-screw technique, a midline entry point is designated. A trough is created in the superior ventral body of C3, and the nearby annulus of the C2–3 disc space is partially removed to allow screw entry into the inferior end plate of the C2 body. A trajectory nearly parallel to the cranial–caudal plane is optimal to ensure that the screw enters the tip, which also reduces the risk for spinal cord injury. The trajectory should be confirmed with intraoperative fluoroscopy. With cannulated screw systems, a Kirschner wire (K-wire) is drilled into the trajectory to guide the placement of hollow screws and tools. It is drilled into C2 until its tip engages the tip of the fractured dens. The K-wire can then be removed and measured to determine the length of the screw. A second K-wire can also be used to measure screw length, if desired. A self-tapping 3.5- or 4.0-mm screw can then be inserted directly over the K-wire. If there is a gap between the fractured fragments, the screw measurement can be adjusted to compensate for the estimated reduction that will occur once the cannulated lag screw is in place. The screw is inserted carefully, with the screw head flush against the C2 body. Screw ends that protrude into the interspace are at risk of becoming loosened or causing fracture. After the screw is in place, the K-wire can be removed.

Minimally Invasive Techniques

Tubular Retractor Systems

During the development of traditional techniques for anterior odontoid fusion, multiple adaptations have been made to retractor systems to prevent soft tissue injury during drilling maneuvers. Apfelbaum and colleagues helped design an anterior cervical retractor with a detachable third blade capable of supporting the posterior wall of the pharynx (Fig. 1c). Tubular drill-protecting guides have also been devised with teeth that can be secured to the targeted entry point. This provides protection of soft tissue by preventing the drill bit teeth from catching encroaching soft tissue around the retractor blades.

With the advent of tubular retractor systems for use in minimally invasive lumbar surgery, our group proposed the application of the same tubular systems for odontoid fusion [3]. An 18-mm tubular retractor (METRx System; Medtronic Sofamor Danek USA, Inc., Memphis, TN) was developed using the same basic principles as those described earlier. A flexible arm-mounting system is attached to the bed rail contralaterally to the surgeon (Fig. 2a). Dissection is performed at the level of the C5 vertebral body, with handheld retractors being used to expose upward to the C2–3 disc space. Then a 16.8-mm dilator tip is placed at the entry point with the desired trajectory as determined using standard biplanar fluoroscopy. After this step is completed, the tubular retractor is placed over the dilator and secured to the flexible arm before removal of the dilator. Final positioning of the tube is confirmed using fluoroscopy, which allows small adjustments of the retractor arm to be made without having to reinsert the dilator. Upon the exposure of this surgical corridor, the surgeon can begin the K-wire insertion and screw placement as described earlier. Subsequent modifications to the tubular retractor include beveling of the distal end of the tube to allow for a flush interface between the retractor and the ventral surface of the vertebral bodies to which it is docked [4] (Fig. 2b, c). A tubular light source has also been added to the tubular retractor to allow better visualization of the working surface.

Percutaneous Screw Placement

In 1999, Kazan et al. [5] described the first percutaneous approach for the placement of anterior odontoid screws, which they tested in cadaveric experiments. This technique was subsequently implemented in human patients with good safety and efficacy [6, 7]. As with the traditional techniques, percutaneous placement relies heavily on biplanar fluoroscopy. With this technique, the entry point is also made at the level of the C5 vertebral body. Most authors suggest injection of 25–30 cm^3 of saline solution into the fascial plane medial to the neurovascular bundle to help separate tissue during placement of the tubular retractor. A small (<1 cm) incision is then made, with the platysmal opening and dissection medial to the sternocleidomastoid muscle. Although the exact techniques may differ by surgeon preference, dissection with a blunt K-wire or Jamshidi needle is used to gain access to the prevertebral space, and the dissection is then directed upward to the C2–3 disc space under fluoroscopic guidance. A telescopic dilating tube system is placed, and the K-wire is positioned along the desired trajectory under fluoroscopic guidance (Fig. 3a–d). A cannulated lag screw is then driven over the K-wire across the fracture. Careful attention to the K-wire is required at all times, particularly during screw drilling, to prevent unwanted advancement toward the brainstem. After placement of the cannulated lag screw, the guidewire is removed.

A limitation of this technique is that the positioning of the K-wire in the appropriate trajectory can be difficult using fluoroscopy alone without direct visualization of the entry

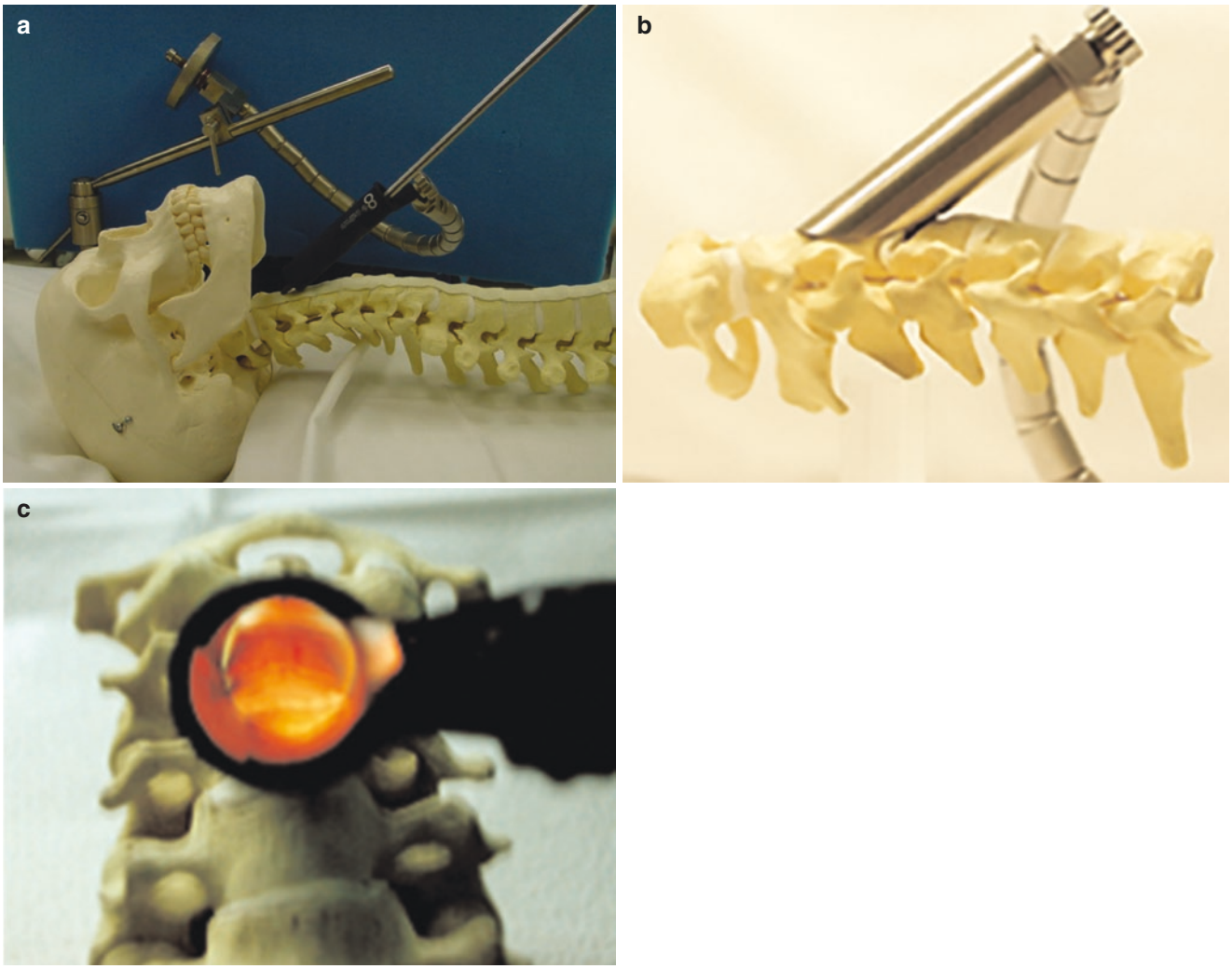

Fig. 2 Tubular retractor system for odontoid fusion. (**a**) Proper orientation of METRx tubular retractor (Medtronic Sofamor Danek USA, Inc., Memphis, TN, USA) for screw placement, with arm rigidly fixed to table. Reprinted with permission from Hott et al.: A new table-fixed retractor for anterior odontoid screw fixation: technical note (3), 98:118–120, 2003. Permission granted by J Neurosurg Spine [3]. (**b**) Modified tubular retractor designed specifically for odontoid screw placement. (**c**) Surgeon's view demonstrating incorporated light source, which improves visualization of entry point. Reprinted with permission from Shalayev et al.: Retrospective analysis and modifications of retractor systems for anterior odontoid screw fixation. 16 (1):Article 14, 2004. Permission granted by Neurosurg Focus [4]

point. Repositioning an undesirable K-wire hole can be particularly difficult, because doing so may compromise screw placement. Some authors have proposed using a two-hole guide tube, so that a second parallel K-wire can be placed at a better trajectory if an inappropriate trajectory is obtained initially [8]. However, to obtain acceptable minimally invasive screw placement, the surgeon must be thoroughly familiar with this surgical methodology.

Intraoperative Navigation

As in many areas of spinal surgery, intraoperative navigation for screw placement is increasingly being used during anterior odontoid fusion. Computed tomography (CT)–based guidance allows for three-dimensional visualization of the proposed entry points and trajectories based on reference points affixed to a radiolucent Mayfield frame (Fig. 3e, f). As a result, many proponents believe that the use of intraoperative CT enhances safety and improves outcomes. Compared to the use of biplanar fluoroscopy, the use of CT for intraoperative navigation decreases radiation exposure to the surgeon and operating room staff at the cost of increased exposure to the patient. In the largest comparison of the two techniques, our group found that navigation cases required more operating room preparatory time but actually led to decreased operation duration and higher rates of good screw positioning with fusion across the fracture

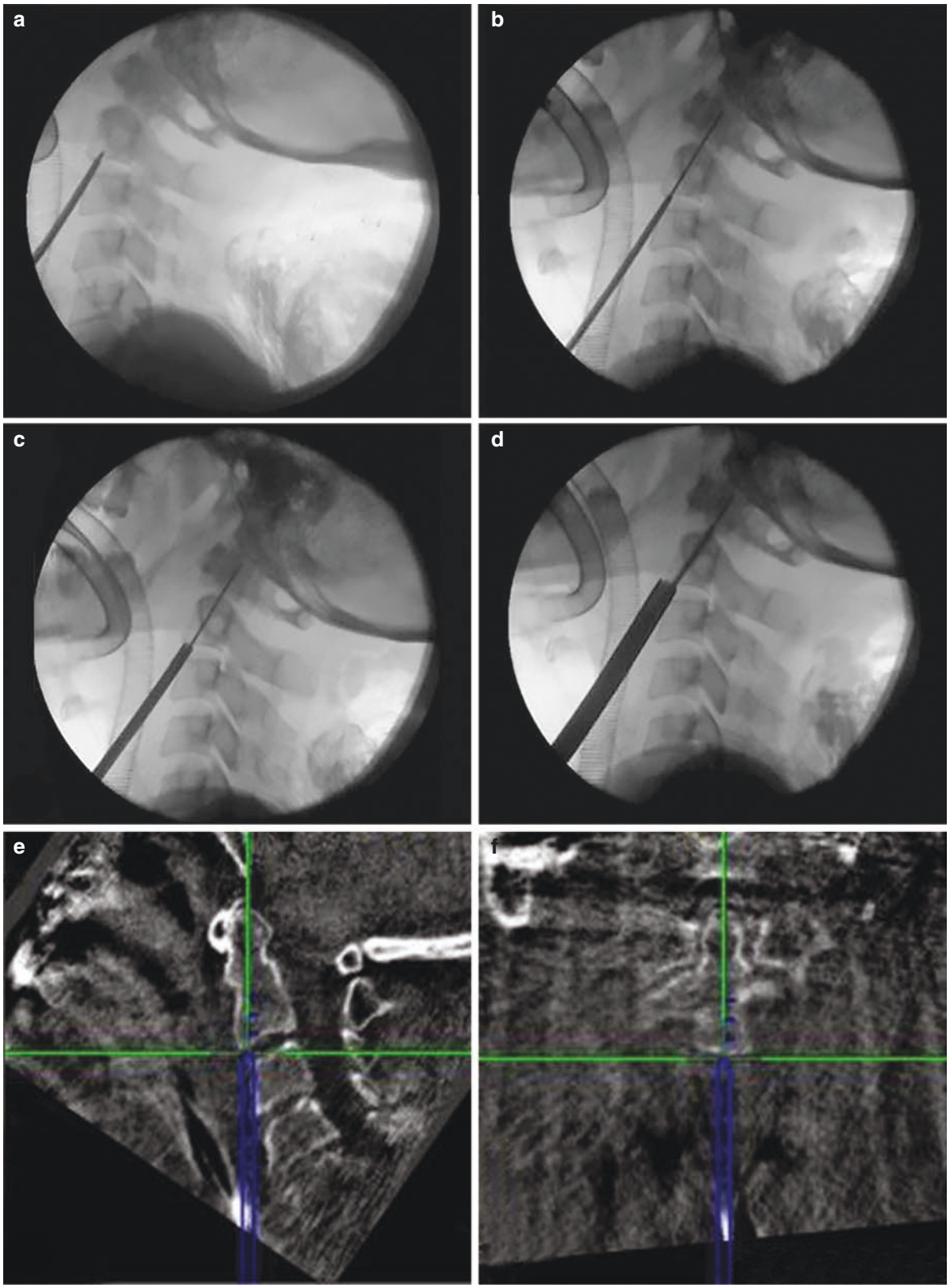

Fig. 3 Image-guided percutaneous odontoid screw placement. Fluoroscopic images show (**a**) a Jamshidi needle placed in the inferior C2 body, (**b**) the K-wire driven into the odontoid, (**c**) progressive dilator tubes placed over the K-wire, and (**d**) screw placement across the fracture from within the largest tube. Intraoperative sagittal (**e**) and coronal (**f**) computed tomography reconstructions also provide surgical image guidance. Parts **a**–**d** reprinted with permission from Sucu et al.: Percutaneous anterior odontoid screw fixation. 51:106–108, 2008. Permission granted by Minim Invasive Neurosurg [7]. Parts **e**–**f** used with permission from Barrow Neurological Institute, Phoenix, Arizona

site [9]. Still, many surgeons will use adjunctive lateral fluoroscopy during drilling of the screw to confirm trajectory and depth.

Results and Discussion

Odontoid fixation is a safe and efficacious method for the fusion of specific types of odontoid fractures in select patients when it is performed by surgeons who are comfortable with the procedure. Recent trends in fine-tuning and optimizing this procedure have been aimed at increasing safety and minimizing morbidity. The evolution of tubular retractor systems and percutaneous methods with dilating tubes allows for smaller incisions, less muscle dissection, and avoidance of damage to surrounding tissue. For surgeons less familiar with the procedure, the addition of CT-based navigation likely improves operative planning, increases the accuracy of screw placement, and facilitates determination of trajectory. Armed with these tools, surgeons should find that anterior screw fixation of odontoid fractures yields excellent fusion results with relatively low risk to the patient.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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CVJ Trauma

Posttraumatic Anatomical Injuries of the Craniovertebral Junction and Treatment Implications: Part I

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Keywords Craniovertebral junction · Anatomy · Trauma Treatment

Introduction

Traumas involving the craniovertebral junction (CVJ) often cause structural modifications in the normal anatomy of the involved areas. Their correct identification is pivotal in clinical practice, since it significantly influences choices between conservative treatment and surgery in the therapeutic decision-making process.

The upper cervical spine is defined by the two most cephalad cervical vertebrae: C1 (the atlas) and C2 (the axis). Both the anatomical conformation and the range of motion (ROM) of this segment are considerably different from those of the subaxial counterpart of the cervical spine. The occipital condyles articulate with the superior surface of the lateral masses of C1. The atlanto-occipital articular unit ensures the greatest proportion of flexion and extension movements of the head.

The main feature of C1 is the absence of an identifiable vertebral body, which is substituted by the odontoid process of the axis. Most of the lateral rotation of the neck actually occurs at the C1–C2 junction; the remaining motion of the cervical spine is distributed among the subaxial spine vertebral motion segments as a fractional amount (~7%) per level and is less in total than the C1–C2 lateral rotation.

This area of the upper cervical spine is extremely mobile, and its stability depends on ligamentous structures. This means that even in cases in which there is no evidence of a fracture of vertebral bony structures, disruption of the ligamentous components may cause vertebral instability and may be sufficient to sustain pathological conditions such as atlanto-occipital dislocation and atlantoaxial subluxation. In fact, radiological investigations should always be performed, especially in comatose patients, to exclude the presence of ligamentous vertebral instability.

Ligaments provide the bulk of stabilization. The anterior longitudinal ligament extends cranially as the anterior atlantoaxial and atlanto-occipital ligaments. The posterior longitudinal ligament continues cranially as the tectorial membrane. The posterior ligamentous complex is composed of the ligamentum flavum and interspinous ligaments. The cephalic extension of the ligamentum flavum is the posterior atlanto-occipital ligament, whereas the cephalic extension of the interspinous ligament is the ligamentum nuchae. The transverse ligament of the atlas holds the dens against the anterior arch of C1; vertically oriented bands of this ligament extend to the anterior foramen magnum and to the posterior body of the axis to form the cruciform ligaments. The alar ligaments extend from the odontoid process to the lateral margins of the foramen magnum, limiting lateral rotation of the skull. The apical ligament, which extends from the tip of the odontoid process to the inner surface of the clivus, adds little structural support (Fig. 1).

Before analysing the posttraumatic anatomical modifications of bony and ligamentous elements of the CVJ, one should remember that some pathological conditions involve the paediatric age group exclusively. This is a consequence of some peculiar anatomical characteristics of the paediatric population that are not identifiable in adults.

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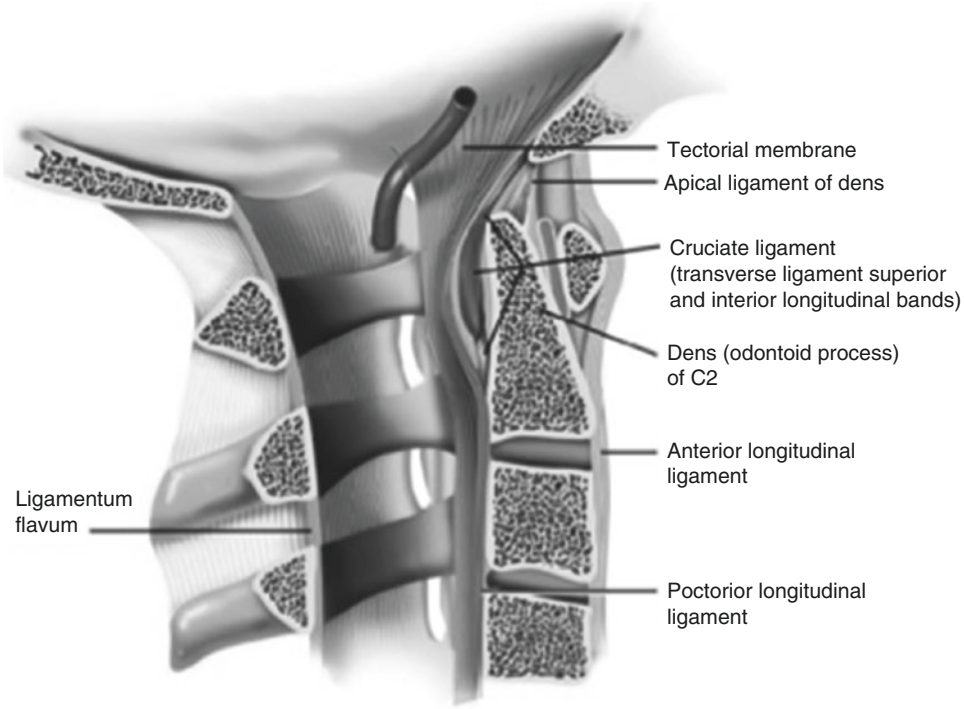

Fig. 1 Relevant ligaments of the craniovertebral junction described in the text

The laxity of the paediatric CVJ ligamentous complex explains why upper cervical spine injuries are so common in the paediatric age group. For the same reason, children are more likely to be subject to spinal cord injuries without radiological abnormalities (SCIWORA).

Computed tomography (CT) has been proved to be most accurate and cost effective than conventional radiography in diagnosis of CVJ injury in moderate-risk to high-risk adults [1–3], although this comes at a cost of increased radiation exposure and an increased risk of thyroid cancer in children. CT scans were thought to be able to improve the efficiency of emergency departments, reducing the time spent in establishing the diagnosis; however, Adelgais et al. [4] showed increased radiation exposure and no increase in the efficiency of management with use of CT scanning rather than conventional radiography in children.

Plain radiography is commonly used in diagnosis of CVJ injuries in both adult and paediatric populations because it is portable, is readily available and carries a relatively low radiation dose. However, open-mouth odontoid images are difficult to acquire in young children, who have a short neck, often wear a collar and are not always able to open their mouth on command.

Lateral projections are more useful than anteroposterior projections for diagnosis of CVJ injuries. Silva et al. [5] compared the sensitivity of the lateral image alone with that of additional images, using multiple-detector CT (MDCT) scanning as a reference standard, and found that the additional images did not increase the sensitivity (which was 73% [95% confidence interval 50–89%] with the lateral view alone) and marginally decreased the specificity (from 92% to 91%). The sensitivity of 73% was considered barely acceptable for a screening study. Other studies have concluded that complementary radiographic projections, such as the transoral projection for odontoid fractures and oblique projections, do not improve the sensitivity of conventional ones. Lateral projections along with anteroposterior projections have been demonstrated to identify approximately 87% of CVJ fractures in the paediatric population [6].

Several measurements—including the Powers ratio, atlantodental interval, atlanto-occipital interval, basion–axial interval and basion–dental interval—have been used in an adult population with good results in terms of sensitivity and specificity. However, these parameters seem to be inapplicable in children because of age-dependent anatomical peculiarities and variations in the ossification process that make these measurements inaccurate [7, 8].

Moreover, values considered physiological in radiographic measurements of the anatomical relationship of the CVJ cannot be applied to MDCT scans. In fact, normal radiographic parameter values have been found to be greater than those measured on CT scans. As a consequence, application of radiographic parameters to CT scans would result in missed diagnosis of a variable percentage of CVJ injuries [8]. However, the capacity to detect cervical fractures is significantly greater with CT scanning than with radiography [7, 9, 10].

Magnetic resonance imaging (MRI) should be used to detect soft tissue injuries (intervertebral disc, ligamentous complex, muscle and joint capsule injuries) and involvement of the spinal cord. MRI plays a pivotal role in implementation of MDCT scan information in surgical planning, as well as in clinical prognostication [11].

This paper is Part I of a two-part report. In the following sections and in Part II of the report, bony and ligamentous alterations of the CVJ are individually described for each component of the region: the occipital bone, atlas and axis.

The Condylar Portion of the Occipital Bone, Atlanto-occipital Dissociation and the Atlanto-occipital Joint Space (Condyle–C1 Interval)

Non-osseous injuries of the craniocervical junction are considerably more common in children than in adults. MRI is crucial in the evaluation process; many cases of juvenile CVJ injuries that are negative on CT scanning are identified with MRI [12, 13]. The injuries range from minor soft tissue injury to cervicomedullary contusion. The disproportionate head size, poor muscle tone, lax ligaments and incompletely developed articulations make the craniocervical junction especially vulnerable in infants and young children (Figs. 2 and 3).

In the guidelines for the management of acute cervical spine and spinal cord injuries, published in *Neurosurgery* in 2013, condyle–C1 interval (CCI) measurements were given a level I recommendation for diagnosis of atlanto-occipital dissociation (AOD) in the paediatric population (level III in adults), with sensitivity and specificity of 100% [14].

The spectrum of CVJ abnormalities detected with MRI was described by Sun et al. in 2000 [15] and includes ligamentous injuries followed by muscular trauma, extra-axial haemorrhage, fractures and spinal cord lesions.

AOD injuries include both atlanto-occipital dislocation and atlanto-occipital subluxation. The mortality rate reported for dislocation injuries is significantly higher than that reported for subluxation [8]. For this reason, much attention has been paid to this region to identify patients who have had subluxation injuries so they can be appropriately managed.

On MDCT multiplanar reconstruction (MPR) images, magnification is negligible and direct visualization of the joint spaces is possible. Specifically, on MPR images, evaluation of the atlanto-occipital joint can be performed accurately. Therefore, assessment of the relationship between the occipital condyle and the lateral mass of the atlas represents the most important method in the detection of AOD.

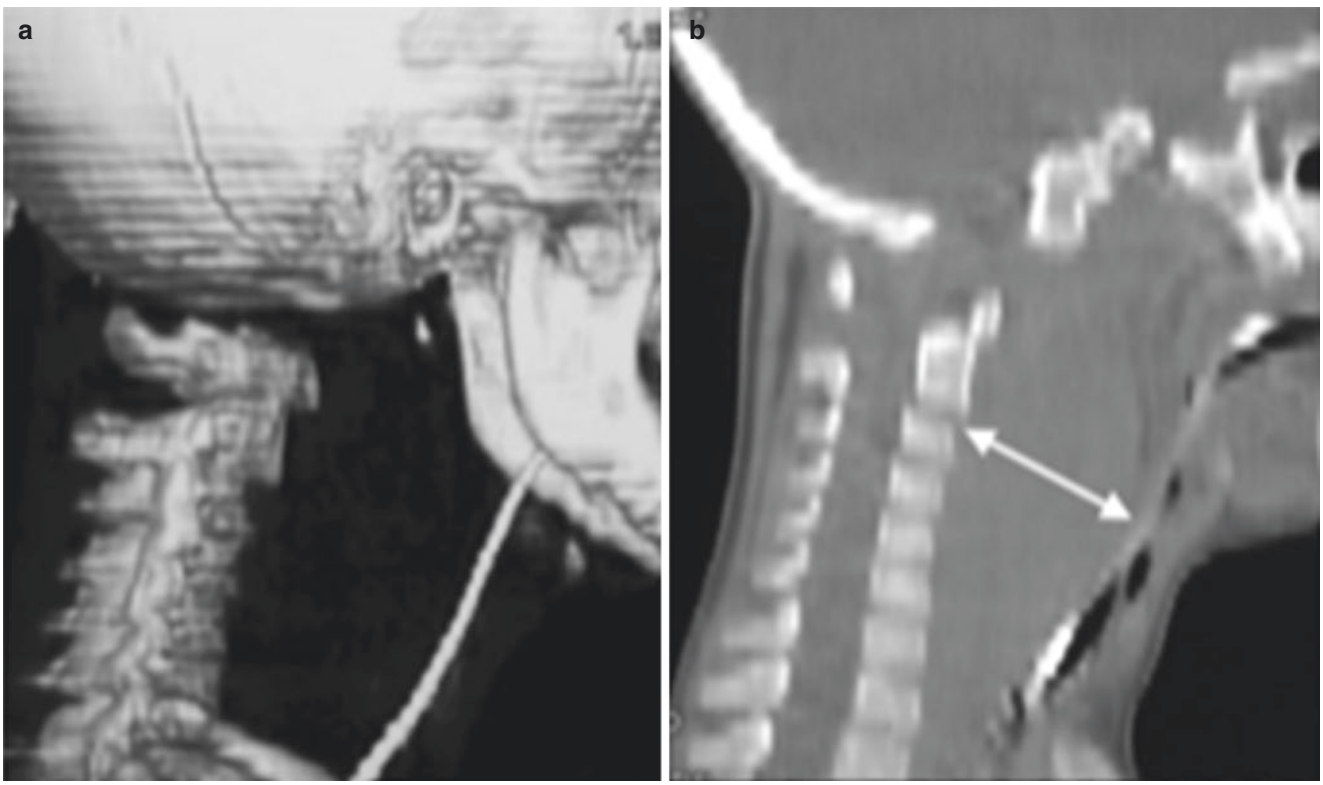

Fig. 2 (**a**) Three-dimensional computed tomography (CT) scan of a young child presenting with a disconnection in C0–C1. (**b**) The same patient, showing swelling of retropharyngeal tissue (*white arrow*), common in this type of traumatic injury

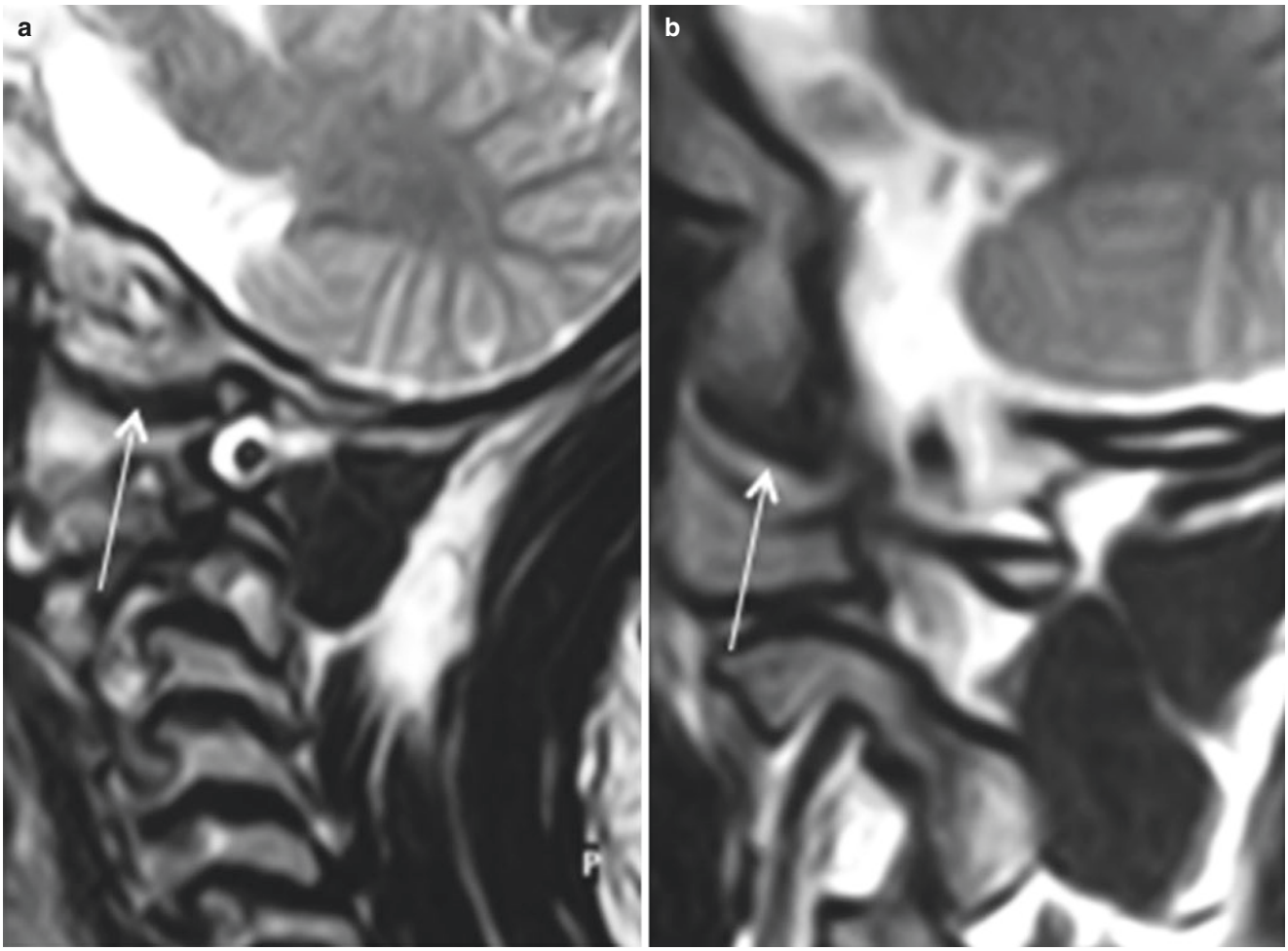

Fig. 3 Sagittal T2-weighted craniocervical magnetic resonance imaging (MRI). (**a**) Enlarged C0–C1 facet (*white arrow*). (**b**) Effusion in the facet, indicating instability of the atlanto-occipital junction (*white arrow*)

In one study of retroclival collections following child abuse [16], the most common type of retroclival collection was located in the subdural compartment (48%). Less common types were retroclival epidural collections (14%) or combined retroclival epidural and subdural collections (10%). More than one quarter (28%) of collections were categorized as indeterminate for the following reasons: the retroclival collection was identified on CT scanning only and no brain MRI was performed; CT scanning showed that the retroclival collection had resolved prior to MRI being performed; and the MRI detail/available sequences did not permit reliable identification of the tectorial membrane and therefore limited the categorization of the retroclival collection.

Tectorial membrane disruption has been reported to be one of the most important factors resulting in formation of a retroclival haematoma (REH). REHs are rare, and their description in the literature is uncommon. They are more commonly diagnosed in the paediatric population; their occurrence in adults is an exceptional event [17]. Most paediatric traumatic REHs have been observed in children harbouring arteriovenous malformations.

The tectorial membrane, transverse ligament and alar ligaments play an important role in stabilizing the craniocervical junction. The tectorial membrane—a superior extension of the posterior longitudinal ligament—extends to the anterior margin of the foramen magnum and covers the caudal portion of the clivus. The tectorial membrane is the major craniocervical stabilizer and plays a significant role in the pathogenesis of REHs.

Several mechanisms have been proposed in order to explain the pathogenesis of clival epidural haematomas. They include stripping of the tectorial membrane from the surface of the clivus, caused by a hyperextension injury, resulting in damage of the membrane itself and bleeding from the injured dura; traumatic disruption of the local vasculature such as the meningohypophyseal trunk or the basilar plexus; and clival fracture or diastasis of the spheno-occipital synchondrosis with dural bleeding [18–20]. Clival subdural haematomas have been rarely reported, and the source of bleeding has not

been clearly understood because of the paucity of vessels identifiable in the clival subdural space [21, 22]. Three different mechanisms have been proposed regarding the unusual occurrence of a subdural haematoma in the clival region:

1. Collection of fluid in the subdural space secondary to dural injury and consequent bleeding.
2. Traumatic arachnoid tear allowing cerebrospinal fluid (CSF) leakage in the subdural space. This would explain the three cases of CSF-intensity subdural collections reported in the aforementioned study [16].
3. Redistribution of subdural fluid/blood from subdural haematomas located along the occipital squama or middle cranial fossa.

The postulated mechanism for tectorial membrane injury is sagittal dislocation of the odontoid process associated with disruption of the transverse ligament, causing detachment of the tectorial membrane from the clivus. In children, the dura mater and tectorial membrane are not firmly attached to the skull; injuries causing stripping of the tectorial membrane could result in traction of the adjacent vascular structures such as the basilar venous plexus and the dorsal meningeal branch of the meningohypophyseal trunk. This may result in collection of blood in the retroclival epidural space. Indirect features of tectorial membrane injuries are stretching, detachment and elevation of the membrane from the clivus, with simultaneous epidural fluid collection extending to the apex of the dens or subdural collection in the form of blood or hygromatous collection [16].

Since a CT scan is not able to clearly identify soft tissues, diagnosis of tectorial membrane injuries with elevation of the membrane itself by the collection of an REH must be performed using MRI, which has higher sensitivity for diagnosis of tectorial membrane injury and is also able to differentiate stretching from disruption of the membrane itself. The paucity of studies regarding traumatic REH and tectorial membrane injury suggests, however, that these lesions have been underdiagnosed to date.

As mentioned above, REHs are rare in the adult population. However some known conditions facilitate the occurrence of this pathological condition; they include: haemophilia, explosive pituitary apoplexy, posterior cranial fossa decompressive craniectomy for cerebellar infarction, and treatment with anticoagulants [17].

Evidence of an intact tectorial ligament, absence of facet fluid and a CCI <2.5 mm on MDCT scanning are signs of stability and suggest that a more conservative therapeutic strategy should be pursued.

In the guidelines for the management of acute cervical spine and spinal cord injuries [14], surgical treatment of AOD with internal fixation and fusion was given a level III recommendation. The most common surgical procedures performed are occipitocervical fixation and, more recently, condylar cervical fixation.

Traction is not recommended in the management of patients with AOD, since it is associated with a 10% risk of neurological deterioration.

Osseous Condylar Lesions

Hanson [23] estimated that the frequency of occipital condyle fracture in seriously injured patients is as high as one or two fractures per 1000 patients.

The classification system used for condylar fracture is that described by Anderson and Montesano [24]. This classification recognizes three different types of fractures: type I comminuted, type II extended to the skull base and type III avulsed. Type III injuries may be associated with disruption of the alar ligaments and tectorial membrane, and may result in craniocervical dissociation. In an extensive retrospective series of 107 fractures in 95 patients, published by Hanson et al. [23], inferomedial avulsions (Anderson and Montesano type III) were the most common type of occipital condyle fracture observed, constituting 80 (75%) of the 107 fractures. Unilateral occipital condyle fractures were found in 73 (77%) of the 95 patients. Bilateral occipital condyle fractures or joint injuries were seen in 22 (23%) of the patients.

Radiographic signs of instability are (1) fragments involving at least 25% of the condylar articulating surface; (2) fragment displacement of 4 mm or more; (3) atlanto-occipital dislocation; (4) subluxation of C0–C1 or C1–C2; (5) C0–C1 or C1–C2 joint widening; and (6) complete transverse fracture through congenitally fused C0–C1 articulation. The consequential treatment (given a level III recommendation) can be synthesized in three options: external cervical immobilization is usually recommended; halo vest immobilization should be considered for bilateral occipitocervical–condylar fractures; and occipitocervical stabilization and fusion are recommended in cases of associated atlanto-occipital ligamentous injury and in those with evidence of instability.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Posttraumatic Anatomical Injuries of the Craniovertebral Junction and Treatment Implications: Part II

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Keywords Craniovertebral junction · Anatomy · Trauma Treatment

Introduction

This paper is Part II of a two-part report. In Part I of the report, injuries of the occipital bone, atlanto-occipital dissociation and the atlanto-occipital joint space were discussed. This part of the report discusses atlantoaxial dislocation and fractures of the atlas and axis.

Atlantoaxial Dislocation

Atlantoaxial dislocation is a pathological condition characterized by loss of stability between the atlas and the axis (C1–C2), resulting in loss of normal articulation. The transverse ligament, which runs across the posterior dens and attaches on either side of the lateral mass of C1, maintains the dens in its physiological position, stabilizing C1–C2 articulation and preventing anterior dislocation of the atlas. Additional stabilization of the dens comes from the alar ligaments, which extend from the odontoid process in a lateral and cephalad direction to the basilar portion of the occiput. The transverse ligament is larger and stronger than the alar ligaments and ensures the greater proportion of the stability, with the alar ligaments providing secondary support [1, 2].

A purely traumatic atlantoaxial dislocation in the absence of other predisposing risk factors is an extremely rare event (Fig. 1a–d). A literature review by Venkatesan et al. in 2012 found only 12 adult case reports published in the literature [3]. The mechanism underlying atlantoaxial dislocation is an abnormal movement of the neck, resulting in disruption of the transverse ligament. Rarely, disruption of the transverse ligament may be accompanied by simultaneous damage of the alar and apical ligaments.

Among odontoid fractures, type II fractures are the most common and are uniquely associated with atlantoaxial dislocation [4] (Fig. 1). Traditionally considered as a congenital anomaly, os odontoideum may in fact be caused by an early traumatic injury in which the odontoid process is completely separated from the axis and then heals, appearing as a separate ossicle. The resulting condition predisposes the patient to dislocation.

Only ten cases of patients who survived a posterior atlantoaxial dislocation without an associated odontoid fracture have been reported to date [5]. However, the incidence of this pathological condition is probably underestimated. In fact, traumatic forces causing the dislocation are likely strong enough to cause a spinal cord injury and the immediate death of the patient. In such cases, an autopsy is not capable of identifying this kind of lesion. This means that the number of posterior atlantoaxial dislocations could be considerably higher than has been reported in the literature. The mechanism underlying posterior atlantoaxial dislocation without involvement of the odontoid process may be correlation of hyperextension and distraction movements. This theory was proposed by Haralson and Boyd [6] and is supported by the finding that these patients have both head and neck lacerations, which are typically found in hyperextension cervical injuries.

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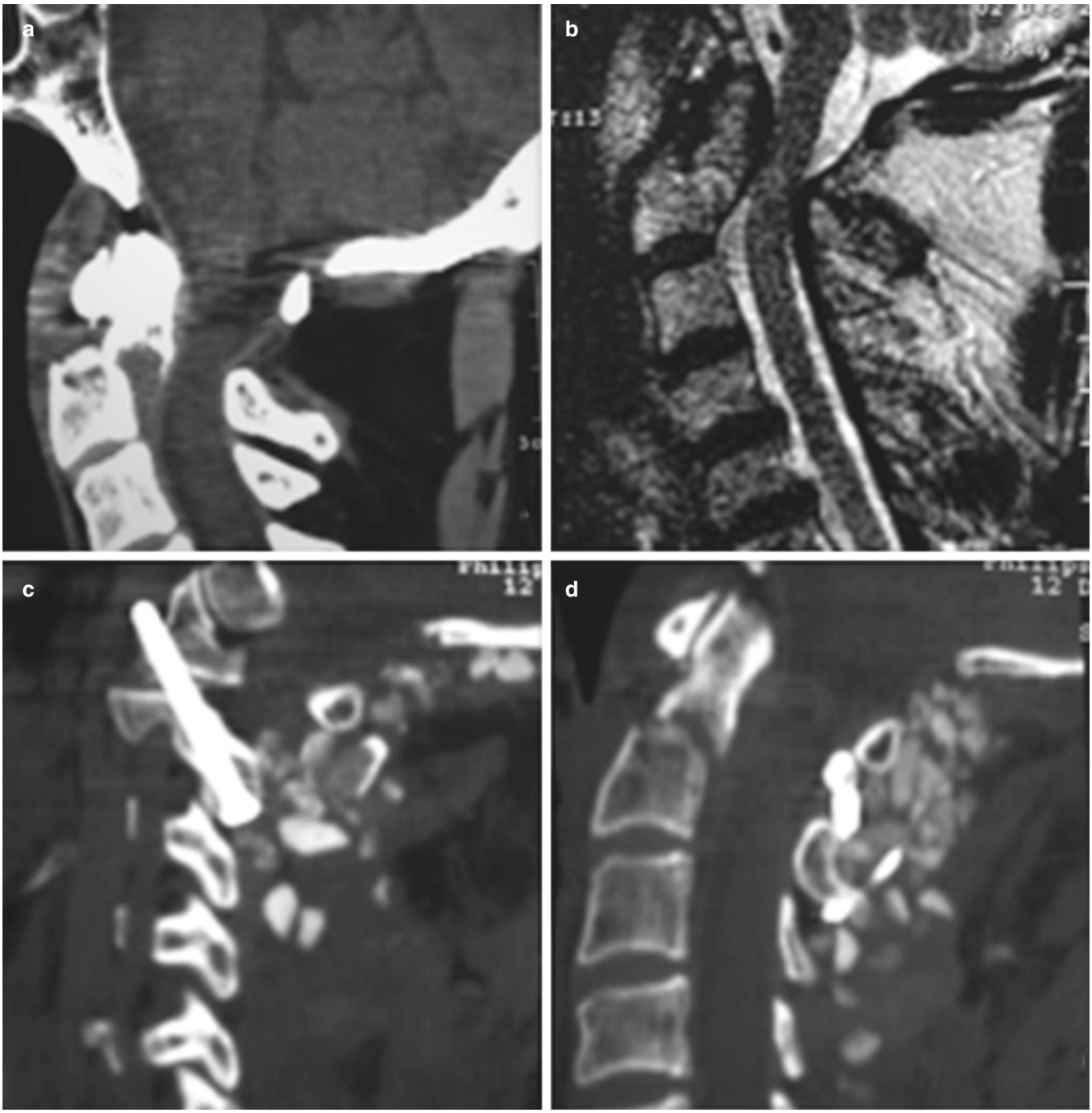

Fig. 1 (**a**) Computed tomography (CT) scan showing posterior atlantoaxial dislocation associated with an Anderson type II fracture. (**b**) Sagittal T2-weighted image of the same patient, showing the odontoid process fractured and migrated into the spinal canal, compressing the high cervical cord region. The patient is affected by severe tetraparesis. (**c**, **d**) The same patient, operated on after 2 days of halo traction (to achieve alignment of C1–C2 and odontoid repositioning), with transarticular C1–C2 screw fixation and posterior C1–C2 fusion with the iliac crest and bony chips

The most common classification, published by Fielding and Hawkins [7] and known as the Fielding classification system, has been widely accepted for clinical application. Unfortunately, it has been found not to have clinical significance in treatment or in grading of the severity of injuries, as the majority of clinical dislocations that are encountered are anterior [8].

The choice of the most appropriate treatment strategy in cases of atlantoaxial dissociation is still a matter of debate concerning conservative versus surgical treatment and, when a surgical option is chosen, which kind of surgical strategy is most suitable.

Most authors advise reserving surgical treatment for symptomatic patients, in whom it is indicated to prevent

neurological deterioration and death [9]. In asymptomatic patients the choice of treatment still represents a matter of discussion [9]. Surgical treatment has been proposed for asymptomatic adult patients with a dislocation greater than 5 mm. Surgical fusion is indicated in children when one or more of the following are present: neurological involvement, persistent anterior displacement greater than 4 mm, deformity present for more than 3 months, or recurrence of deformity following 6 weeks of immobilization. In young adults, fusion is recommended when moderate displacement is seen in flexion and extension cervical radiographs, or when instability (with or without pain) is present.

Because there is a paucity of universally accepted guidelines and a lack of patient-derived outcome data [10], more research on the relative risks of surgery versus neurological deterioration due to asymptomatic atlantoaxial dislocation is needed.

Isolated Fractures of the Atlas

Fractures of the atlas overall account for 25% of atlantoaxial complex bony injuries, 10% of cervical spine injuries and 2% of all spinal injuries. Injury of the cervical spine occurs infrequently in paediatric populations, and although C1 injuries represent only 1–2% of paediatric traumas and 2–10% of all cervical injuries in this population, the associated mortality is 16%. In one epidemiological study the authors documented 1537 cases over a 15-year period and maintained that these types of fracture are increasing among the elderly in Sweden. The annual incidence nearly doubled over the course of the study period and in 2011 was nearly 17 per million people [11].

Fractures are currently divided and classified in relation to whether there is exclusive involvement of the bone or simultaneous involvement of bone and ligaments [12], and obviously they are treated according to the risk of instability.

The atlas fracture types are subdivided into stable fractures and unstable fractures. The site can be the anterior and posterior arch for stable fractures or unstable fractures. The instability is represented by transverse ligament rupture. A third type of fracture comprises fractures of the lateral mass atlas.

In these fractures the lateral displacement of the transverse process can compress the neurovascular structures exiting from the jugular foramen against the styloid apophysis, causing Collet–Sicard syndrome (unilateral paralysis of the cranial nerves from IX to XII) [13]. A remarkable radiological sign is reduced space between the styloid process and the transverse process of the atlas ipsilateral to the cranial nerve deficit demonstrated on multiplanar computed tomography (CT).

The treatment option are listed in Table 1.

Table 1 Treatment of atlas fractures

Atlas fracture type	Treatment option
Anterior or posterior arch fractures (Type I)	Collar
Anterior and posterior arch (Type II, burst)	
Stable (transverse atlantal ligament intact)	Collar, Halo
Unstable (transverse atlantal ligament disrupted)	Halo, C1–C2 stabilization and fusion
Lateral mass fractures (Type III)	
Comminuted fracture	Collar, Halo
Transverse process fractures	Collar

LMD lateral mass displacement

Axis Fractures

The most widely used classification is as follows: fractures of the odontoid process, hangman's fractures, and miscellaneous non-odontoid and non-hangman's fractures.

Odontoid Fractures

Historically, odontoid fractures have been divided into three types.

Classification systems are useful for defining treatment algorithms. One of the most widely used ones is that suggested by the Anderson and D'Alonzo classification. On the basis of their relative stability, type I and III fractures can often be immobilized by a collar. However, there is no consensus on the treatment of type II fractures. Immobilization options may include a collar, halo device, or anterior or posterior internal fixation. Greater patient age, the extent and direction of fracture displacement, delay in diagnosis, and comminution of the fracture all negatively influence union rates.

This crucial treatment decision is further complicated by the difficulty of exact distinction between type II and type III fractures.

Some authors have described an intermediate typology of odontoid fracture called a 'shallow' or 'high' type III fracture. Thus, the distinction between types II and III is not always so obvious, and some authors advocate surgical fusion for both type II and type III fractures [14, 15]. To address this limitation, more precise parameters have been established to better distinguish type II from type III fractures [4]. Thus, oblique fractures in the anterior–posterior plane with mild C2 vertebral body extension and no C2 facet involvement are still classified as type II fractures. However, if the fracture extends into at least one of the superior articular facets of C2, the fracture is classified as a type III fracture. A second limitation of the Anderson and D'Alonzo description of type II fractures is the lack of distinction between fractures that have a broad range of different morphologies and associated treatment

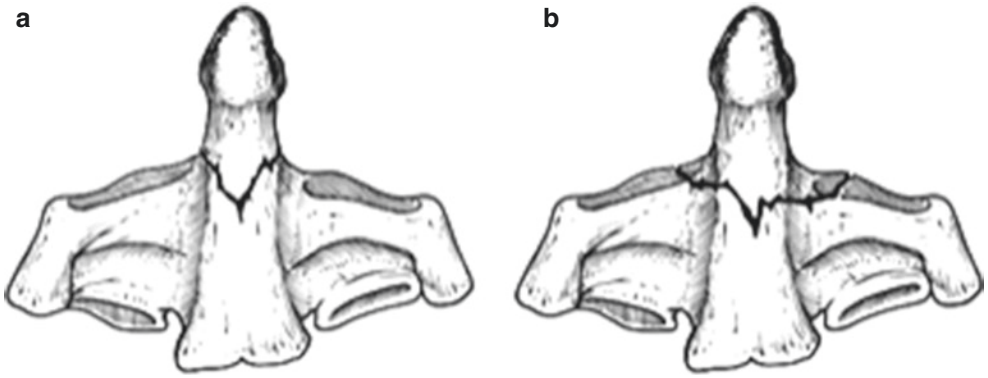

Fig. 2 The Grauer modification of the odontoid fracture classification delineates the difference between (**a**) type II fractures (with involvement of the C2 superior articular facet) and (**b**) type III fractures (without such involvement)

considerations. For example, fracture line obliquity, displacement and comminution clearly affect treatment recommendations. The different types of fracture stabilization that are now advocated for different subgroups of odontoid fractures highlight this result. To address this limitation, Hadley et al. [8] introduced a new fracture subclass, defined as a type II fracture complicated by an additional chip fracture fragment at the anterior or posterior aspect of the base of the odontoid process. This subclass, which represents 5% of type II odontoid fractures, has been observed to progress into non-union, regardless of initial fracture displacement, patient age or neurological status.

Nonetheless, there is still a wide array of fracture patterns that are classified as type II fractures. For this reason, Grauer et al. [4] proposed a treatment-oriented type II subtype classification system, consisting of type IIA, IIB, and IIC fractures (Fig. 2). The first type is defined as a minimally displaced or nondisplaced type II fracture with no comminution. These fractures are generally treated with external immobilization. The second type is a displaced fracture extending from anterior–superior to posterior–inferior or a transverse fracture. These fractures are amenable to anterior screw fixation following fracture reduction, assuming adequate bone density. The third type is a fracture line extending from anterior–inferior to posterior–superior or a fracture with significant comminution. These fractures are generally treated with posterior atlantoaxial stabilization.

Traumatic Spondylolisthesis of the Axis (Hangman's Fracture)

Bilateral fracture of the axis pars interarticularis was first described in 1866 by Haughton [16].

Garber [17] coined the term 'traumatic spondylolisthesis of C2' to describe this modern-day fracture.

In 1965, Schneider et al. [18], in a well-known paper, popularized fractures of the lamina, articular facets, pedicles or pars of the axis vertebra as hangman's fractures. They showed how old nomenclature could adapt to changes in types of trauma such as those caused by falling, diving or motor vehicle accidents. Today, the management strategies and surgical indications for hangman's fractures are still controversial, particularly for type II and type III fractures. The classification system proposed by Levine and Edwards [19] added flexion–distraction as a mechanism of injury (type IIA), four injury. The criteria used to determine the lesion's instability are either clinical or anatomical, as described by Li et al. [20].

The treatment criteria related to the type of lesions are clearly evidenced in the guidelines for the management of acute cervical spine and spinal cord injuries [21].

Combination Fractures of the Atlas and Axis

In their paper, Gleizes et al. reported that this combination of fractures was relatively common and required a high level of surveillance to be detected.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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The Decision-Making Process in Traumatic Lesions of the Craniovertebral Junction: An Evidence-Based Approach? Part I

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Keywords Craniovertebral junction · Injuries · Evidence

Evidence-based medicine is a rather young concept, which entered the scientific literature in the early 1990s.

This approach is intended to counteract the empirical approach to research and strengthen epistemology on the basis of the strongest scientific papers (meta-analyses, systematic reviews and randomized controlled trials) [1].

Elements of the Decision-Making Process

In this paper we summarize the essential elements of the decision-making process. Each anatomical region is then discussed separately. This part of the paper (Part I) covers atlanto-occipital dislocation or dissociation, and isolated condylar fractures. Part II of the paper covers isolated and combination fractures of the atlas and axis.

The first topic is the processing of radiographic instability criteria based on x-rays, computed tomography (CT) scanning and magnetic resonance imaging (MRI). These depend substantially on identification of types of lesions, taking into consideration the following:

Lesion type and epidemiology: Bone injury, ligamentous injury or a combination of the two.

Bone displacement: Listhesis, angulation and/or subluxation.

Ligament ruptures: Avulsion, stretching, retroclival collection.

Measurements: Distances and intervals between bony structures of the craniovertebral junction (CVJ) or bony fragments of the involved vertebrae.

Patient age: In paediatric patients (mostly children aged <10 years), injuries of the C0–C2 region are the predominant form of cervical injury. In elderly patients, treatments options can be different from those in younger adults. In recent years, the incidence of cord injury caused by child abuse has increased [2, 3].

The paediatric population (the younger patients are affected more than older with differences in physiological development and evolving anatomy) is particularly susceptible to craniocervical trauma [4, 5].

Degree of neurological involvement: Complete, incomplete or focal neurological deficits (Fig. 1).

Atlanto-occipital Dislocation or Dissociation

Atlanto-occipital (AO) dislocation or dissociation (AOD), once considered generally rare, is fatal in its most severe forms, showing disruption of craniocervical ligaments [2, 4]. In autopsy studies it represent 12% of identified cervical injuries; the most common mechanism is a pedestrian being struck by a motor vehicle.

Bellabarba et al. classified lesions on the basis of their stability, as follows: (1) stable lesion (immobilization treatment); (2) stable, but the stability must be demonstrated with a traction test (obviously, every effort must be made to identify hidden instability); and (3) high unstable and potentially fatal lesion.

These patients frequently present with vascular injuries and must be screened for blunt cerebrovascular injury.

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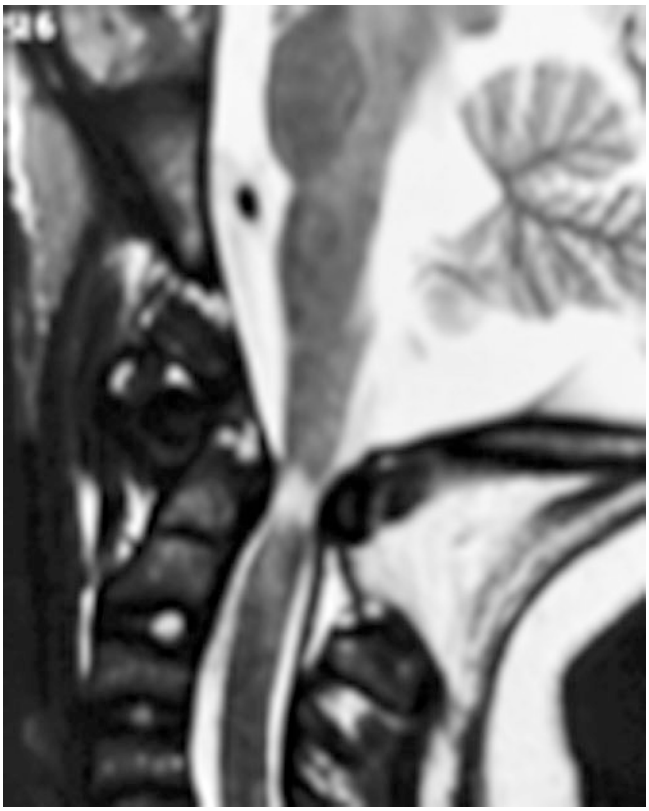

Fig. 1 Six-year-old child involved in a road traffic accident. No bony injury is identified by computed tomography (CT) scanning, but a clear-cut abnormal high-intensity signal in the cervicomedullary region is identified by magnetic resonance imaging (MRI)

Sun et al. stated in 2000 [6] that involvement of the tectorial membrane (TM) (Fig. 2) was a critical threshold for unstable ligamentous injury of the CO–C2 region because (1) all spinal cord injury (SCI) occurred with TM injury; (2) all TM injuries also involved AO joint disruptions; (3) cases treated without fusion had progressive MRI changes; and (4) abnormal CO–C2 measurements were found only with TM injury. Other entities that have received increasing consideration in recent years are retroclival subdural collection (the vast majority) and epidural collection, described after trauma in general and particularly in child abuse cases. In the literature, TM rupture seems to play a major role in the formation mechanism.

With regard to this injury, the guidelines for the management of acute cervical spine and spinal cord injuries, published in 2013, give a level I recommendation for CT scanning to determine the condyle–C1 interval (CCI) in paediatric patients and a level III recommendation for CT scanning in adults.

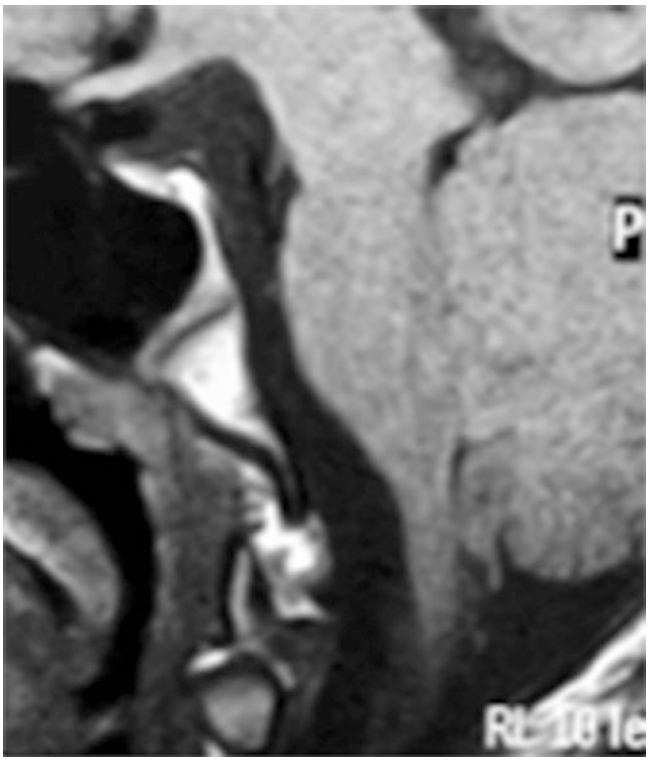

Fig. 2 Young child involved in a road traffic accident. Magnetic resonance imaging (MRI) shows apical ligament rupture and stretching of the tectorial membrane

Isolated Condylar Fractures

Occipital condyle fractures (OCFs) are considered common nowadays, since the introduction of CT into clinical practice. In these lesions, however, it is advisable to evaluate abnormalities of the lower cranial nerves on admission, which are frequently described, and calculate the patient's Glasgow Coma Scale (GCS) score (high frequency of loss of consciousness).

The following recommendations given in the 2013 guidelines are only rated as level III: (1) treatment with external cervical immobilization; (2) consideration of more rigid external immobilization in a halo vest for bilateral OCFs; and (3) halo vest immobilization or occipitocervical immobilization and fusion in patients with associated AO ligamentous injury or evidence of instability (Fig. 3).

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

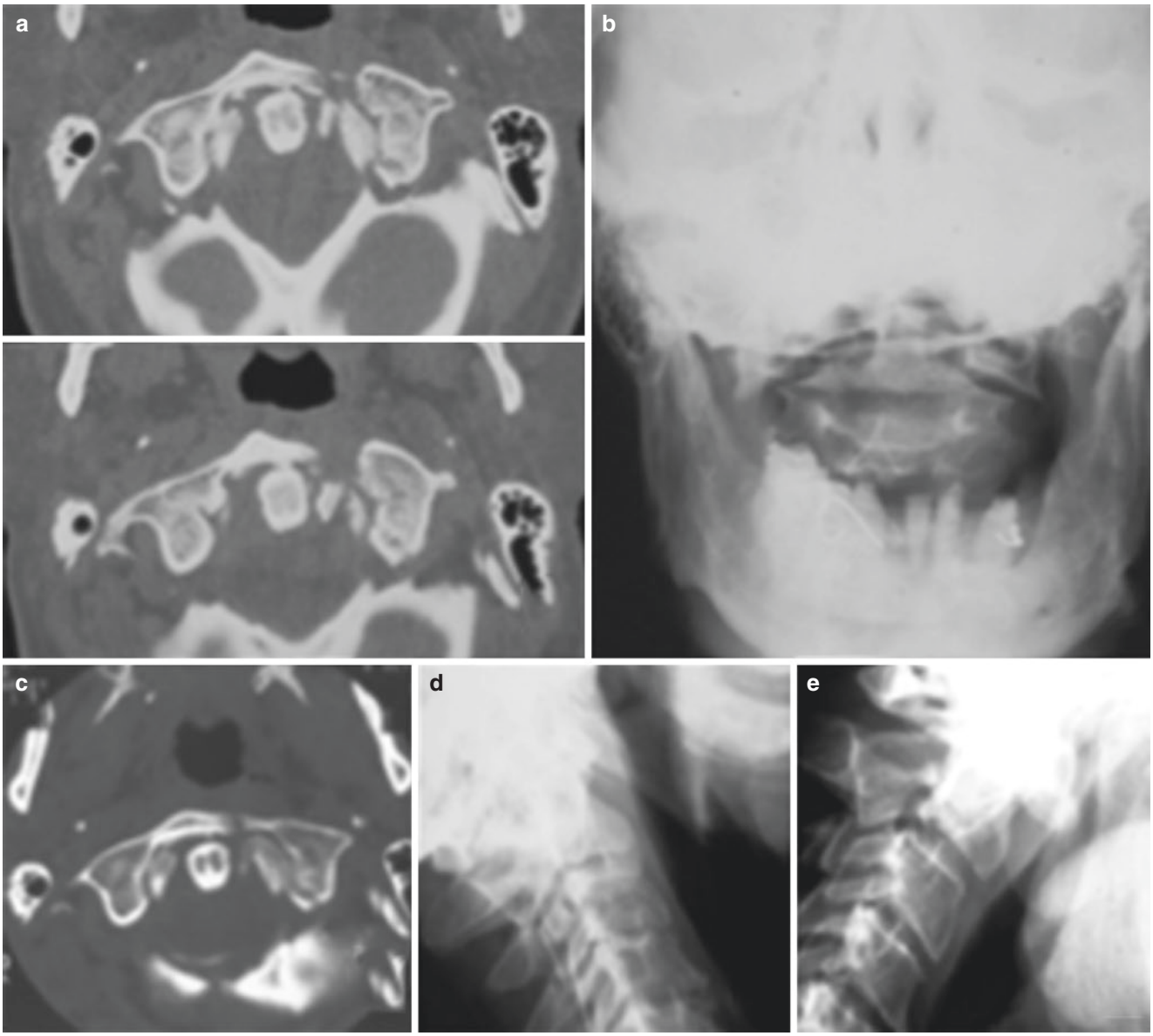

Fig. 3 Fifty-five-year-old man involved in a road traffic accident. (**a**) Computed tomography (CT) scanning shows a left condylar fracture associated with a left lateral articular mass of the atlas fracture. (**b**) The Spence distance is increased slightly. (**c**–**e**) Control CT scans and X-rays show optimal fusion and stability (dynamic x-rays normal) after 1 year, achieved by conservative treatment (wearing of a Philadelphia collar only)

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The Decision-Making Process in Traumatic Lesions of the Craniovertebral Junction: An Evidence-Based Approach? Part II

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Keywords Craniovertebral junction · Injuries · Evidence

Introduction

This paper is Part II of a two-part report. Part I of the report covered atlanto-occipital dislocation or dissociation, and isolated condylar fractures. This part of the report covers isolated and combination fractures of the atlas and axis.

Isolated Fractures of the Atlas

Historically, Jefferson has been considered the first author to classify these fractures, although a recently published paper described an Italian surgeon named Quercioli as the first author to identify a quadripartite fracture of the atlas [1].

The classification is extremely variable, but basically these fractures are divided into three main types:

Type 1: Only one arch involved (anterior or posterior)
Type 2: Both arches (anterior and posterior) involved
Type 3: Lateral mass involved with or without the arches

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In fracture type 3, the involvement of cranial nerves from IX to XII (Collet–Sicard syndrome) has been reported [1].

The instability is due to injury of the transverse ligament, which is shown by magnetic resonance imaging (MRI) in 60% of patients [2]. Dynamic x-rays, performed cautiously, may be required to confirm this instability.

Isolated Fractures of the Axis

These fractures are divided into three types [1–11]:

Type 1: Odontoid fractures
Type 2: Traumatic spondylolisthesis of the axis
Type 3: Combination fractures of the atlas and axis

Odontoid Fractures

The old subdivision described by Anderson and D'Alonzo is still usually valid and is reported in the first part of this paper. The most critical issue is the type II fracture because some authors have recognized subtypes such as Hadley type IIA (with fragments of bone comminution at the base of the odontoid process). Similarly, Grauer et al. described three subtypes:

Subtype A: Similar to Anderson type II
Subtype B: Oblique fracture line displacement from superior to posterior inferior
Subtype C: Transverse fracture line and inferior to posterior superior comminution

Surgery is generally indicated in comminuted fractures and/or when radiographic instability signs are present. In the field of type II odontoid fractures, level II evidence has been reported for surgical stabilization in patients over 50 years

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old. Immobilization is recommended in all fracture types [8, 12, 13] with acceptable alignment, while anterior or posterior surgery [14–16] is required when the atlas to dens interval is >5 mm.

Traumatic Spondylolisthesis of the Axis

This term was coined by Garber in 1964 [17] to describe this modern-day fracture, which is similar to that sustained by judicial hanging (*hangman's fracture*), but Schneider, in a well-known 1965 paper, popularized the old term [18].

The incidence of this particular type of fracture in the context of cervical spine injuries was studied and reported by Greene et al. in 1976 [8]. They reported a 4% incidence of hangman's fractures out of a total of 1820 cervical fractures. Concomitant cervical fractures, atlas fractures and hangman's fractures have been reported in the literature, with a variable incidence. Cord injury is rare, and spinal instability is not frequently reported. There are at least eight classifications to define the fracture subtypes. Only level III evidence is reported for surgery; cervical immobilization in a halo device is the most common type of treatment, and surgery is reserved for cases of relevant angulation and/or disruption of C2–C3, or cases of failure of alignment following external immobilization (Figs. 1–3).

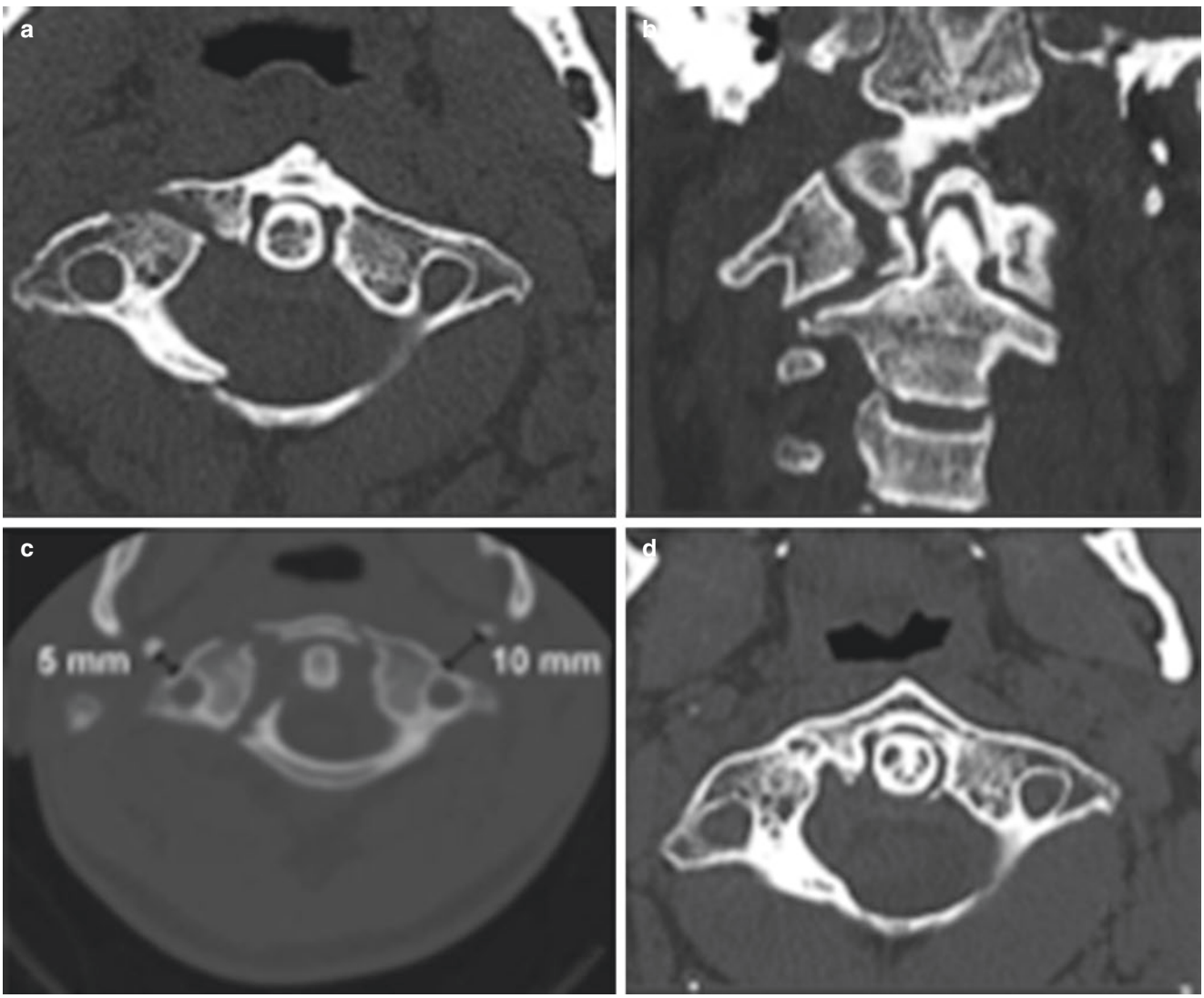

Fig. 1 (**a**) Axial and (**b**) coronal computed tomography (CT) scans showing an atlas fracture involving the anterior and posterior rings of the atlas. The transverse ligament is intact, as demonstrated by the normal atlantodental interval. In this fracture type the lateral mass of the atlas can be displaced laterally, compressing vascular and nerve structures against the styloid process and causing Collet–Sicard syndrome. (**c**) Bone window CT scan showing an approximation of the distance from the lateral mass of the atlas to the styloid process due to displacement of the lateral mass of the atlas. **d** Bone fusion after some months

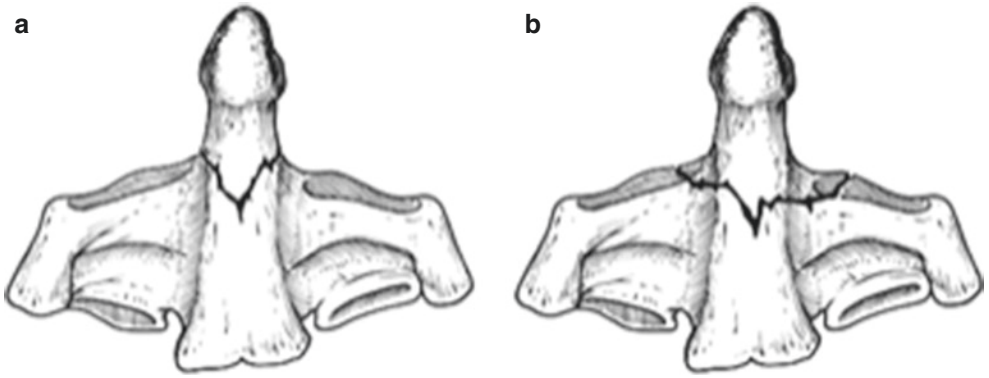

Fig. 2 The Grauer modification of the odontoid fracture classification delineates the difference between (**a**) type II (with involvement of the C2 superior articular facet) and **(b)** type III (without such involvement)

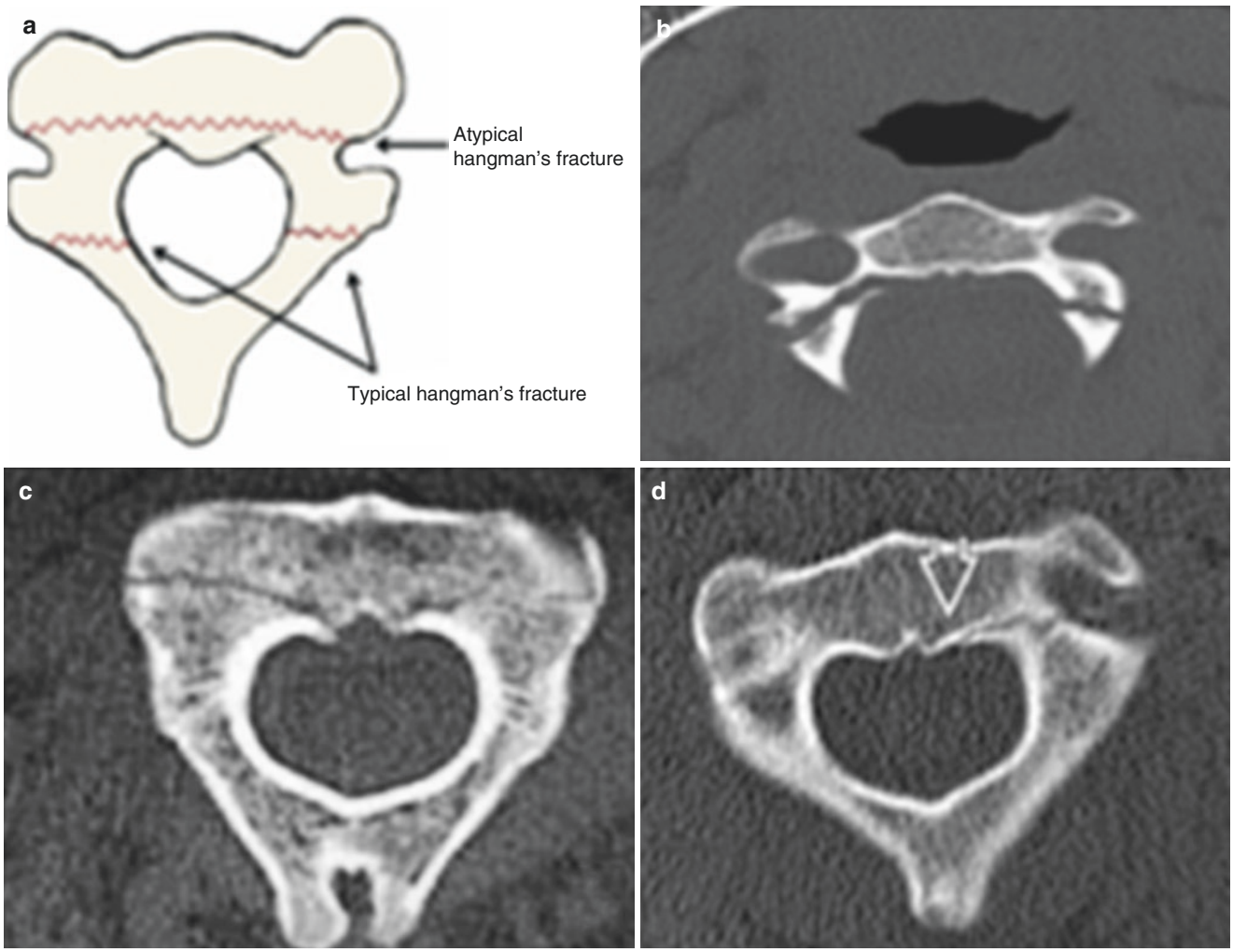

Fig. 3 (**a**) Schematic illustration and distinction between (**b**) typical and (**c**, **d**) atypical hangman's fractures (not involving the isthmus of the axis)

Combination Fractures of the Atlas and Axis

The most frequent combinations are:

1. Hangman's fracture associated with odontoid fracture
2. Odontoid fracture associated with C2 lateral mass fracture
3. Jefferson fracture associated with odontoid fracture

Neurological deficits are more prevalent in combination fractures than in C1–C2 fractures alone. The recommendations are, however, level III. It should be noted that in surgical practice, it is difficult to conduct randomized controlled trials, as they can present ethical conflicts that make it very difficult to draw useful conclusions.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Type II Odontoid Fracture: a case series highlighting the treatment strategies

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Abstract *Background:* A type II odontoid fracture, if unstable, can cause spinal cord damage. In this case, it is essential to choose the correct treatment—but the issues of what the correct treatment is and which of the different surgical options is best are quite controversial. In this paper we present strategies for treatment of type II odontoid fracture.

Materials and Methods: Thirty consecutive cases of type II odontoid fracture were treated at the Division of Neurosurgery at Villa Sofia Hospital in Palermo (23 cases) and at the Neurosurgical Clinic, University Hospital of Palermo (seven cases), from January 2011 to August 2016. Four patients were treated with external immobilization. Twenty-six patients underwent a surgical procedure.

Results: There was no mortality related to the surgical procedure. One patient had a pre- and postoperative neurological deficit, and remained tetraparetic. Follow-up radiological studies in the surgically treated group showed bone union in 21 patients and stable fibrous union in one.

Conclusion: In our and other authors' experience, when the direction of the fracture line is down and forward, external immobilization can be sufficient for healing. Anterior odontoid screw fixation can be considered the treatment of choice for unstable odontoid fractures (with a horizontal, down and back, or comminuted fracture line) without dislocation or with dislocation less than 7 mm.

When the odontoid fracture is associated with a Jefferson fracture or dislocation greater than 7 mm, stabilization of C1–C2 may be necessary. In this case, placement of screws in the dens and in the joints through a single approach represents the most valid technique.

In the case of an inveterate fracture of the dens with severe C1–C2 dislocation, the surgical operation that offers the best prospects is posterior stabilization, utilizing the Guo technique.

Keywords Type II odontoid fracture · C1–C2 instability · Odontoid screw · C1–C2 articular screw

Introduction

Odontoid fractures represents about 20% of all cervical fractures. Of these, the Anderson and d'Alonzo type II fracture (on the base of the dens) is the most common, occurring in more than 60% of cases; when it is unstable, it can cause spinal cord damage. In this case, it is essential to choose the correct treatment [1–3]—but the issues of what the correct treatment is and which of the surgical options is best are quite controversial [4–7]. We present a strategy for treatment of type II odontoid fractures.

Materials and Methods

Thirty consecutive cases of type II odontoid fracture were treated at the Neurosurgery Division of Villa Sofia Hospital in Palermo (23 cases) and at the Neurosurgical Clinic, University Hospital of Palermo (seven cases), from January 2011 to August 2016. There were 19 males and 11 females.

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The median age was 58.3 years (range 12–89 years). The direction of the fracture line was oblique, down and forward without dislocation in four patients, and they were treated with external immobilization (a sternal–occipital–mandibular immobilizer [SOMI] brace). In 18 cases the fractured dens had no dislocation or had a dislocation less than 7 mm (the direction of the fracture line was horizontal in six cases and oblique, down and back in 12 cases), and they were treated with odontoid screw fixation. The fractured dens was associated with a Jefferson fracture in two cases and with a dislocation greater than 7 mm in five patients. These seven patients were treated with anterior screw fixation of the dens and bilateral C1–C2 anterior transarticular screw fixation. Finally, one patient had an inveterate odontoid fracture with severe lateral C1–C2 luxation and was treated with manual reduction followed by posterior stabilization utilizing the Guo technique (bilateral C1–C2 transarticular screws, C1 laminar hook fixation and bone graft fusion). Clinical and radiological follow-up was performed in all but three of the patients.

Results

The follow-up of the patients ranged from 4 months to 5 years. In the conservatively treated group, bone fusion was observed in three patients and fibrous union in one. In the surgically treated group there were no deaths except for one woman in a coma due to a severe head injury, who died from pneumonia after 40 days. One patient, who had an inveterate fracture, had a pre- and postoperative neurological deficit and remained tetraparetic. In the early postoperative period, 11 patients experienced mild dysphagia, which required no treatment beyond dietary modification. In the patients undergoing surgery, radiological studies showed bone union in 21 patients and stable fibrous union in one.

Case Illustration

Case 1

This 17-year-old boy sustained a head and neck injury in a diving accident. No neurological deficit was noted. A computed tomography (CT) scan showed a type II odontoid fracture. The direction of the fracture line was oblique, down and back without dislocation. He was treated with odontoid screw fixation. A postoperative CT scan demonstrated correct placement of the screw (Fig. 1). One year later, dynamic radiography showed no dislocation, and bone fusion was evident.

Case 2

This 75-year-old man sustained a head and neck injury in an accidental fall from body height. The neurological examination was normal. A CT scan disclosed a type II odontoid fracture, with the direction of the fracture line being oblique down and back, with 10-mm posterior dislocation of the fractured dens and of the C1 joint facets with respect to C2. The patient underwent manual reduction under the guidance of image intensification. A subsequent CT scan showed good alignment of the fracture and of the joint facets; therefore, anterior screw fixation of the odontoid fracture and of the bilateral C1–C2 joints was performed. A postoperative CT scan showed correct placement of the screws (Fig. 2). In the first 10 days the patient had slight dysphagia. At follow-up after 9 months a CT scan documented bone fusion of the fracture.

Case 3

This 68-year-old man presented with immediate tetraparesis after mild cervical trauma. He stated that he had experienced neck pain after a head injury 5 months earlier. A neuroradiological study showed an inveterate odontoid fracture with severe lateral C1–C2 dislocation. The patient underwent manual reduction under the guidance of image intensification. A CT scan showed good alignment of the fractured dens and of the C1–C2 articular facets. Posterior stabilization with the Guo technique (bilateral C1–C2 transarticular screws, C1 laminar hook fixation and bone graft fusion) was performed. A postoperative x-ray and CT scan showed the alignment of C1–C2 and correct placement of the implant system (Fig. 3). At follow-up after 6 months, the patient's neurological deficits were unchanged and a CT scan documented bone union in relation to the bone graft.

Discussion

Type II odontoid fractures in young people tend to be due to high-energy trauma, but in the elderly population they can occur through low-impact mechanisms because the dens becomes significantly less robust with age. Some of these fractures can be considered unstable, with a risk of spinal cord damage. But when exactly can this fracture be considered unstable? On the basis of studies by Roy-Camille et al. [8] and clinical experiences at the Hôpital Pitié-Salpêtrière in Paris [9], we agree that type II odontoid fractures are unstable and need surgical treatment when they are (1) horizontal, (2) oblique down and back, or (3) comminuted. Therefore, in these cases—and if the fractured dens is not dislocated or is dislocated by less than 5 mm—it is essential to choose the correct treatment, and this issue is quite controversial [4–6].

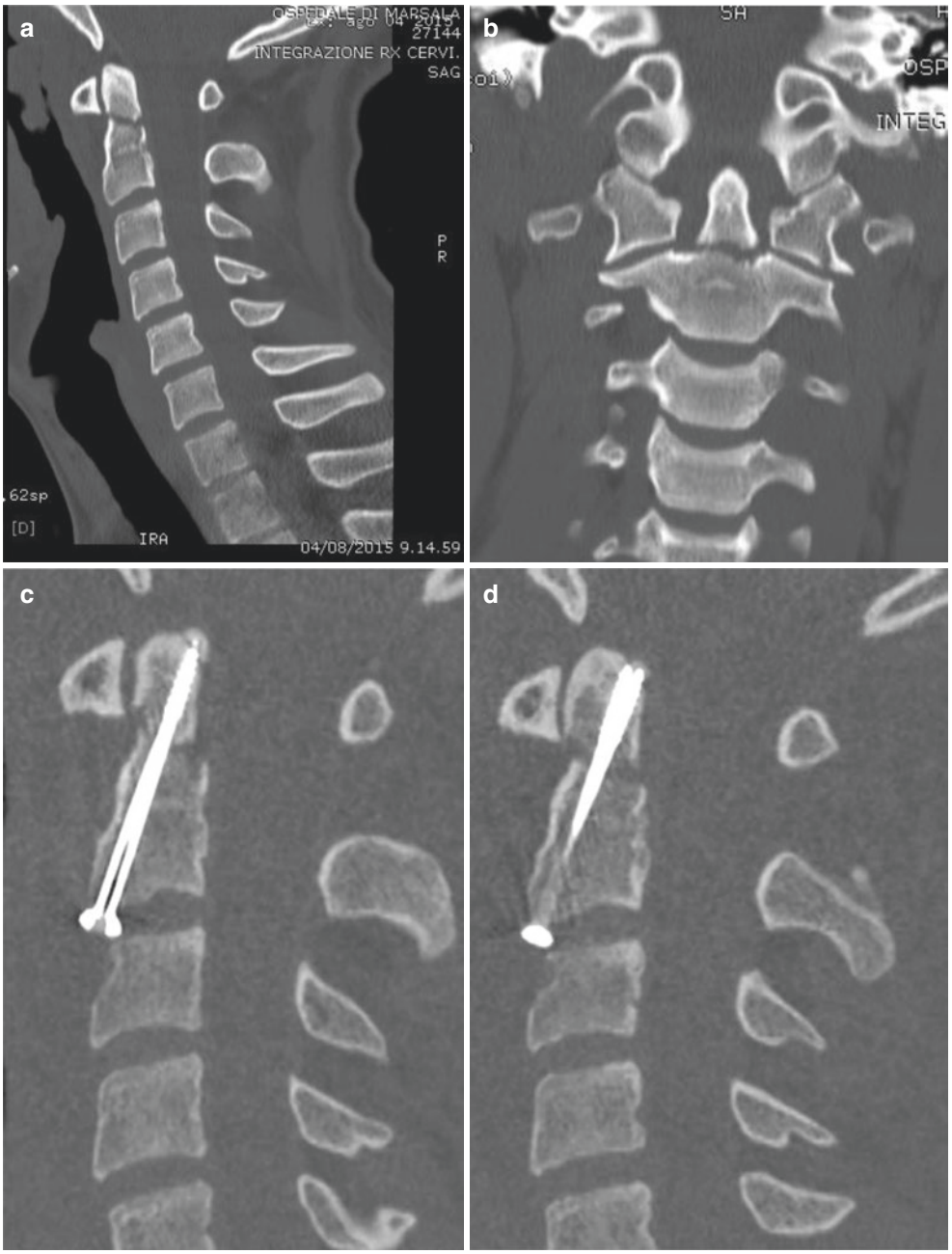

Fig. 1 Sagittal and coronal computed tomography (CT) reconstruction showing an oblique down and back odontoid fracture (**a**, **b**). Postoperative CT reconstruction revealing correct screw placement (**c**, **d**)

Conservative treatment with use of external immobilization (a halo vest, SOMI brace or cervical collar) guarantees fusion of the fracture in only a small proportion of patients—particularly in elderly patients—with a risk of dislocation and spinal cord compression [1]. Some authors have reported deaths due to cardiopulmonary compromise in elderly patients with odontoid fractures treated with halo vest immobilization. For these reasons there is a trend for many of these cases to be treated surgically [1, 10]. Posterior cervical fusion has historically been the first option for patients with odontoid fractures and for patients with C1–C2 instability. One of the earliest types of fixation for C1–C2 fusion was described by Gallie. It involved fixation of the posterior arch of C1 and of the lamina or spinous process of C2, using a cerclage wire technique with an onlay bone graft, but the failure rate was high. A modification of this technique by Brooks [11] also had limited success, with a 30% failure rate. Also, use of Halifax clamps with C1 and C2 laminar

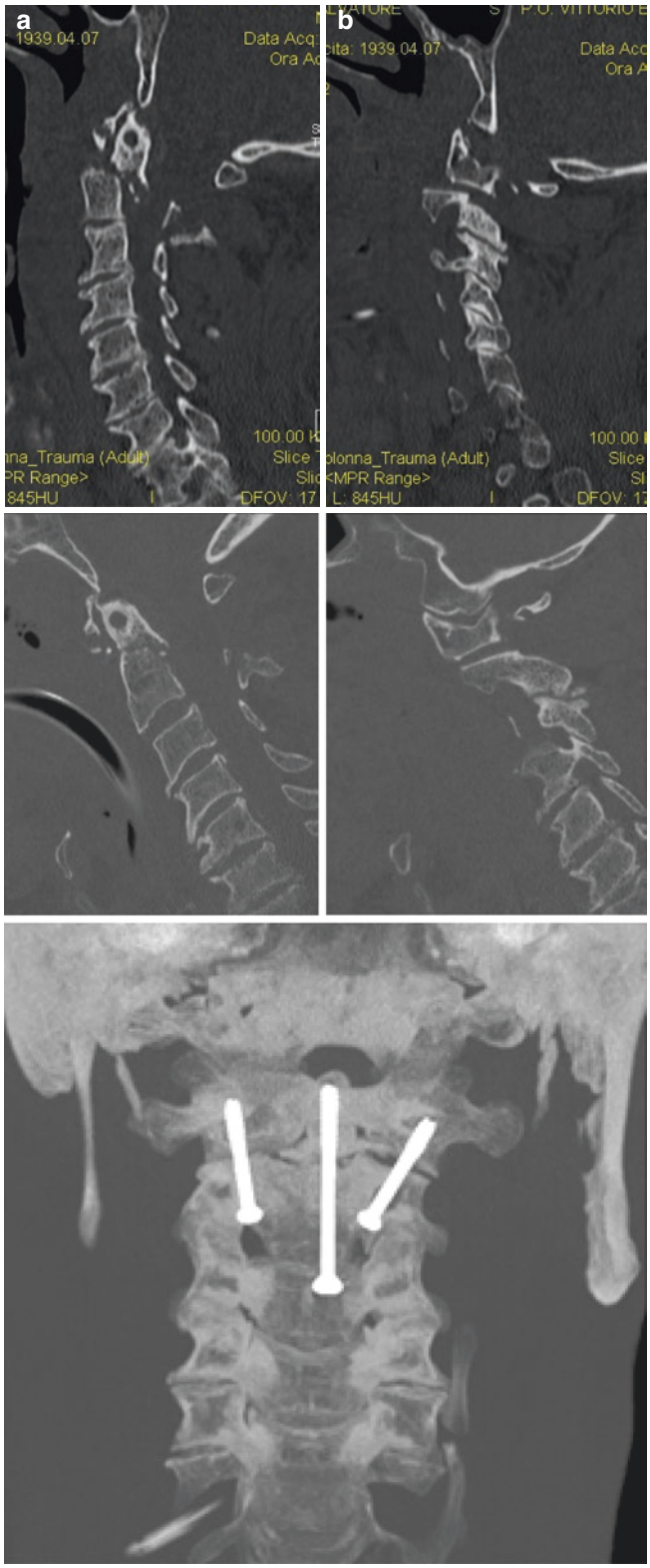

Fig. 2 Computed tomography (CT) scan on admission, showing an odontoid fracture with 10-mm posterior dislocation of the fractured dens and of the C1 articular facet with respect to C2 (**a**, **b**). CT reconstruction after manual reduction, demonstrating good alignment (**c**, **d**). Postoperative three-dimensional CT scan (**e**)

hooks, united by screws, had a 30% failure rate [12]. These discouraging results prompted research into new approaches; therefore, since the early 1980s, other techniques have been devised for treatment of odontoid fracture and C1–C2 instability.

In 1982, Bohler [13] presented the surgical technique of anterior screw fixation of odontoid fractures. This is an osteosynthetic technique that provides immediate stability, promotes healing and may preserve C1–C2 rotational motion. It also offers several advantages, including reduction of soft tissue trauma, a decreased risk of vertebral artery injury, lack of requirement for bone grafting, a shorter operating time and a shorter hospital stay. The most significant complication is represented by dysphagia. Use of this technique may not be possible in patients with a barrel-shaped chest, a short neck and impossibility of extending the neck. The fusion rate, using this technique, is 81–90%. In 1986, Magerl [14] proposed a new technique for C1–C2 fusion, using posterior transarticular screws associated with cerclage wiring and bone graft. With this technique a 90–100% fusion rate was achieved. Later, similar results were reported with use of posterior transarticular screws without cerclage wiring and bone grafting, avoiding the risks of passage of sublaminar wire and of graft migration [15]. However, insertion of posterior screws can be difficult or impossible in anatomical conditions such as a narrow pars interarticularis or a high-riding foramen transversarium, which places the vertebral artery at high risk of injury. This technique also carries risks of spinal cord and vertebral artery injury, screw breakage and infection. In 2001, Harms [16] presented a technique of C1–C2 fixation with bilateral insertion of polyaxial-head screws into the lateral mass of C1 and into the pedicle of C2, with rod fixation; with this technique it is also possible to treat irreducible fractures with a 100% fusion rate. But, of course, this is also a technically demanding operation. The most recently presented technique for posterior stabilization of C1–C2 was published in 2014 by Guo et al. [17]: use of bilateral C1–C2 transarticular screws with C1 laminar hook fixation and bone graft fusion. This operation is more advantageous than the Magerl technique because it avoids sublaminar passage of wiring, and it is more advantageous than the Harms technique because it provides three-point fixation instead of two-point fixation. The risks and technical difficulty of C1–C2 posterior stabilization had stimulated surgeons to research an easier and less dangerous approach to treat C1–C2 instability, so the surgical technique of C1–C2 anterior transarticular screw fixation has been disseminated [18–20]. This technique offers several advantages: the positioning of the patient is much simpler; the surgical approach is less traumatic with a lower infection rate; and the risks of spinal cord and vertebral artery injury are lower. The good clinical results

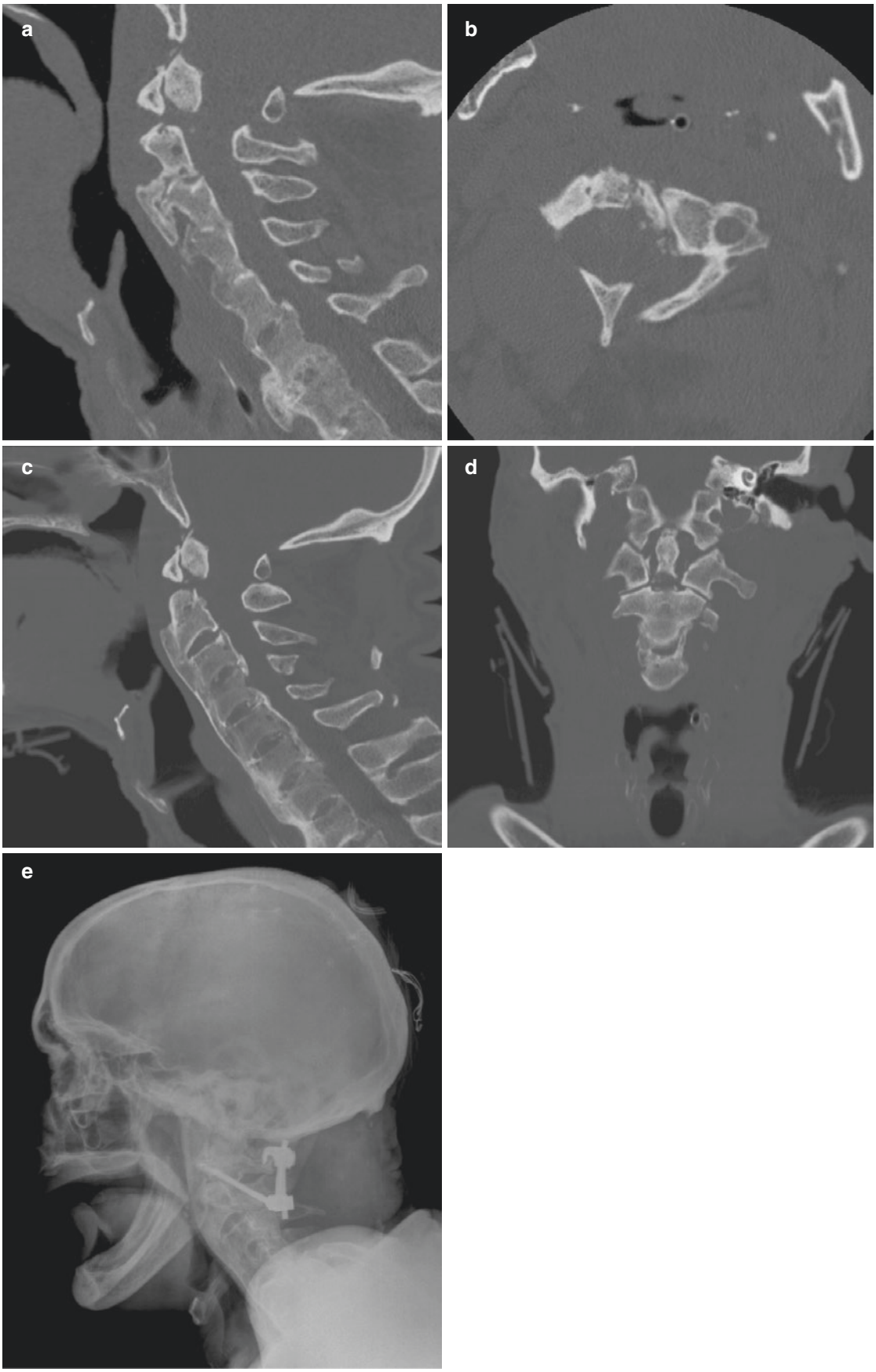

Fig. 3 Computed tomography (CT) scan on admission, revealing an inveterate odontoid fracture with lateral dislocation of C1 on C2 (**a**, **b**). CT scan after manual reduction, showing good alignment (**c**, **d**). Postoperative lateral x-ray (**e**)

achieved with this technique have been confirmed by a biomechanical study published by Sen et al. [21], who demonstrated that there was not a large difference in the strength of C1–C2 fixation between use of anterior transarticular screws and use of posterior transarticular screws alone. Furthermore, when C1–C2 instability is combined with odontoid fractures, treatment of both can be done through a single anterior approach. But what causes C1–C2 instability? Current opinions cite transverse ligament injury, odontoid fracture dislocation greater than 5 mm and associated fractures of C1 and C2, while less importance or attention is given to C1–C2 joint injury. The physiological C1–C2 range of motion in flexion and extension is minimal—only 13 grades—and occurs on the sagittal plane without loss of alignment of the articular facets. So is C1–C2 joint integrity conceivable if the dislocation of the fractured odontoid process is greater than 7 mm? Moreover, can it be speculated that dislocation of the fractured odontoid process, which sometimes exceeds 1 cm, happens without slippage of both C1 lateral masses forward or backward with respect to the C2 facets and without capsular ligament injury? As shown in Fig. 4, T2-weighted magnetic resonance imaging (MRI) performed

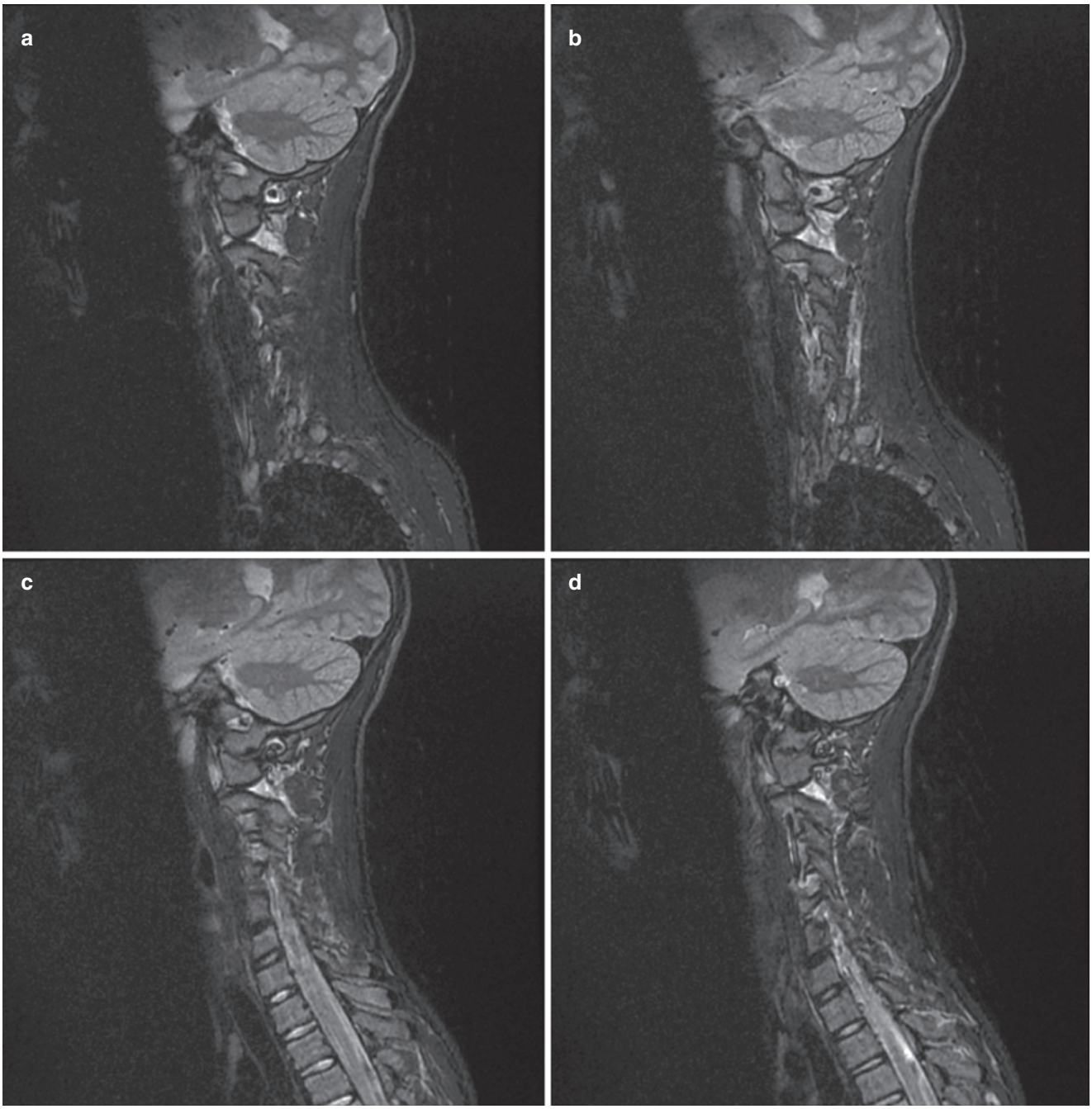

Fig. 4 Sagittal T2-weighted magnetic resonance imaging (MRI) of case 2: the high intensity of the tissues back to the C1–C2 joints is an expression of distension and rupture of the capsular ligaments

in case 2 showed high signal intensity of the tissues posterior to the C1–C2 joints, expression of distension and rupture of the capsular ligaments. Therefore, C1–C2 joint instability in odontoid fractures is an underestimated event, in our opinion. Unrecognized C1–C2 joint instability could be the cause of some failures of odontoid screw fixation: non-union, breakage of the anterior portion of the C2 vertebral body, and screw breakage.

Therefore, if the dislocation of the fractured dens is greater than 7 mm, an accurate CT reconstruction of the joints can reveal C1–C2 subluxation, and MRI can reveal distension and rupture of the capsular ligaments of the C1–C2 joints. In this case, odontoid screw fixation alone may be insufficient and stabilization C1–C2 may be necessary. Moreover, in cases of inveterate fracture of the dens with severe C1–C2 dislocation, as in our patient, the surgical operation that offers the best prospects is posterior stabilization according to the Guo technique [17]. In fact, in such cases, screw placement in the dens cannot cause bone fusion and healing for the interposition of fibrous tissue in the rims of an ancient fracture, and anterior or posterior transarticular screws may be insufficient for healing, whereas the Guo technique, involving three-point fixation, may be an appropriate treatment.

The numbers in the present case series are too small to permit us to affirm that our treatment strategy is definitely effective. However, the results we achieved appear to indicate that this is a promising direction. Moreover, the 7-mm cut-off point for dislocation of the fracture and of the C1–C2 articular facets that we have considered as a boundary for deciding between more simple odontoid screw fixation and anterior placement of screws in the dens and the C1–C2 articular facets could be too low. But to solve this question, more studies will be necessary.

Conclusion

Despite the frequency of type II odontoid fracture, the most appropriate treatment is still a matter of discussion. In our and other authors' experiences, when the direction of the fracture line is down and forward, external immobilization can be sufficient for healing. For us, anterior odontoid screw fixation can be considered the treatment of choice for unstable odontoid fractures (with a horizontal, down and back, or comminuted fracture line) without dislocation or with dislocation less than 7 mm.

In our opinion, the presence of C1–C2 joint injury in odontoid fractures is underestimated, and this could be the cause of some failures of anterior odontoid screw fixation. Therefore, when an odontoid fracture is associated with C1–C2 dislocation greater than 7 mm, stabilization of C1–C2 could be necessary. C1–C2 instability is now commonly treated with posterior screw fixation, but these technically demanding operations can be limited by anatomical conditions and carry severe risks. Therefore, we—like many authors—think that C1–C2 anterior transarticular screws can be considered an effective alternative procedure. However, if C1–C2 instability is associated with a type II odontoid fracture, screw placement in the dens and in the joints through a single approach represents the most valid technique.

Finally, in cases of inveterate fracture of the dens with severe C1–C2 dislocation, the surgical operation that offers the best prospects is posterior stabilization utilizing the Guo technique. To date there is insufficient evidence to establish a standard or guideline for odontoid fracture management [6]. A randomized trial or serial case–control studies will be necessary. Our work must be considered preliminary, and other studies are necessary to confirm this treatment strategy.

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Learning Curve in Surgical Treatment of Odontoid Fixation for a Series of Type II C2 Fractures

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Keywords Type II odontoid fracture · C1–C2 instability · Odontoid screw · C1–C2 articular screw · Learning curve

Introduction

The craniovertebral junction (CVJ) is a complex anatomical area upon which most of the motion of the upper cervical spine depends [1]. Because of its unique range of motion, the CVJ is subject to several types of traumatic injury; it has been shown that odontoid fractures are the most common ones in the general population and are the most common isolated spinal fractures [2]. Accounting for up to 18% of all cervical fractures, odontoid fractures are the most common ones in elderly patients [3], in whom they account for up to 60% of spinal cord injuries [4].

Three different types of odontoid fracture were standardized and classified by Anderson and D'Alonzo [5]. Type I is an avulsion fracture at the tip of the odontoid process, above the transverse ligament; type II is a fracture of the body of the odontoid process, between the transverse ligament and the base of the odontoid process; and type III is a basilar fracture extending to the axis vertebral body. Conservative treatment is widely supported for type I and III odontoid fractures, but there is no clear consensus on the optimal treatment of type II fractures [4, 6]. Because of the high risk of cord damage and the lower fusion rates, type II odontoid fractures are often treated surgically by challenging yet life-saving surgery. To reduce the high mortality rate seen with conservative external immobilization, several surgical techniques have been proposed to stabilize and fix odontoid fractures [6, 7]. Although the choice of surgical treatment is still a matter of debate, it has been shown that use of anterior odontoid screws guarantees immediate stabilization of the fracture, produces osseous fusion rates of 88–100% over the course of a year and preserves the neck's range of motion, sparing the patient from the need for C1–C2 fusion [6].

Although anterior screw fixation of odontoid fractures may be the treatment of choice in younger patients with favourable fracture geometry to preserve neck motion [4], the complex and potentially dangerous anatomy of the CVJ exposes the spinal surgeon to a higher risk of complications. In this regard, it is vital that the proficiency goals and the point at which a surgeon is expected to become proficient are well defined, as in other spinal surgical procedures [8]. To assess this, we have to look at the concept of the learning curve to define when a surgeon may be considered proficient in autonomously fixing odontoid fractures through an anterior cervical approach. The learning curve is defined as the time after which a surgical procedure can be performed with safety and efficiency, and it is often related to the operating time [9]. To define a learning curve for anterior fixation of odontoid fractures, we retrospectively evaluated 25 consecutive cases of type II odontoid fracture treated through anterior placement of transodontoid screws, and we analysed the operating time and the patient's radiological exposure as markers of surgical skill improvement.

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Materials and Methods

Between January 2011 and August 2016, 25 consecutive type II odontoid fractures were jointly treated at the Neurosurgical Unit of Villa Sofia Hospital (20 cases) and the University Hospital of Palermo (five cases) by two surgeons (EF and RM). Of the 25 patients, 16 were male and nine were female; the median age was 58.3 years (range 12–89 years). Eighteen fractures were treated by surgical fixation with anterior transodontoid screws, since they either were not dislocated or were backward dislocated by less than 7 mm. Seven fractures were treated by anterior surgical fixation with both transodontoid screws and C1–C2 transarticular screws, since five of them were dislocated by more than 7 mm and two of them were associated with Jefferson fractures.

Surgical Procedure

Each patient was placed in a supine position and the cervical spine was hyperextended to reduce odontoid dislocation and facilitate the anterior cervical oblique screw trajectory. More accurate screw placement can be obtained by use of biplanar fluoroscopic guidance. A transverse skin incision was performed between C5 and C6, then the C2–C3 intervertebral disc was exposed. To facilitate the insertion of a K-wire with a guide through the oblique trajectory, the superior central portion of the C3 body was drilled. Then the superior central portion of the C2–C3 annulus was removed to expose the inferior lip of the C2 body. With use of fluoroscopic guidance, a K-wire was placed with a guide through the inferior edge of the C2 body, facing the disc space and through the fracture line and the apical cortex of the odontoid fracture fragment. Then the drill bit was inserted through the C2 body and a cannulated titanium screw was placed into the tip of the odontoid process, through the apical cortex. To ensure solid fixation, bicortical purchase is suggested. Autologous fibrin glue was used to enhance haemostasis and fusion [7, 10–12]. When instability of the C1–C2 joint was suspected, a screw might be inserted, with an oblique and middle lateral trajectory, blocking both of the C1–C2 joints. All procedures were performed by the spinal surgeons using the same technique. For each procedure, different indexes were retrospectively evaluated: the measured outcomes included the operating time, the length of hospital stay and the patient's radiological exposure.

Results

The follow-up duration ranged from 4 months to 5 years. Among the patients who were treated surgically, there was only one death, which was due to a complicated odontoid fracture with concomitant traumatic brain injury. No neurological deficits attributable to the surgical procedure were noted. In the early postoperative course, mild dysphagia was noted in 11 patients as the only adverse event, and it required no treatment other than dietary modification. Stable osseous fusion was achieved in 21 patients and fibrous union in one.

The mean surgical time was 98 min (range 64–144 min), and the mean fluoroscopy time was 36.6 s (range 29.8–58.8 s). A linear regression analysis showed an association between the case number and the operating time: the higher the case number, the shorter the operating time.

Among the 25 cases of type II odontoid fracture treated by anterior transodontoid screw fixation, the first ten cases had a mean operating time of 114 min (range 96–144 min). In contrast, the latter 15 cases had a shorter mean operating time of 86 min (range 64–104 min). It can be shown that the first ten cases corresponded to the learning phase of the surgeon's learning curve, when his skills were improving swiftly and the operative time shortened proportionately. During the latter 15 cases, no shortening of the operating time was observed. In regard to the radiological exposure, in the first ten cases the mean exposure time was 46.1 s (range 42.6–58.8 s), whereas in the latter 15 cases it was only 31.2 s (range 29.8–37.2 s).

Discussion

Although surgical treatment of type II odontoid fractures is still a matter of debate, anterior transodontoid screw placement is the treatment of choice to ensure direct osteosynthesis and potentially reduce loss of C1–C2 motion [13]. Several surgical techniques have been investigated over the years to assess their learning curves. In particular, different spinal surgery techniques have been developed in recent decades and have been validated in the light of their learning curves [8, 14–16]. The term "learning curve" began to be used during the 1970s and defines the acquisition process of a new surgical technique [15]. On average, the acquisition of a new technique progresses swiftly at the early stage of the learning curve and gradually and asymptotically slows down at the steady state of the proficient stage. At this stage of the learning curve, the operating time, which has understandably shortened during the early stage, according to the surgeon's experience, tends to decrease its trend, reaching an asymptote [15]. The operating time is generally advocated as a reference in assessment of a learning curve, since it depends mostly on the surgeon's skills, although additional factors (such as anaesthetic complications and the input of the surgical team, theatre nurses and fluoroscopy technicians) can influence it.

In our retrospective study of 25 consecutive anterior odontoid fixation procedures, a sharp decrease in the operating time was evidenced during the first ten cases. After that, the pace decreased its trend, reaching an asymptotic steady state during the latter 15 cases. According to the results of our lin-

ear regression analysis, there was a clear end point at the tenth case, at which stage a surgeon could be deemed proficient. We consider that the operating time at the first stage of the learning curve was longer because of the surgeon's carefulness in preventing complications. When the surgeon became confident with the procedure, the operating time progressively shortened until differences in the operating time depended no longer on the surgeon's skills but only on external factors. According to our results, this happened after the tenth anterior transodontoid screw fixation procedure.

As a further predictor of surgical skill acquisition and progression of the learning curve, we also retrospectively evaluated the radiological exposure time for each of the 25 treated patients. Since the operating time can be influenced by several external factors, we assumed that the radiological exposure time would be most precisely related to the proficiency of the surgeon and would indirectly reflect the patient's safety and the invasiveness of the surgical procedure. Similarly, after the first stage, during which the radiological exposure time shortened sharply, our retrospective analysis evidenced a clear turning point at the tenth odontoid fixation in the linear regression analysis, since a progressive decrease in the shortening trend was noted. This evidence suggests that the operative learning curve for a spinal surgeon performing anterior transodontoid screw fixation can be assumed to be complete after ten type II odontoid fracture fixation procedures. During this learning period the presence of a senior surgeon can ensure rapid and effective development of the young surgeon's skills with ample safety for the patient, until the young surgeon becomes self-confident and proficient with the procedure [14]. However, complications can occur even when a surgeon has mastered the techniques [15]. In order to overcome these, efforts to refine the technique should be continued even during the last stage of the learning curve of the surgeon, who—like a ship's captain—sails alone in the sea of spinal surgery.

Conclusion

Anterior transodontoid screw fixation is a life-saving surgical procedure for treatment of unstable type II odontoid fractures. Like other new procedures, it is characterized by a two-stage learning curve. At the first swift stage, the operative time understandably shortens on every surgical occasion, and it can be described as the "learning stage"; the second stage is characterized by a levelling-off of the decrease in the operative time, due to the surgeon's skill acquisition, and it can be described as the "proficient stage". In our retrospective analysis, the turning point appeared to be the learner surgeon's tenth anterior screw fixation procedure for odontoid fracture. To make the first learning stage proceed swiftly and safely, we advise young surgeons to be accompanied by an elder proficient surgeon during the procedure.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Functional Outcome After Odontoid Fractures in the Elderly

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Abstract While several papers on mortality and the fusion rate in elderly patients treated surgically or non-surgically for odontoid fractures exist, little information is available on quality of life after treatment. The aim of treatment in these patients should not be fracture healing alone but also quality of life improvement.

A literature search using PubMed identified seven papers including information on functional evaluation of 402 patients.

Patients treated with anterior screw fixation had a good functional outcome in 92.6% of cases. This percentage seemed to decrease in octogenarians. Less information was available for patients treated with posterior approaches; it would seem that up to a half of such patients experienced pain and limitations in activities of daily living after surgery. Patients treated with a halo device had a functional outcome that was worse (or at least no better) than that of patients treated with surgery, with absence of limitations in activities of daily living in 77.3% of patients. Patients treated with a collar had a good functional outcome in the majority of cases, with absence of limitations in activities of daily living in 89% of patients.

More studies are needed for evaluation of functional outcome, especially in patients treated with a collar, a halo device or a posterior approach.

Keywords Odontoid fracture · Elderly patients · Outcome evaluation · Functional outcome · Treatment · Surgery · Collar · Halo

Introduction

The *Global Health and Aging* report presented by the World Health Organization states that the number of people aged ≥65 years is growing from about 524 million in 2010 to about 1.5 billion in 2050, especially in developing countries [1].

Patients in this age group are at high risk of cervical spine injuries, especially at the C0–C2 level (50% of cases with cervical spine injury) [1]. While Anderson type I and III odontoid fractures are mainly treated conservatively, the treatment strategy for type II odontoid fractures represents a much discussed topic among experts in the field.

In fact, because of the presence of osteoporotic bone, comorbidities and a watershed area for blood supply, solid fusion of type II fractures in these patients is not the rule.

The reported fusion rate in surgically treated patients is 85–100% [2–9]. Some authors have reported a pseudarthrosis rate of 85% in patients treated conservatively [10].

A recent review comparing elderly patients treated for odontoid fractures found that when posterior surgery and anterior surgery were compared, the short-term mortality, long-term mortality and complication rates did not differ significantly [8].

Indeed, when patients treated with a halo device were compared with those treated with a hard collar, no significant differences in short-term mortality or complication rates were identified [8].

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Moreover, fracture fusion itself does not necessarily mean a good patient outcome in terms of quality of life.

While several papers on morbidity, mortality and fusion rates in elderly patients treated surgically or non-surgically for odontoid fractures have been published in recent years, little information is available on quality of life after fracture treatment.

The aim of this review was to obtain information on functional outcome after treatment of odontoid fractures in the elderly by comparing the available studies on this topic.

Materials and Methods

A systematic literature search using PubMed (www.pubmed.gov) was performed on 5 January 2017. The search terms included the pathology ('odontoid fracture', 'upper cervical fracture', 'axis fracture'), the patients ('elderly', 'geriatric population', 'octogenarians') and the outcome evaluation ('quality of life', 'Neck Disability Index', 'outcome', 'Barthel Index', 'SF-36', 'Smiley–Webster scale'). The Boolean operators 'OR' and 'AND' were used. No language or publication year restrictions were used.

Nine-hundred and one papers were initially identified (see Table 1 for details). The exclusion criteria included abstracts, editorials, letters, case reports, case series of fewer than five patients, review articles, meta-analysis articles, studies focused on non-human subjects, non-clinical studies, unrelated studies and studies that did not contain treatment information. After an initial review of the 901 articles, 894 papers were excluded. The reference lists of the included articles were reviewed, and no further papers were identified from them. Therefore, our review finally yielded seven articles (Table 2).

Table 1 Search strategy

Search n	Queries	Items found
#1	odontoid fracture OR dens fracture OR dens rupture OR odontoid rupture OR C2 rupture OR C2 fracture OR axis fracture OR axis rupture OR upper cervical spine fracture	5473
#2	elderly OR elderly patients OR elderly people OR elderly 65 OR elderly 70 OR elderly 75 OR elderly 80 OR elderly 85 OR elderly 90 OR elderly 95 OR elderly 100 OR octogenarians OR geriatric population	4555233
#3	neck disability index OR mortality OR outcome OR barthel index OR functional evaluation barthel OR Quality of life OR Robinson criteria OR Smiley-Webster scale OR Cervical spine outcomes questionnaire OR SF-36 OR activities of daily living	2436839
#4	#1 AND #2 AND #3	901

Results

The seven papers we identified included information on 402 patients. The characteristics of these studies are summarized in Table 2. All of these papers were published after 2007 and included elderly patients treated for type II dens fracture. Most patients were treated with anterior screw fixation (in six of the seven papers; 163 cases) [2–4, 6, 9, 11]. One-hundred and five patients were treated with a posterior approach: 80 patients with the Harms technique, seven patients with the Magerl technique, two patients with another posterior technique and 16 patients with C1–C2 wires and bone fixation) [5, 9]. Fifty-four patients were treated with halo immobilization [4, 9] (in one series an additional 16 patients received a halo device after posterior C1–C2 wire and bone fixation) [5], and 80 patients were treated with a collar [5, 9]. The mortality rate was quite high, ranging from 4.2% to 25.8%. The mortality rates ranged from 9.7% to 33.3% in surgically treated patients and from 6.3% to 25.8% in patients treated with a collar or a halo device.

The complication rates ranged from 12.5% to 31% with surgery and from 18% to 36% in patients treated with a collar or a halo device. The fusion rate was higher in patients treated with surgery.

Regarding functional evaluation, several evaluation tools were used: the Smiley–Webster scale/Robinson criteria in three papers [2, 3, 7], the Cervical Spine Outcome Questionnaire (CSOQ) in two papers by the same authors [4, 5], the Neck Disability Index (NDI) and the 36-Item Short Form Survey (SF-36) [9] activities of daily living and pain evaluations.

Three of the seven papers dealing with anterior screw fixation used the Smiley-Webster scale/Robinson criteria for functional evaluation: taken together, these studies analysed the outcome of 95 patients [2, 3, 7]. Most patients (89 of 95 cases, 93.7%) had an excellent/good outcome. Only six patients had a fair or poor outcome (6.3%); four of these patients were in the series reported by Henaux et al., which included only patients over 80 years of age [2].

The series reported by Kohlhof et al. also included patients treated with anterior screw fixation [6]. These authors used a subjective evaluation tool; they concluded that all 24 patients in their series had no pain at 6-month follow-up and that all patients returned to their previous daily activities. In the paper by Joestl et al., 29 of 32 patients treated with anterior screw fixation survived for at least

Table 2 Published series on functional evaluation after treatment of elderly patients with odontoid fracture

Author (year)	No of patients	Age	Fracture type	Treatment	Outcome: short-term mortality	Outcome: long-term mortality	Complication rate	Fusion rate	Functional evaluation	Functional outcome
Platzer (2007) [7]	41	≥65	II	Ant screw	9.7%	NR	22%	88%	Smiley-Webster, pain evaluation, ADL	mean Smiley-Webster score: 1.50; Pain frequency: 5/41–12.2%; Severe pain: 3/41–7.3%; limitations in ADL: 5/41–12.2%; Movement restrictions 4/41–9.7%
Hou (2011) [3]	43	≥65	II	Ant screw	NR	18.6%	NR	85.7%	Robinson criteria (same as Smiley-Webster), ROM	Robinson: Excellent 17 (39.5%), good 24 (55.8%), fair 2 (4.7%). ROM: 5 pts-11.6%-less than 25% ROM limitations; 3 pts-6.9%-more than 25% ROM limitations
Henaux (2012) [2]	11	≥80	II	Ant screw	0	18%	0	44.4% (fibrosis in other cases)	Smiley-Webster	Smiley-Webster: Excellent 5 (45.5%), good 2 (18.2%), fair 1 (9%), poor 3 (27.3%)
Kohlhof (2013) [6]	24	62–98	II	Ant screw and cement	2/24 (8.3%)	8/24 (33.3%)	12.5%	84.2%	Subjective evaluation	6 month: none complained pain. All pts returned to normal activities
Vaccaro (2013) [9]	159	≥65	II	101 surgery (12 ant screw, 80 C1C2 Harms 7 C1C2 Magerl 2 Other) 58 conservative (6 Halo, 52 collar)	NR	20 Month: 13.9% surgical group, 25.8% conservative group	30% surgical group 36% nonsurgical group	95% surgical 79% conservative	NDI, SF36	NDI: better in surgical group (28vs 31.6 at 12 month f-up); NDI worsened in both groups: 14.7 points in conservative group and 5.7 points in surgical group. SF-36 better in surgical group
Joestl (2016) [4]	80	≥65	II	32 Ant screw vs 48 Halo	12.5% ant screw, 6.2% halo	12.5% ant screw, 6.2% halo	28% ant screw vs 25% Halo	90% ant screw, 77% Halo	Cervical Spine Outcome Questionnaire	81% returned to previous activities. Ant screw group: better pain severity, disability and phychological distress; no difference in physical symptoms
Joestl (2016) [5]	44	≥65	II Non-unions after previous treatment	16pts: posterior C1C2wires and bone and Halo; 28 pts collar			23% (31% surgical group, 18% collar group)	100% fusion in surgical group, 92.8% preudoarthrosis in conservative group	Cervical Spine Outcome Questionnaire, Pain frequency, Pain severity, ADL	ROM impairment: 5 patients (31.5%) surgery vs 2 pts (7%) conservative; Occasional pain: 5 pts (31%) surgery, 5 pts (18%) conservative; Regular Pain: 2 pts (12.5%) surgery, 0 conservative; Severe pain: 2 pts (12.5%) surgery, 0 conservative; ADL limitations: 8 pts (50%) surgery, 3 pts (11%) conservative; functional disability and psychological distress: worse for conservative

1 month after surgery and were therefore available for follow-up functional evaluation [5]. Of these 29 patients, four (13.8%) had some limitations in certain sport/profession activities and five (17.2%) experienced either occasional pain (four patients) or regular pain (one patient). Patients who were treated with anterior screw fixation (12 cases) in the series reported by Vaccaro et al. could not be included in this outcome evaluation, since those authors considered patients treated with anterior screw fixation together with patients treated with posterior approaches [9]. In general, 137 of 148 patients (92.6%) treated with anterior screw fixation from the series reported by Platzer et al., Hou et al., Henaux et al., Kohlhof et al. and Joestl et al. had a good outcome (as measured by 'excellent' and 'good' scores on the Smiley–Webster/Robinson scales, 'absent' or 'rare' pain evaluations and a return to previous activities) [2–4, 6, 7].

Regarding patients treated with posterior approaches, information on functional outcome was available on 89 patients reported by Vaccaro et al. and 16 patients reported by Joestl et al. [5, 9]. Nonetheless, these two series could not be compared with each other, since the evaluation tools they used were markedly different. Moreover, Vaccaro et al. evaluated the outcome of patients treated with posterior approaches (89 patients) and with anterior screw fixation (12 cases). They observed that both the NDI and the SF-36v2 Bodily Pain outcomes remained significantly better in the surgical group compared with those in the nonsurgical group [9]. Of the patients treated with a posterior approach in the series reported by Joestl et al., 5/16 patients (31.5%) had range-of-movement (ROM) impairment, 5/16 patients had occasional pain and 2/16 patients (12.5%) had regular severe pain. Moreover, 8/16 patients (50%) had limitations in activities of daily living [5].

The remaining patients in these series reported by Vaccaro et al. [9] and Joestl et al. [4, 5] were treated conservatively with a halo device or a collar. In the series reported by Joestl et al., 10/44 patients (22.7%) treated with a halo device had limitations in daily activities. Of these patients, three (6.8%) had limitations in all daily activities. Occasional pain was experienced by 7/44 patients (15.9%) and regular pain by 2/44 patients (4.5%) [4].

In another series reported by the same authors [5], 3/28 patients (11%) treated with a collar had no severe pain but experienced limitations in some daily activities.

Another series we did not include in this analysis was reported by Butler et al. [12]. Using the CSOQ, these authors compared patients younger and older than 65 years who were treated with a halo device or a collar; the series included 14 patients over 65 years of age. The authors observed that older patients had worse physical symptoms, functional disabilities and psychological distress. Unfortunately, the authors did not specify how many patients were treated with a halo device or a collar.

Discussion and Conclusion

While several papers discussing morbidity, mortality and fusion rates in elderly patients treated for dens fracture have been published, little information is available on the functional outcome of these patients. Elderly patients often have pre-existing comorbidities and several limitations in activities of daily living; therefore, both the trauma causing an odontoid fracture and the fracture treatment itself may result in further impairment of quality of life. Healing of a bony fracture alone cannot be considered a success if the treatment causes a deterioration in the quality of life. Our literature search revealed that only seven papers analysed the functional outcome of elderly patients treated for fractures of the odontoid process.

The outcome tools that were used in these studies were the SF-36, NDI, CSOQ and Smiley–Webster scale/Robinson criteria activities of daily living.

The NDI is similar to the Oswestry Low Back Pain Disability Index. The NDI is a questionnaire with ten items: personal care, pain, headaches, reading, concentration, lifting, work, recreation, sleeping and driving. The scores range from 100 (the worst score) to zero (the best score) [13].

The SF-36 is widely used in geriatric populations, since it is a tool measuring the patient-reported multidimensional health status. It describes the health status in terms of eight items: physical functioning, physical limitation, general health, pain, emotional status (well-being), role limitation–emotional, social activities and energy/fatigue. The final scores range from 100 ('no disability') to zero ('maximum disability') [14].

The CSOQ is a tool designed and used for assessing the outcome of patients with neck pain. It includes information on pain severity, physical symptoms, demographics, functional and psychological disability, healthcare utilization and general satisfaction [15]. Like the SF-36, this tool is not simple to administer and requires input from trained doctors.

The Smiley–Webster scale (referred to as the modified Robinson criteria in the paper by Hou et al. [3]) is a very easy scoring system to use. It assesses patient outcome by combining pain and return to former activity levels. The outcome is expressed as 'excellent', 'good', 'fair' or 'poor'.

The objective of our study was not to compare the ease of use and appropriateness of the different evaluation scales. Nonetheless, it is our opinion that evaluation tools should have a clinical impact on treatment decision-making and should be easily explainable to patients before treatment. A question frequently asked by patients before surgery is 'What is the percentage of success?' not 'How much will the mean score of this tool increase after treatment?' Some of the aforementioned scales have been created and validated for

other purposes where a multidimensional evaluation of elderly patients was needed.

Our literature review revealed that patients treated with anterior screw fixation had a good functional outcome in the great majority of cases (92.6%). This percentage seemed to decrease in octogenarians.

Less information was available regarding patients treated with posterior approaches, but it would seem that up to half of these patients experienced limitations in activities of daily living after surgery.

In this analysis the total number of patients treated non-surgically was 134 (80 patients treated with a collar and 54 patients treated with a halo device). Moreover, it was not always possible to compare patients treated with a halo device and those treated with a collar, as some authors analysed the outcome of these two groups together. Nonetheless, it would seem that patients treated with a halo device had a functional outcome that was worse (or at least no better) than that of patients treated with surgery [9], with no limitations in activities of daily living being reported in 77.3% of patients in the series reported by Joestl et al. [4].

The only available series analysing patients treated with a collar, which included just 28 patients, revealed that the functional outcome was good in the majority of cases, with no limitations in activities of daily living in 89% of patients—comparable to the outcome of patients treated with anterior screw fixation [5].

Our review revealed that the available studies on elderly patients with type II odontoid fractures did not compare the functional outcome of different approaches (surgical and non-surgical), since different and non-comparable outcome evaluation tools were used and different treatment modalities were analysed together [9].

Another limitation of this review was that the follow-up varied widely in these series. We have therefore referred to outcomes as described by the authors of the papers.

In conclusion, it seems that patients treated with anterior screw fixation had the best functional outcome, as did patients treated with a collar. Patients treated with a halo device seemed to have the worst functional outcome.

Further studies are needed to confirm these findings. Such studies should evaluate patients separately, using easily comparable scales (such as the Smiley–Webster scale). The most complicated scales (the NDI, CSOQ and SF-36) should be categorized in order to define groups of patients with excellent, good, fair or poor outcomes. Moreover, more studies are needed to evaluate the outcome of elderly patients treated with a collar, a halo device or a posterior approach.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards Yes financial support was received for this work.

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CVJ Infections

Management of Craniovertebral Junction Tuberculosis Presenting with Atlantoaxial Dislocation

Granit Molliqaj, Philipp Dammann, Karl Schaller, Ulrich Sure, and Enrico Tessitore

Abstract Tuberculosis (TB) rarely involves the craniovertebral junction (CVJ). Atlantoaxial dislocation (AAD) is one of the most commonly encountered lesions in craniocervical TB. The incidence of TB and its craniovertebral manifestation is increasing even in developed countries because of intercontinental migration and increased prevalence rates of immunosuppression conditions. While the treatment of craniovertebral TB is well standardized and relies on conservative measures, the treatment of TB with AAD is disputable. In this paper we present a review of the literature and elucidate our approach to craniovertebral TB with AAD through a case illustration.

Keywords Tuberculosis · Craniovertebral junction · Atlantoaxial dislocation · Craniocervical fixation

Introduction

Tuberculosis (TB) rarely manifests at the level of the spine (occurring in fewer than 1% of TB cases [1]). Involvement of the craniovertebral junction (CVJ) in TB is even rarer and occurs in only 0.3–1% of all spinal TB cases [2]. However, with 10.4 million new incident cases of TB worldwide (of which 580,000 are multidrug resistant), the TB epidemic is larger than previously estimated, according to a 2016 World Health Organization report [3]. With eastern hemisphere countries accounting for the majority of new cases, western hemisphere countries face significantly smaller numbers of TB cases. Nevertheless, the incidence of CVJ-TB is increasing even in western countries because of intercontinental migration and increased prevalence rates of acquired immune deficiency syndrome [4, 5].

Atlantoaxial dislocation (AAD) is the most commonly observed CVJ lesion in patients with TB [6]. It is associated with destruction of bone, ligaments and the articular process, causing local instability [7].

While treatments for CVJ-TB without spinal instability are well standardized and rely on conservative measures such as external immobilization and antibiotics, the treatment of cases with AAD is still controversial, ranging from collar immobilization to surgery. In this paper we present a review of the literature and elucidate our approach to CVJ-TB with AAD through a case illustration.

Materials and Methods

A search for all relevant PubMed-listed publications from 1990 to the present was performed, using the following search terms: ‘tuberculosis’, ‘craniocervical junction’, ‘craniovertebral junction’ and ‘atlantoaxial dislocation’. Only English-language literature was chosen. If relevant studies were found in the reference lists of selected articles, they were also included if they met the following inclusion criteria: either (1) clinical series of at least 20 adult patients with CVJ-TB with a standardized treatment algorithm and standardized outcome assessment; or (2) systematic reviews of CVJ-TB.

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Results

A total of 55 relevant articles were found. Of these, five papers [4, 6, 8–10] met our inclusion criteria (see Table 1). The five clinical series comprised analysis of a total of 197 patients affected by CVJ-TB (see Table 2).

The initial clinical findings corresponded in all series. Neck pain was the dominant initial symptom. Signs of myelopathy were frequently present (occurring in 48–80% of patients). Radiographic evidence of AAD, which is defined by an atlantodental interval (ADI) >3 mm, occurred in 54–100% of patients in the different series [4, 6, 8–10]. Basilar invagination is defined by the presence of the odontoid tip above the McRae line drawn on a lateral skull radiograph or in a midsagittal view on a computed tomography (CT) scan. The initial grading of TB at the CVJ differed in all studies. Arora et al. [8] used the disability grading for CVJ malformation proposed by Di Lorenzo [11], combined with the radiological grading system proposed by Lifeso (see below) [2]. The Di Lorenzo system is based on assessment of neurological deficits and disability: grade 1 corresponds to neck pain only; grade 2 represents an autonomous patient with a minor disability; at grade 3 the patient is not fully independent in terms of daily life activities (DLA); and at grade 4 the patient is completely dependent in terms of ADL [11]. Behari et al. [9] also used the Di Lorenzo grading. Gupta et al. used a combination of the Nurick disability scale and their own specific radiological grading system [6]. Shukla et al. [4] utilized the grading system proposed by Lifeso (see below) [2]. Teegala et al. [10] proposed their own grading system based on a neck movement score, motor score and radiological score (assessing bony destruction, cord compression and retropharyngeal collection). Corresponding to the different grading systems used, the descriptions of preoperative radiological findings varied widely.

The postoperative clinical outcome was defined in all series as improvement, stability or deterioration of functional scores over time [4, 6, 8–10]. Common disability scoring systems (the Nurick scale, Frankel scale and Ranawat score) are also routinely used in assessment of patients with CVJ-TB [9, 10, 12–14]. An improved outcome was observed in the majority of patients (91–100%); however, it was not further detailed in most series. Furthermore, complete relief of initial neck pain was mentioned in all series, accounting for all patients. Mortality ranged between 0% and 4%, in all cases being attributed to general progression of systemic TB (see Table 2).

The recommended treatment algorithms for CVJ-TB varied in the different series [4, 6, 8–10]. The majority of treatment decisions were based on the presence of neurological deficits, associated anterior compression (with a granuloma or abscess) and reducible/non-reducible AAD [4, 9, 10]. Gupta et al. considered only the presence of AAD and bony alterations, proposing a specific grading system on this basis [6].

Table 1 Number of articles and inclusion criteria

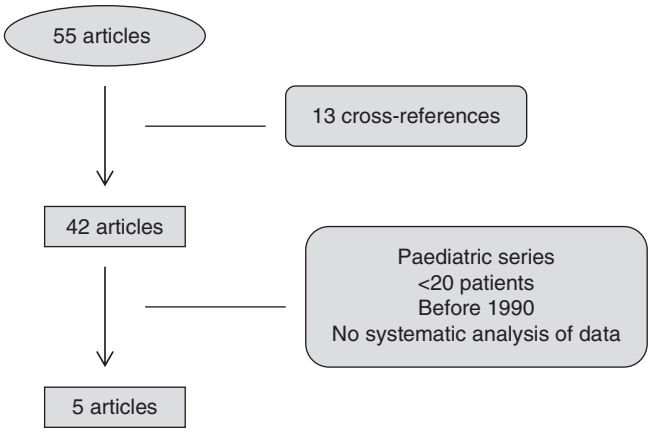

Table 2 Recent series of craniovertebral tuberculosis

Authors	Year	Cases	Period (years)	Symptoms neck pain	Symptoms myelopathy	Grading preop.	AAD	ATT ent	Mortality	Neurological outcome improved or stable
Behari et al. [9]	2003	25	8	25/25 (100%)	18/25 (72%)	Disability score (1–4)	17/25 (68%)	All	1/25 (4%)	24/25 (96%)
Shukla et al. [4]	2005	24	14	24/24 (100%)	16/24 (67%)	Radiological score	14/24 (58%)	All	0/24 (0%)	24/24 (100%)
Gupta et al. [6]	2006	51	27	48/51 (94%)	41/51 (80%)	Radiological score	51/51 (100%)	All	1/51 (2%)	48/52 (91%)
Teegala et al. [10]	2008	71	9	71/71 (100%)	34/71 (48%)	Combined score	38/71 (54%)	All	0/71 (0%)	71/71 (100%)
Arora et al. [8]	2011	26	6	26/26 (100%)	17/26 (65%)	Combined score	17/26 (65%)	All	0/26 (0%)	26/26 (100%)

AAD atlantoaxial dislocation, *ATT* anti-tuberculous therapy

Discussion

Radiological Features and Classification Systems

Over time, progression of TB at the CVJ may lead to atlantoaxial instability and dislocation. A summary of typical radiological findings in CVJ-TB can be found elsewhere [15]. Goel et al. recently described the evolution of the disease in three stages, emphasizing that bony structures are the first ones to be destroyed, causing displacement and subsequent rupture of ligaments [7]. The initial stage is characterized by unilateral implication of the facet of C2 without destructive deformation, causing mainly neck pain. At the second stage, the disease progresses with destruction of the atlantoaxial joint by inflammation and necrosis, extending over the rest of the C1 and C2 vertebra and leading to ineffectiveness of the transverse and alar ligaments through destruction of their bony attachments. Symptoms include neck pain, muscle spasms, torticollis and potential neurological deficits. At the third stage, the contralateral atlantoaxial joint is involved, and AAD with a neurological deficit is frequently seen.

In 1987, Lifeso et al. [2] proposed a three-stage radiological classification system (including treatment recommendations). At stage 1 there is ligamentous integrity but bony destruction is discrete, without signs of C1–C2 dislocation. Slight odontoid invagination may be present. At stage 2 there is AAD including discrete bony destruction and ligament rupture, with or without C2 invagination of the odontoid process. Patients at this stage may require fusion. At stage 3 there is AAD with significant bony destruction and closure of the anterior arch of the axis. Fixation may be required between the occiput, the axis and C3 [13].

Apart from having the potential to show CVJ instability, magnetic resonance imaging (MRI) or CT scan findings are mandatory to confirm suspicion of CVJ-TB, especially in western European countries, where the disease is extremely rare. The typical radiological presentation for TB in the CVJ includes destruction of the vertebral body with the presence of an abscess with an irregular and large periphery, inside which there is calcification. This should differentiate TB from other inflammatory, rheumatoid, tumorous or infectious lesions of the CVJ [16].

Diagnosis

Independently of the radiological aspect of CVJ-TB, establishing a formal diagnosis usually relies on staining or culture of *Mycobacterium tuberculosis*. Most authors agree that at least a needle aspiration or biopsy should confirm the diagnosis [4, 6, 8–10, 17]. However, other diagnostic criteria exist to provide an objective diagnosis of CVJ-TB. A diagnosis can be retained when a patient presents with a cervical collection on imaging with or without instability and bony destruction. For the diagnosis to be confirmed, the patient should present with one additional major criterion or four minor additional criteria [10]. The major criteria are polymerase chain reaction positivity, and histopathological or microbiological evidence of TB. The minor criteria include radiological evidence of pulmonary TB, a positive Mantoux test result, a history of TB, an erythrocyte sedimentation rate >50 mm/h, constitutional symptoms such as weight loss or nocturnal fever, and a clinicoradiological response to antitubercular therapy (ATT).

Treatment

Prolonged systemic ATT is naturally the mandatory baseline treatment in CVJ-TB and should follow the guidelines for TB treatment [3]. Collection of pre- or paravertebral fluid (tubercular pus) by needle aspiration is also recommended (see case illustration in Sect. 4.5) and may require decompression of nervous structures [6–8]. However, surgical decompression and/or posterior fixation of the CVJ are controversial, and expert opinions on the optimal treatment have shifted in recent decades [6, 7, 10]. In our review of the five largest and most recent series, we found overall that there were two opposed treatment approaches for CVJ-TB and AAD: (1) a tailored treatment algorithm including surgical options (ranging from simple decompression to radical surgical correction) based on radiological findings and the patients' initial clinical condition; and (2) a tailored treatment algorithm relying solely on conservative measures such as traction and collar immobilization. The two algorithms essentially differ in the treatment of patients with extensive radiological changes (such as fixed or reducible AAD) and a significantly impaired neurological condition. While the former approach advocates surgical fixation (with or without decompression), the latter approach advocates long-term rigid external immobilization (with or without traction). Regarding the clinical outcomes in the presented series, equally favourable results have been reported with both approaches [4, 6, 8–10]. Overall, all authors discuss the need to balance surgical risks (especially wound healing in immune-deficient patients) with the inconvenience of long-term rigid external immobilization. Arora et al. and Gupta et al. recommend conservative treatment in all patients with CVJ-TB, regardless of the neurological condition and the

presence or absence of spinal instability [6, 8]. On the other end, Behari et al. and Shukla et al. suggest surgical treatment in patients with severe deficits as a result of spinal cord injury induced by AAD [4, 9].

Teegala et al. described an interesting approach to management of these patients. They classified patients into three grades of severity according to clinical and radiological evaluations of limitation of cervical motion, weakness and imaging of lesions [10]. Patients with a severe clinicoradiological disability (grade 3) benefited from anterior decompression and posterior fixation. Patients with grade 1 or 2 clinicoradiological disability were initially treated conservatively, but those whose condition evolved poorly or who had AAD underwent surgical treatment as well [10]. Of course, all patients were treated with a four-drug ATT regimen from the time of diagnosis.

Table 3 Craniovertebral junction tuberculosis grading system

Parameter	Score
Restriction of active neck movement score[a]	
No	1
Yes	2
Motor score	
No	1
Minimal (MRC[b] motor power ≥4)	2
Severe (MRC motor power ≤3)	3
Radiological score	
Retropharyngeal collection ≥7 mm in anteroposterior diameter at C2 body level without evidence of bone destruction or radiological instability	1
Retropharyngeal collection evidence of bone destruction (involvement of 1 Dennis vertebral column), thecal sac compression without cord compression, or cord changes	2
Severe bone destruction (involvement of >1 Dennis vertebral column), with cord compression and/or cord signal changes	3

Based on composite score, patients were divided into 3 grades in Teegala et al series [10]: Grade 1: score of 3–4; Grade 2: score of 5–6; Grade 3 score of 7–8

We propose to split the score on 2 grades: Grade 1: score <6; Grade 2 ≥6

[a]Patients who could not flex more than 50% of mentosuprasternal distance in a military erect position

[b]*MRC* Medical Research Council

Treatment Algorithm

On the basis of the literature we analysed, we propose a comprehensive treatment algorithm taking into consideration the most relevant criteria analysed in the different studies we reviewed (see Fig. 2).

We remain convinced that consideration of both clinical and radiological findings is necessary for good management. Moreover, some patients present with a discrepancy between their radiological severity and clinical performance. Having reviewed the literature we selected and analysed, as discussed above, we consider that consideration of these two aspects in the therapeutic decision-making tree may avoid delays in diagnosing patients and offering them adequate treatment to avoid morbidity associated with long-term neck brace immobilization, appearance of new neurological deficits or death induced by neural squeeze. This algorithm is based on our interpretation of the existing literature, with adaptation to the patient's clinical situation and severity of illness, in order to standardize treatment of CVJ-TB on the basis of integration and consideration of the current data.

Like Teegala et al., we propose to first evaluate disease severity at the outset by using their clinicoradiological grading system [10]. This grading system assesses limitation of cervical motion, weakness and imaging of lesions (see Table 3). However, we propose to simplify the grading by dividing patients into two groups on the basis of their clinicoradiological score. Patients with a score ≥6 should undergo surgical treatment, while patients with a score <6 should receive conservative treatment.

According to this scoring system, a score of ≥6 points means that the radiological and clinical situation has to be considered severe. To obtain a score of ≥6 the patient must at least have a motor deficit and bony destruction, with a risk of cervical stenosis. Teegala et al. noted that some patients with a score of 5–6 were initially treated conservatively but subsequently required surgery for their residual disease. To avoid the occurrence of a new neurological deficit after the diagnosis is made, and to minimize the need to compromise the patient's daily life with long-term neck brace immobilization, we consider that patients with a score of ≥6 could safely benefit from surgical treatment. Of course, if we take as reference points the clinical series of Arora et al. and Gupta et al., we can concede that some patients would be 'overtreated' with our algorithm [6, 8].

In patients with a clinicoradiological score <6, conservative management should be tried, comprising an ATT regimen for at least 6 months and neck stabilization with a rigid collar the time that bone fusion occurs on follow-up at 3 weeks, 6 weeks, 3 months, 6 months and 1 year. Any patient who shows no evidence of clinicoradiological

improvement or who presents with neurological impairment will have to join the surgical treatment arm. Patients with a clinicoradiological score ≥6 should undergo surgical treatment. The type of surgical treatment will depend on the status of cervical stability. In our opinion, posterior fusion surgery should be reserved for patients presenting with clear spinal instability (AAD) and major bone/ligamentous disruption to prevent further spine misalignment and neurological deterioration, and to reduce instability-related pain. If AAD is reducible by traction, surgical treatment should be limited to posterior fixation involving the destroyed segments. In the event of condyle involvement and disruption, the occiput should be included in the fixation (see case illustration in Sect. 4.5). If the AAD is fixed, a posterior approach should be followed by transoral or transnasal odontoid resection for anterior decompression. We recommend immobilization in a hard collar for 6 weeks after the surgical procedure. Any attempts to conservatively treat cases with AAD and instability may result in a secondary deformity, which may lead to chronic neck pain. Of course, all patients must be prescribed a four-drug ATT regimen from the outset in accordance with infectious disease guidelines.

Case Illustration

A 35-year-old woman, originally from Pakistan, presented with neck pain and severe tetraparesis. A CT scan showed a vertical AAD, cranial settling of the odontoid process (which was beyond the McRae line and compressing the spinal cord) and lateral subluxation to the left (Fig. 1a, b). The anteriorly displaced C1 posterior arch impinged on the spinal cord as well. The left C1 lateral mass and condyle were completely destroyed, such as the transverse ligament. MRI confirmed spinal cord compression and showed tissue with intense contrast enhancement surrounding the atlantoaxial region and a prevertebral retropharyngeal abscess (Fig. 1c, d). The abscess was transorally drained by an ear, nose and throat (ENT) surgeon (using open drainage), and tissue sampling confirmed TB. The patient was then submitted to gentle traction with Gardner tongs with a 3 kg weight, which allowed partial reduction of the cranial settling (reducible AAD) and improvement of the patient's neurological condition. Then, posterior C1 arch decompression and occiput–C4 stabilization and fusion were performed. The patient was prescribed antibiotic treatment with isoniazid, ethambutol, rifampicin and pyrazinamide for a total of 6 months. CT scanning (Fig. 2a) and MRI (Fig. 2b, c) at 1-year follow-up confirmed the absence of disease recurrence and a well-decompressed spinal cord. A 2-year x-ray control showed stability of the implants (Fig. 2d).

Conclusion

Tuberculosis rarely involves the craniocervical junction. Neck pain and neurological deficits are the most common findings. Computed tomography (CT) scanning and magnetic resonance imaging (MRI) may show a retropharyngeal vertebral abscess or granulation tissue, bony and/or ligamentous disruption, and spinal instability (atlantoaxial dislocation [AAD]). While the treatment of cases with mild symptoms and instability is well defined and relies on traction, external immobilization and antibiotics, the management of patients with severe pain and neurological deficits is still controversial. We recommend surgical treatment in cases with severe AAD and signs of spinal cord compression. Posterior-only fixation is recommended in cases of reducible AAD, while patients with fixed AAD should undergo both posterior fixation and anterior decompression. Antibiotic treatment is mandatory and should be started as soon as possible in the postoperative period (Fig. 3).

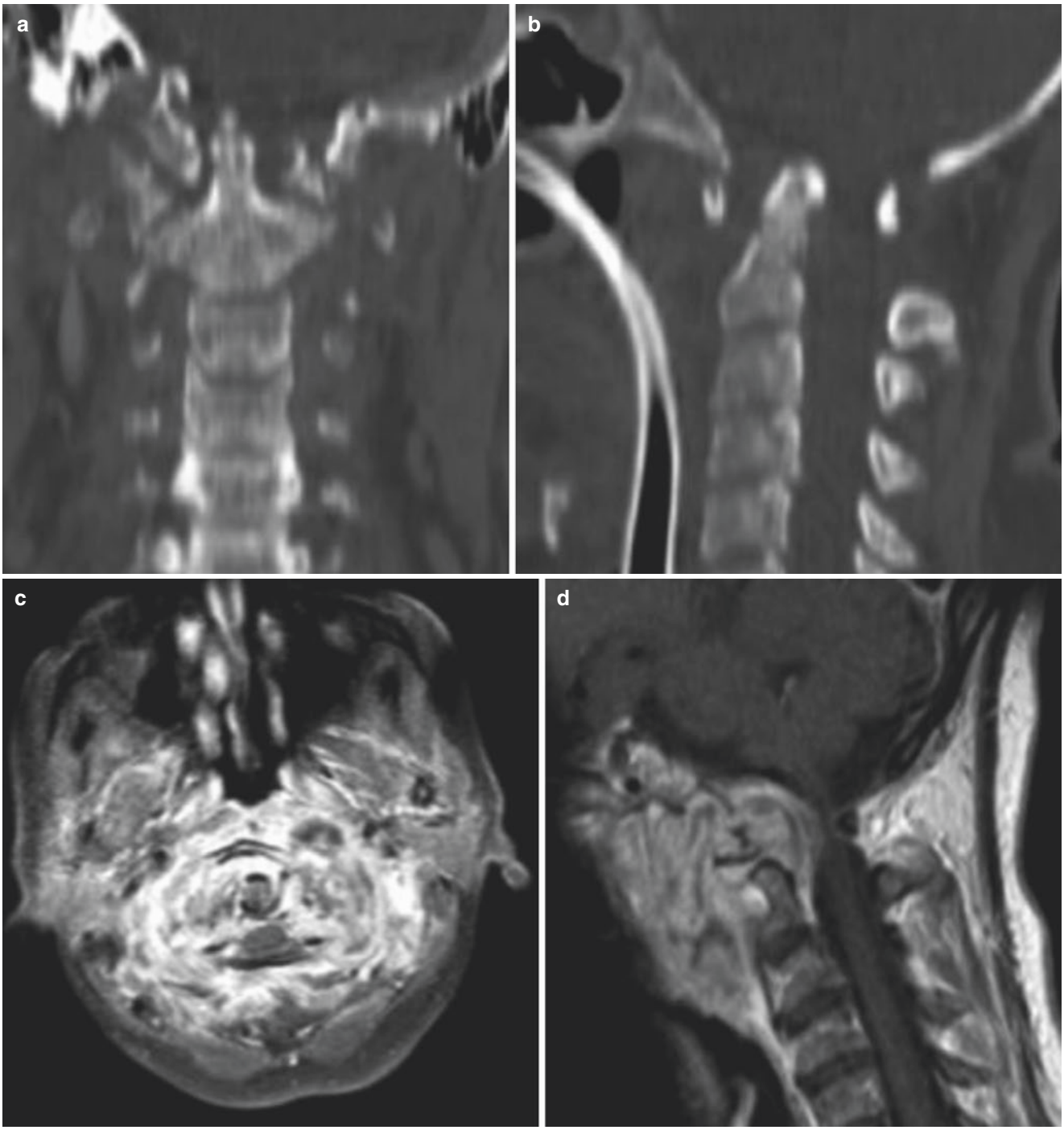

Fig. 1 (**a**) Preoperative coronal computed tomography (CT) scan showing atlantoaxial dislocation (AAD) with lateral subluxation caused by a left C1 lateral mass and condyle destruction. (**b**) Preoperative sagittal CT scan showing AAD with odontoid cranial settling (the odontoid tip is well above the McRae line). (**c**) Axial T1 gadolinium-enhanced magnetic resonance image (MRI) showing atlantoaxial and soft tissue infiltration. (**d**) Sagittal T1 gadolinium-enhanced MRI showing a prevertebral retropharyngeal abscess and spinal cord compression by pathological retro-odontoid soft tissue

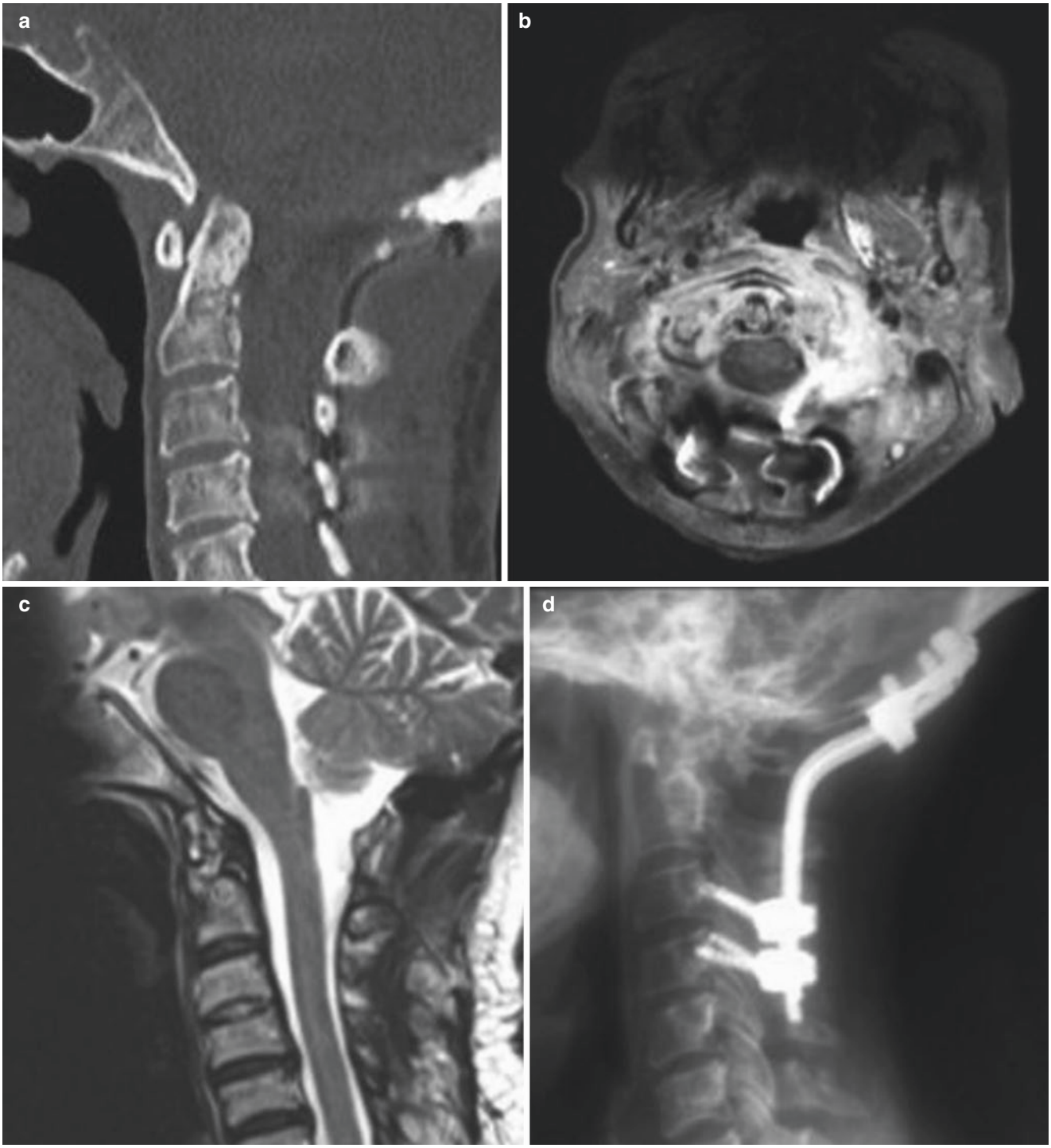

Fig. 2 (**a**) Postoperative sagittal computed tomography (CT) scan showing good alignment and posterior decompression. (**b**) Axial T1 gadolinium-enhanced magnetic resonance image (MRI) at 1-year follow-up, showing almost complete resolution of tissue infiltration. (**c**) Sagittal T2 MRI at 1-year follow-up after successful spinal cord decompression. (**d**) Two-year X-ray control showing stability of the implants and good spinal alignment

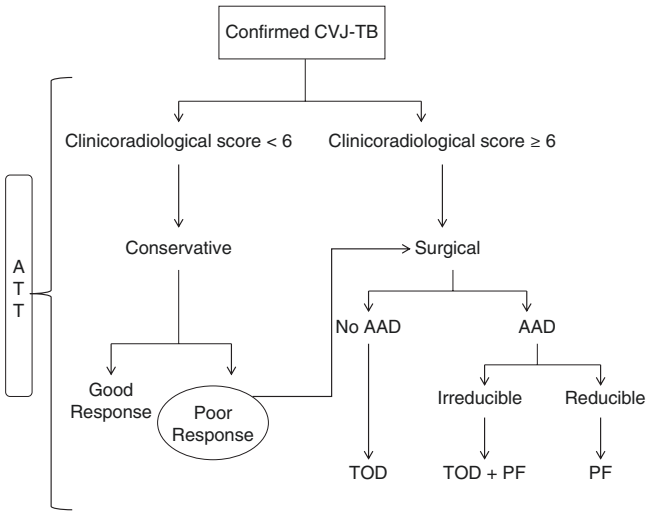

Fig. 3 Treatment algorithm flow chart showing the management approach proposed by the authors. *AAD* atlantoaxial dislocation, *ATT* antitubercular therapy, *CVJ* craniovertebral junction, *PF* posterior fixation, *TB* tuberculosis, *TOD* transoral decompression

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Extensive Spinal Epidural Abscesses Resolved with Minimally Invasive Surgery: Two Case Reports and Review of the Recent Literature

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Abstract *Purpose:* An extensive spinal epidural abscess is a rare condition and causes significant morbidity and mortality. Few authors have described this uncommon entity, which requires early diagnosis and optimal treatment to avoid devastating complications. The purpose of this study was to evaluate a minimally invasive technique for treatment of an extensive spinal epidural abscess by describing two cases. Furthermore, we conducted a review of the recent literature on the management of this rare condition.

Methods: We report two cases of spinal abscesses extending to the whole epidural space, successfully treated by use of a minimally invasive technique consisting of multilevel laminotomy and catheter irrigation to decompress and drain the epidural space.

Results: This technique is able to decompress the spinal cord, isolate the pathogen and evacuate the abscess. No complications, late spine deformity or dura penetration were observed in our patients.

Conclusion: Urgent surgical decompression, in combination with long-term antibiotic treatment, is generally considered the treatment of choice for an extensive spinal epidural abscess. A minimally invasive technique can be very useful as a surgical option.

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Keywords Extensive spinal epidural abscess · Holocord abscess · Minimally invasive surgery

Introduction

A spinal epidural abscess (SEA) is an uncommon condition with potentially devastating consequences, with a major incidence in the sixth and seventh decades of life. Historically, the reported rates of SEA have ranged from 0.2 to 1.2 cases per 10,000 hospital admissions [1]. An SEA is defined as extended when it involves more than five vertebral levels; the estimated mortality rate is around 15% [2, 3]. Nowadays the prevalence is increasing because of increases in predisposing conditions such as diabetes mellitus, intravenous drug abuse (IVDA), chronic renal failure, systemic immunodeficiency and previous spinal surgery. The aetiology is bacterial contamination due to haematogenous spread in half of all cases, contiguous spread in one third of cases and other unidentified causes in the remaining cases [4]. Approximately 50% (range 11–75%) of patients are initially misdiagnosed at the time of presentation [5, 6]. The combination of a low incidence and non-specific symptoms such as back pain or localized spinal tenderness can make early recognition difficult. Unrecognized SEA may progress not only to a potentially irreversible neurological deficit but also to life-threatening sepsis. The first cause of death is represented by septic shock. Patients with SEA often have numerous comorbidities; severe thrombocytopenia or a massive infection often worsen the patient's outcome [7–10]. The accuracy of diagnosis has been improved by the use of magnetic resonance imaging (MRI); in fact, this technique represents the gold standard for diagnosis of SEA [11]. Although rapid surgical decompression and specific antibiotic therapy make up the cornerstone of therapy for SEA, the ideal management of this condition remains controversial. We report two cases of extensive SEA treated successfully with a minimally invasive technique.

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Furthermore, we conduct a review of the recent literature to investigate the features and the kinds of treatment for this rare condition.

Case 1

A 62-year-old man presented at our emergency department with an acute exacerbation of chronic back pain and low-grade fever. The clinical examination showed no signs of neurological symptoms in the four extremities or sphincter disturbances. There were no sensory deficits. On admission the patient presented with a very limited range of motion of the lumbar spine and diffuse tenderness on palpation over his back. He had a history of previous surgical debridement for a flesh-eating disease of the right leg, arising from a total knee arthroplasty. In the previous 10 days the patient had received high-dose steroid therapy for low back pain. Laboratory tests showed a small increase in the white cell count (13,400 cells/mm^3), with an erythrocyte sedimentation rate of 57.2 mm/h (normal value 0–15 mm/h) and a C-reactive protein (CRP) level of 54.7 mg/dL (normal value 0–3 mg/dL). Plain radiographs of the cervical, thoracic and lumbar spine showed no abnormal findings. The patient clinically deteriorated over the next 6 h after admission to the spinal surgery division and showed a fever of 38.6 °C and progressive flaccid tetraplegia with subsequent breathing difficulty requiring ventilatory support. A whole-spine MRI examination was performed, showing an epidural abscess extending from the craniocervical junction (CCJ) to T8, located anterior to the cord, and from T12 to S1, located in the anterior and posterior epidural space, determining a compromise in the canal diameter and cord compression (Fig. 1). T2W sagittal sections showed L1/L2 discitis. The patient was taken to the operating room for an emergency evacuation. With use of a minimally invasive anterior approach, drainage of the upper cervical epidural abscess was performed with a 1.3-mm-diameter smooth catheter inserted from the C2–C3 disc space into the bulbar region under fluoroscopic guidance. Through a posterior midline approach, at the same time, we performed minimally invasive laminotomy and flavectomy on the left side in the T11–T12, L1–L2 and L3–L4 spaces, in an attempt to preserve the midline structures (Fig. 2). The pus was drained, under mild continuous suction, using a 2.7-mm-diameter silicone catheter, inserted caudally and cranially into the epidural space at the laminotomy sites. The catheter was inserted through each laminotomy site and advanced for up to 5 cm gently in both upward and downward directions (Fig. 3). Irrigation was performed with several litres of normal saline until clear fluid was obtained. Samples were obtained and sent for Gram staining and culture. The cervical wound was closed over a drain, which was retained for 3 days. Microbiological examinations were positive for methicillin-sensitive *Staphylococcus aureus*. Targeted antibiotic therapy was administered, and the patient showed improvements in ventilation and neurological condition. Three-month follow-up MRI studies showed complete resolution of the epidural abscess without vertebral malalignments, thus avoiding the need for fixation (Fig. 4). At 2-year follow-up, the patient had no clinical, radiological or laboratory evidence of residual or recurrent spinal infection.

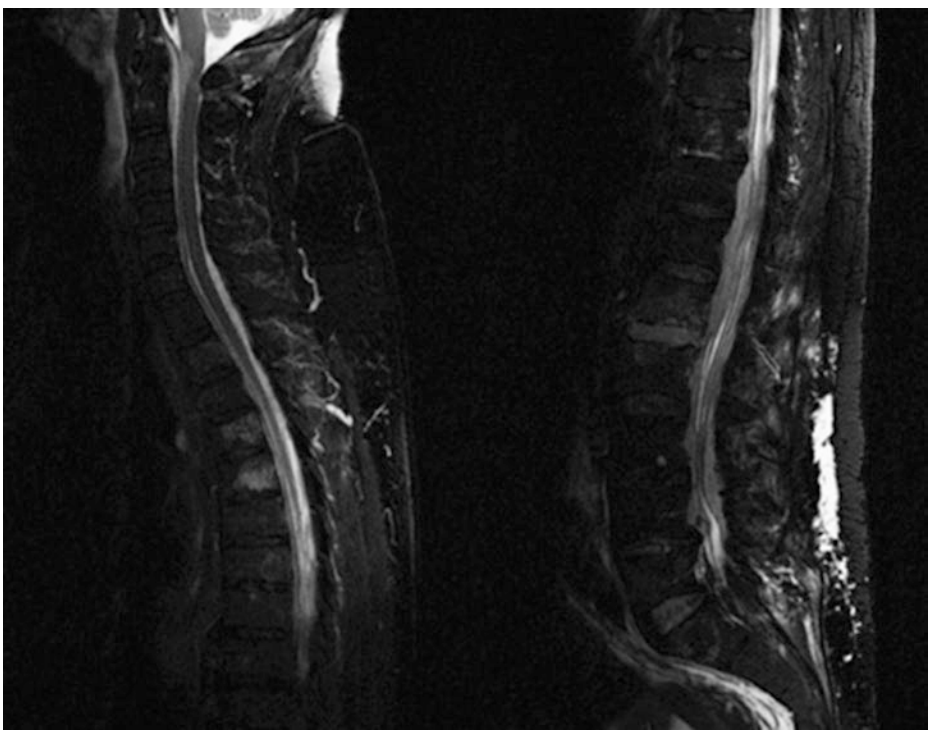

Fig. 1 Magnetic resonance imaging (MRI) showing an epidural abscess extending from C1 to T8 and from T12 to S1

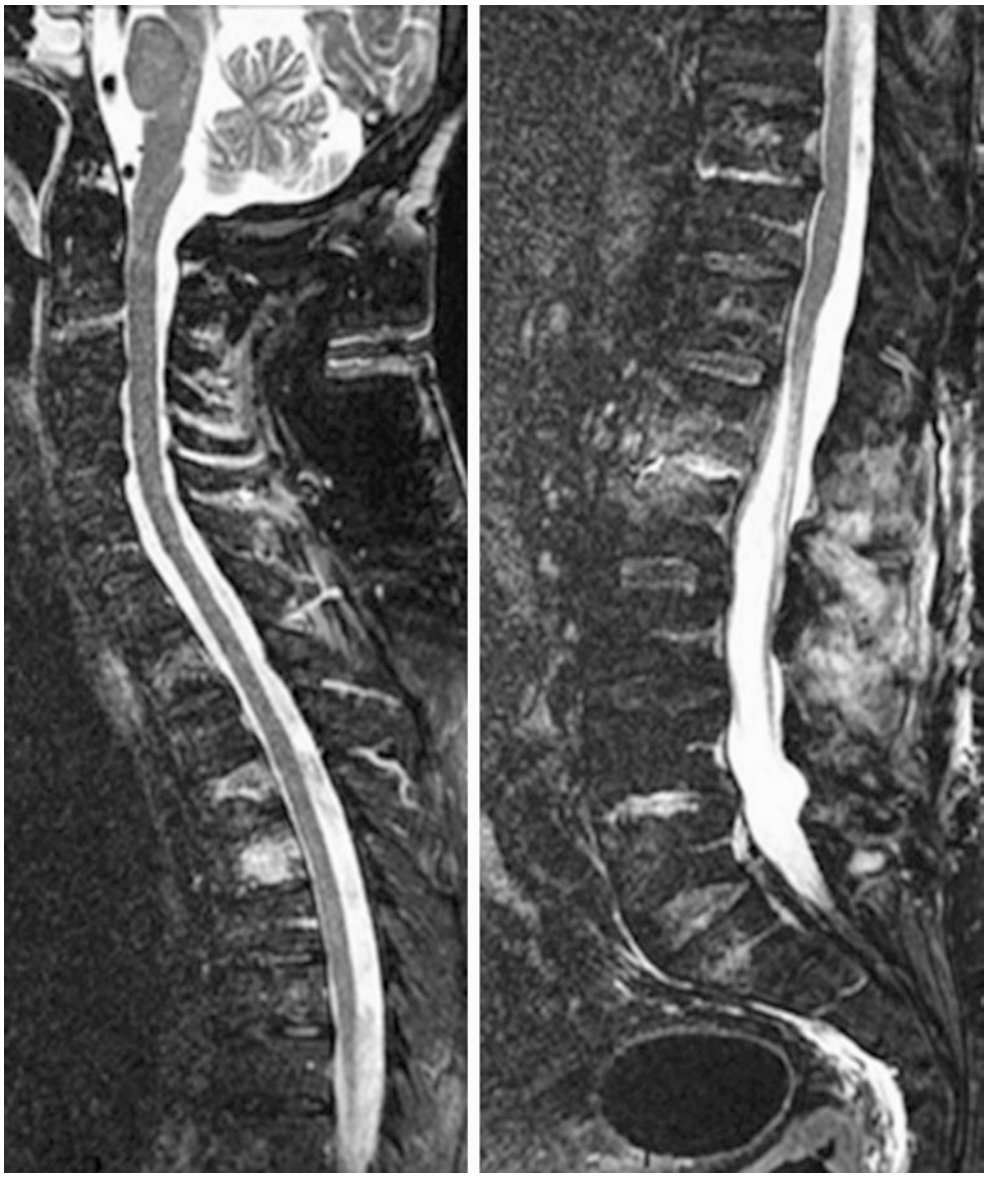

Fig. 2 Unilateral laminotomies to preserve midline structures and exposition of the epidural space

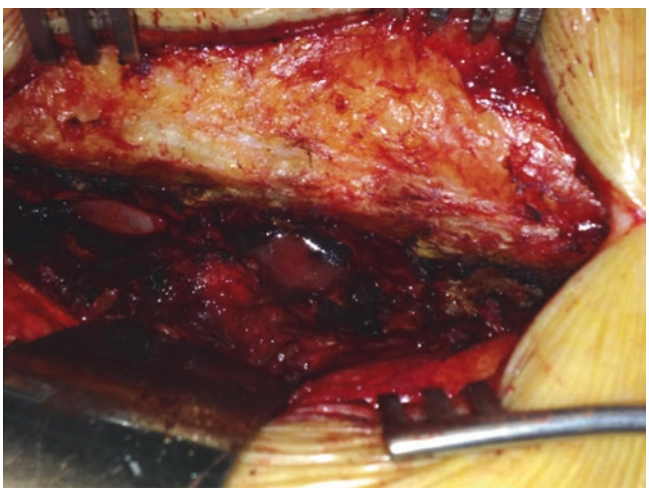

Fig. 3 Pus drainage, under mild continuous suction, using two 2.7-mm-diameter silicone catheters, inserted caudally and cranially into the epidural space at the laminotomy sites

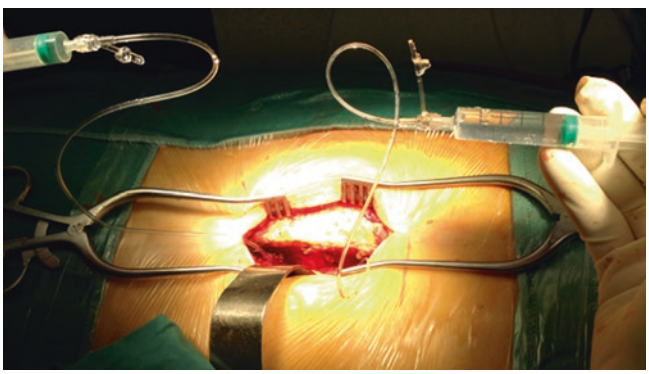

Fig. 4 Postoperative magnetic resonance imaging (MRI) showing evacuation of the spinal epidural abscess and satisfactory decompression of the neurological structures

Case 2

A 72-year-old female complained of increasing pain in the thoracolumbar spine, which was not responsive to anti-inflammatory therapy. She did not present with any neurological disorders at the four extremities or the sphincter, or any sensory deficit. She had severe obesity (body mass index (BMI) 44 kg/m^2) and a history of lupus erythematosus (LES), which had been treated with steroids for 20 years, but this treatment had been suspended 5 years ago. She was admitted to our emergency department with low back pain, fever, hypotension and oliguria. A complete blood count, CRP level and procalcitonin level were obtained, showing a white blood cell count of 18.57 × 10^9/L with neutrophilia, a CRP level of 204 mg/dL (range 0–3 mg/dL), and a procalcitonin level of 24 (range <0.05). She was admitted to the department of internal medicine with a diagnosis of septic shock. In this department, more tests were required. Plain radiographs of the cervical, thoracic, and lumbar spine showed no findings. A total-body computed tomography (CT) scan showed multiple renal abscesses and a swollen appearance of the piriformis muscle. She clinically deteriorated over the next 5 days after admission, showing a reduction in her conscious state (with a Glasgow Coma Scale (GCS) score of 11) and a fever of 39 °C. A whole-spine MRI (with gadolinium) was performed, showing an epidural abscess extending from T1 to L4, located posterior to the cord and causing a compromise in the canal diameter and cord compression. T2W sagittal sections showed L1/L2 discitis and enhancement of the paraspinal muscles, particularly to the right, where there were three abscess cavities at the level of L1. The patient underwent emergency surgical spinal decompression. With use of a minimally invasive technique, two levels of unilateral laminotomies on the left side at T7 and L2 were performed in an attempt to preserve the midline structures. The pus was drained, under mild continuous suction, using two 2.7-mm-diameter silicone catheters, inserted caudally and cranially into the epidural space, at the laminotomy sites. Both catheters were passed into the posterior epidural space under the legamentum flavum, upward and downward. Irrigation was performed with several litres of normal saline until clear fluid was obtained. Samples were obtained and sent for Gram staining and culture. The microbiological examination was positive for *Escherichia coli.* Targeted antibiotic therapy was administered, and the patient showed an improved neurological condition. A 3-month follow-up MRI showed a clear reduction of the abscess involving the posterior epidural space from T1 to L4. At 6-month follow-up, the patient had achieved an excellent neurological recovery, with thoracolumbar pain reduction, and radiographs showed no signs of instability that required additional surgical treatment.

Literature Review and Discussion

In the emergency department, a diagnosis of SEA is often not considered when a patient shows no neurological deficit. Knowledge of the aetiopathology and natural history of this disease may decrease the number of misdiagnosed cases. Bacteria gain access to the epidural space by three mechanisms: per continuitatem from a neighbouring infected structure (10–30%), through haematogenous dissemination (50%) or through iatrogenic inoculation (15%). In 30–40% of cases, no source can be identified [10]. A large variety of pathogens have been found as causative agents for SEA, with *S. aureus* involved in 60% of cases, Gram-negative rods in 10%, *Streptococcus* spp. in 9% and anaerobes in 2% [10].

Heusner et al. [12] described the clinical features and progression of SEA, occurring in four stages. At the first stage, the patient has back pain, fever and tenderness; at the second stage, radicular pain, nuchal rigidity/neck stiffness and reflex changes appear; at the third stage, the patient presents with sensory abnormalities, motor weakness, and bowel and bladder dysfunction; at the last stage, paralysis occurs. Despite the availability of increasingly refined methods of imaging, this staging system remains a valuable tool because it can allow diagnosis of SEA before the appearance of irreversible neurological damage [3, 12]. Neurological deficit can be caused by direct mechanical compression or indirect vascular occlusion by septic thrombophlebitis. Patel et al. [13] reported that diabetes mellitus, leucocytosis (a white blood cell count >12.5 × 10^9/L), positive blood cultures and a CRP level >115 mg/dL represent the four risk factors associated with failure of medical treatment of SEA.

In cases of a suspected acute epidural abscess, it is mandatory to perform a whole-spine MRI to exclude the possibility of multisegmentary involvement. Contrast-enhanced MRI represents the gold standard because it allows us to identify abscess extension in the sagittal, coronal and axial planes. Epidural abscesses usually show hyperintense signals on T2-weighted images with enhancement in postcontrast studies; homogeneous enhancement on T1-weighted sequences obtained with gadolinium and hyperintensity throughout the lesion on T2-weighted sequences are more consistent with phlegmon; a bright edge with a hypointense nucleus on T1-weighted images is indicative of liquid pus [14–17]. Use of MRI is essential to distinguish phlegmon from liquid pus in order to choose the better surgical approach. Lumbar puncture plays a less important role in diagnosing SEA and should not be performed routinely. The result of Gram staining of cerebrospinal fluid (CSF) is frequently negative, so we believe that CSF sampling is to be avoided if we suspect SEA, as that procedure could introduce pathogens into the thecal sac, causing fulminant meningitis.

Above the foramen magnum the dura is tenaciously adherent to the bone. Below this level there is a real epidural space, posterior and lateral to the spinal cord, that extends along the entire length of the spine. This space is poorly represented in the cervical region and largest in the thoracic and lumbosacral regions, and it is filled with fat, arteries and venous plexus. SEA is typically located in the 'true' posterior space. Anteriorly there is only a potential epidural space because the dura is adherent to the vertebral bodies from the foramen magnum down to L1. For this reason the majority of SEAs are localized posteriorly. In rare cases the epidural abscess can involve more than five vertebral levels (a so-called extensive epidural abscess) or the entire spinal canal from the cervical region to the sacrum (a so-called holocord or holospinal epidural abscess).

Only a few cases of extensive or holocord SEA have been described. We performed a review of the recent literature in the PubMed, Embase and Google Scholar databases by conducting a keyword search for the terms 'extensive spinal epidural abscess', 'holospinal epidural abscess' and 'holocord abscess'. We included only studies published between 2006 and 2016 that reported cases of spinal abscess involving five levels or more and confined in the epidural space. Cases in which there was previous spinal surgery, subdural extension of the abscess or without indication on treatment, and publications with no abstract available, were excluded. Therefore, we selected just 19 articles (including the cases described in this paper), for a total of 22 patients (Table 1). The average age was 53.6 years (range 25–77 years), and 17 male versus five female patients were affected (male to female ratio 3.4:1). Most of the patients (n = 13 (59%)) with extensive SEA had at least one of the known predisposing factors, including diabetes mellitus (27%), a history of IVDA (18%), cancer (9%), hepatitis C virus (HCV), immunodeficiency, alcoholism, obesity, chronic renal failure, Crohn's disease or cardiovascular disease. Regarding the spinal levels involved, the epidural abscess extended from the cervical region to the lumbar region in seven patients (40.9%), from the cervical region to the sacrum in eight patients (36%), from the cervical region to the thoracic region in four patients [14, 36] and from the thoracic region to the lumbar spine in one patient. The abscess was associated with *S. aureus*–positive cultures of the drained fluid or blood culture in most of the cases we reviewed (14 patients (63.6%)), *Streptococcus* spp. in two patients (9%), and *E. coli* and *Aeromonas hydrophila* in two patients; in four papers the pathogen was not specified. In all cases the clinical suspicion was confirmed by MRI of the entire spine.

Both the diagnosis and the treatment of this disease should be done as early as possible [37, 38]. The treatment of SEA is controversial, with reports of surgical and non-surgical treatment [39–43]. In Table 1 we give a brief summary of the surgical procedures performed in 20 of the overall 22 patients. Urgent surgical decompression in combination with long-term antibiotic treatment is generally considered the treatment of choice for extensive SEA. Some factors, however, such as a high surgical risk and a minor neurological deficit, favour non-surgical treatment. In the case described by Van Bergen [23], given the extent of the abscess and the numerous septations on the one hand and the minor neurological deficit on the other hand, it was decided to not perform a surgical procedure, keeping in mind that in the event of neurological deterioration, surgery would be considered. In the same way, Killen [34] reported successful non-surgical treatment of a 77-year-old patient, in whom a course of antibiotics resulted in complete radiological resolution of the abscess and a full neurological recovery. However, in this case the clinical examination did reveal a mild neurological deficit: globally reduced power (Medical Research Council (MRC) grade 4/5) in all muscle groups of the lower limbs and normal power in both upper limbs. Many surgical strategies have been described in the literature to evacuate an SEA and decompress the spinal cord, from open decompression and late closure to limited decompression (1-level laminectomy or hemilaminectomy with the use of a Fogarty embolectomy catheter) [3] and percutaneous CT-guided needle aspiration [44]. Some authors have reported performing a wide decompression through multilevel laminectomies [45], inducing postoperative instability of the spine, which could be followed by postsurgical kyphosis, important blood loss, increased postoperative pain and a prolonged recovery time [17]. CT-guided needle aspiration could cause extension of the infection to the subdural area by accidental penetration of the dura [41]. Other authors have recommended less invasive techniques involving placement of small catheters within the epidural space [3, 19, 22, 25, 32, 33, 35, 36, 41]. These side effects could be avoided by the minimally invasive technique used in our two cases. We believe that use of a minimally invasive technique in patients affected by extensive SEA is both effective and safe. It is possible through multiple laminotomies and fenestrations introducing a flexible catheter that is able to drain the epidural abscess while preserving the stability of the posterior elements of the spine. It is essential to perform unilateral decompression in order to not compromise the posterior ligamentous complex. This procedure is associated with minimum blood loss, faster functional recovery, reduction of postoperative pain and no need for subsequent stabilization surgery. This less invasive technique can treat the infection, and it can also avoid a long-term deformity. In the literature, cases of neurological deficits are not reported due to the positioning of such catheters into the epidural space. In the first case we have described, the abscess was unusually localized in the anterior cervical epidural space, making the choice of surgical approach more complex; we performed an anterior approach to the C2–C3 disc space and drainage with a 1.3-mm smooth catheter,

Table 1 Review of the recent literature

Study	Sex	Age (years)	Risk factors	Level involved	Pathogen	Conservative treatment	Surgical treatment	Duration of follow-up	Neurological sequelae
Payer and Walser [18]	Male	70	Gastric cancer	C2–T8	MSSA	IV flucloxacillin for 5 weeks	Multilevel unilateral laminar fenestrations from C2 to T8; irrigation and drainage of the abscess	2 months	None
Urrutia and Rojas [19]	Male	36	None	C2–sacrum	*Peptostreptococcus*	IV cefotaxime and clindamycin	Limited laminectomies in the cervical (C3–C5), thoracic (T5–T6) and lumbar spine (L4–L5); irrigation with a paediatric urinary catheter	5 years	None
Chang et al [20]	Male	45	None	C3–lumbar tract (C4–C6 osteomyelitis)	MSSA	Hyperbaric oxygenation and 6 weeks of IV oxacillin	Anterior cervical corpectomies; internal spinal fusion with a mesh cage with morselized autogenous bone grafts and a titanium plate	3 months	Yes
Gorchynski et al. [21]	Male	33	None	C1–L5	MRSA	IV vancomycin and oral rifampicin	Laminectomy of the entire spine; drainage (no other information available)	1 year	Yes
Smith and Kavar [22]	Male	25	Crohn's disease	C2–S1	NK	IV antibiotics	C5, T8 and L3 laminectomies; saline washout at each laminectomy site, using a ventricular catheter	NK	Yes
Van Bergen et al. [23]	Male	50	None	C2–L3	MSSA	IV flucloxacillin for 5 weeks	None	2 months	None
Elsamaloty et al. [24]	Male	53	Diabetes mellitus	C1–L1	MSSA	IV methicillin	Multilevel laminectomies performed at C5–C6, T2–T3 and L1–L2; irrigation	6 months	Yes
Tahir et al. [25]	Female	38	IVDA, HCV	C1–sacrum	MSSA	IV vancomycin and ciprofloxacin for 4 weeks	T4–T5 and L3–L5 laminotomies and flavectomy; soft silicone catheter advanced for up to 3–5 cm gently in both upward and downward directions to flush the epidural space with saline	1 year	NK
Deshmukh [26]	Female	59	Alcohol abuse, IVDA, progressive breast cancer	C2–T3 (C5–C6 spondylodiscitis)	MSSA	Ceftriaxone (2 g twice daily) for 8 weeks and a cervical collar for 3 months	Anterior contiguous 5/7-mm midline trough corpectomies in the vertebrae and intervening disc spaces from C2 to T3; drainage	1 year	Yes
Lehman and Lenke [27]	Female	38	IVDA	C7–L4	NK	NK	Thoracic laminoplasty, hinged open; multiple laminotomies in the lumbar spine; irrigation	NK	NK
O'Brien et al. [28]	Male	71	Diabetes mellitus	CCJ–sacrum	MSSA	IV piperacillin/ tazobactam	None	2 months	None
Lau et al. [29]	Male	50	Diabetes mellitus	CCJ–L5 (L3–L4 osteomyelitis)	MSSA	IV broad-spectrum antibiotics	Decompression through a transoral odontoidectomy; removal of the anterior arch of C1; partial corpectomy of C2; laminectomies of C3–C6 and C1–C2; posterior cervical fusion	13 months	Yes

Lee et al. [30]	Male	72	Diabetes mellitus (T7 vertebroplasty 5 weeks later)	C4–T8 (T6–T8 osteomyelitis)	*Aeromonas hydrophila*	IV ampicillin/ sulbactam	Multilevel laminectomies from C5 to T8; posterior instrumentation from T3 to T10 (to stabilize pregress T7 vertebral fracture)	2 years	None
Velpula et al. [31]	Male	67	None	CCJ–T4 (C6/C7 discitis)	NK	IV antibiotics	Anterior cervical decompression	NK	Yes
Shiu et al. [32]	Male	69	Diabetes mellitus, cardiovascular disease	Whole spinal canal (L3–L4 osteomyelitis)	*Streptococcus intermedius*	IV antibiotics	Vertebroplasty of L4; laminectomy of T6 and T8; subcutaneous drainage tube	2 months	Yes
	Male	46	None	C1–L5	MSSA	IV broad-spectrum antibiotics	Segmental laminectomies of C3–C6, T6–T9 and L2–L3 for abscess drainage; a red rubber catheter was used to irrigate the epidural spaces	1 month	Yes
Smith et al. [33]	Male	51	None	C1–S1	MRSA	Linezolid and rifampicin for 3 months	Laminectomy of L3; a catheter was used to wash out the epidural space rostral and caudal to the laminectomy; C1–C2 laminectomy 6 days postoperatively to drain residual ventral collection at the CCJ	11 months	Yes
Killen et al. [34]	Female	77	None	C2–L1	MSSA	IV flucloxacillin and oral fusidic acid for 4 weeks, then oral flucloxacillin for a further 5 weeks	None	1 year	None
Abd-El-Barr et al. [35]	Male	51	Diabetes mellitus	C2–S2	MSSA	IV daptomycin	Apical skip laminectomy (2/3 levels, centred at the apexes of the natural spinal lordosis and kyphosis in the cervical, thoracic and lumbar spine); epidural irrigation (saline + vancomycin) using a small-bore paediatric feeding tube	1 year	None
	Male	46	IVDA	C1–L4	NK	NK	Apical skip laminectomy and irrigation (as in the above case)	1 year	None
Proietti (2016)	Male	62	None	CCJ–S1 (L1–L2 discitis)	MSSA	IV antibiotics	Anterior approach to the C2–C3 disc space; drainage with a 1.3-mm smooth catheter; posterior laminotomies on the left side at T11/T12, L/2 and L3/L4; irrigation using a 2.7-mm-diameter silicone catheter inserted caudally and cranially	2 years	None
	Female	72	Obesity, LES	T1–L4 (L1–L2 discitis)	*Escherichia coli*	IV antibiotics	Posterior unilateral laminotomies at T7 and L2; irrigation with two 2.7-mm silicone catheters inserted caudally and cranially into the epidural space	6 months	None

CCJ craniocervical junction, *HCV* hepatitis C virus, *IV* intravenous, *IVDA* intravenous drug abuse, *LES* lupus erythematosus, *MRSA* methicillin-resistant *Staphylococcus aureus*, *MSSA* methicillin-sensitive *Staphylococcus aureus, NK* not known

followed by irrigation through posterior skip laminotomies in the thoracic and lumbar regions. Most of the authors in our review reported good results obtained by emergency surgical treatment in patients affected by extensive SEA, in combination with intravenous antibiotic therapy. However, neurological sequelae persisted in most cases, especially when the pretreatment deficit was severe. Delays in diagnosis and treatment of extensive SEA lead to poor results; moreover, if surgical intervention occurs after a long course of failed medical management, the ability to recover motor function is significantly limited [13].

Conclusion

An extensive spinal epidural abscess is a rare but life-threatening condition, which requires early diagnosis and prompt management. The main pathogenic mechanism seems to be bacterial haematogenous dissemination, in most cases involving *S. aureus*. Magnetic resonance imaging is crucial to confirm the clinical suspicion and to show the real extension of the abscess. The recent literature mostly recommends surgical decompression followed by intravenous antibiotics in patients with neurological abnormalities. Patients with a high surgical risk and a minor neurological deficit should receive non-surgical treatment. The minimally invasive technique used in our cases is very useful to treat an extensive epidural abscess of the spine. Thanks to this approach, spinal surgeons can treat most extensive spine epidural abscesses, decompressing the spinal cord and isolating the pathogen, with fewer side effects.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

The patients discussed in this paper have expressed their consent for the presentation of their cases and the processing of their personal data.

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Complications of Halo Placement

Giuseppe Talamonti, Alberto Debernardi, Massimiliano Visocchi, Fabio Villa, and Giuseppe D'Aliberti

Abstract *Background:* The halo vest is widely used throughout the world to manage craniovertebral and cervical instabilities. It can be used for postoperative immobilization or as an alternative to surgical fixation.

Method: In this paper we present some cases of severe complications from our own practice and review the literature on halo complications.

Results: Like any therapeutic manoeuvre, halo placement may be followed by various complications. In the meantime, modern techniques of fixation offer safe and immediate stabilization.

Conclusion: The halo vest remains a formidable method for cervical immobilization. However, it should not be used a priori instead of surgery.

Keywords Cervical fixation · Cervical immobilization · Complications · Cranio-vertebral joint · Halo vest

In the late 1950s, halo vests became available for the treatment of cervical instability. These devices rapidly gained wide acceptance for the management of a number of cervical diseases. Conservative (nonsurgical) treatment became possible in several types of cervical fractures [1]; adequate and effective postoperative immobilization allowed improved surgical results and permitted development of new surgical strategies for management of injuries, malformations, tumours, infections and inflammation. However, as the use of the halo vest has proliferated worldwide, knowledge has also increased regarding its limitations, failures and possible complications.

In the meantime, the enormous development of surgical techniques and instrumentation have radically changed cervical spine surgery.

In this paper, we present some cases of halo complications that have led us to reconsider the spread of halo indications.

Case Presentation

Case No. 1

This 7-year-old boy presented with a 3-month history of streptococcal pharyngitis and progressive development of cervical pain and torticollis. Neuroradiological assessments (Fig. 1a, b) documented Grisel's syndrome [2] (nontraumatic atlantoaxial subluxation), which was identified as grade 3, according to the Fielding classification [3]. Accordingly, treatment consisted of antibiotics and halo vest placement. A couple of weeks later, the patient was again referred to us because of local skin swelling and intense pain triggered by light finger pressure. A computed tomography (CT) scan revealed a depressed skull fracture without any intracranial haemorrhage (Fig. 1c). The relatives denied any traumatic event. The halo vest was removed, the fracture was repaired and C1–C2 fusion was performed during a single procedure. The results were excellent (Fig. 1d, e).

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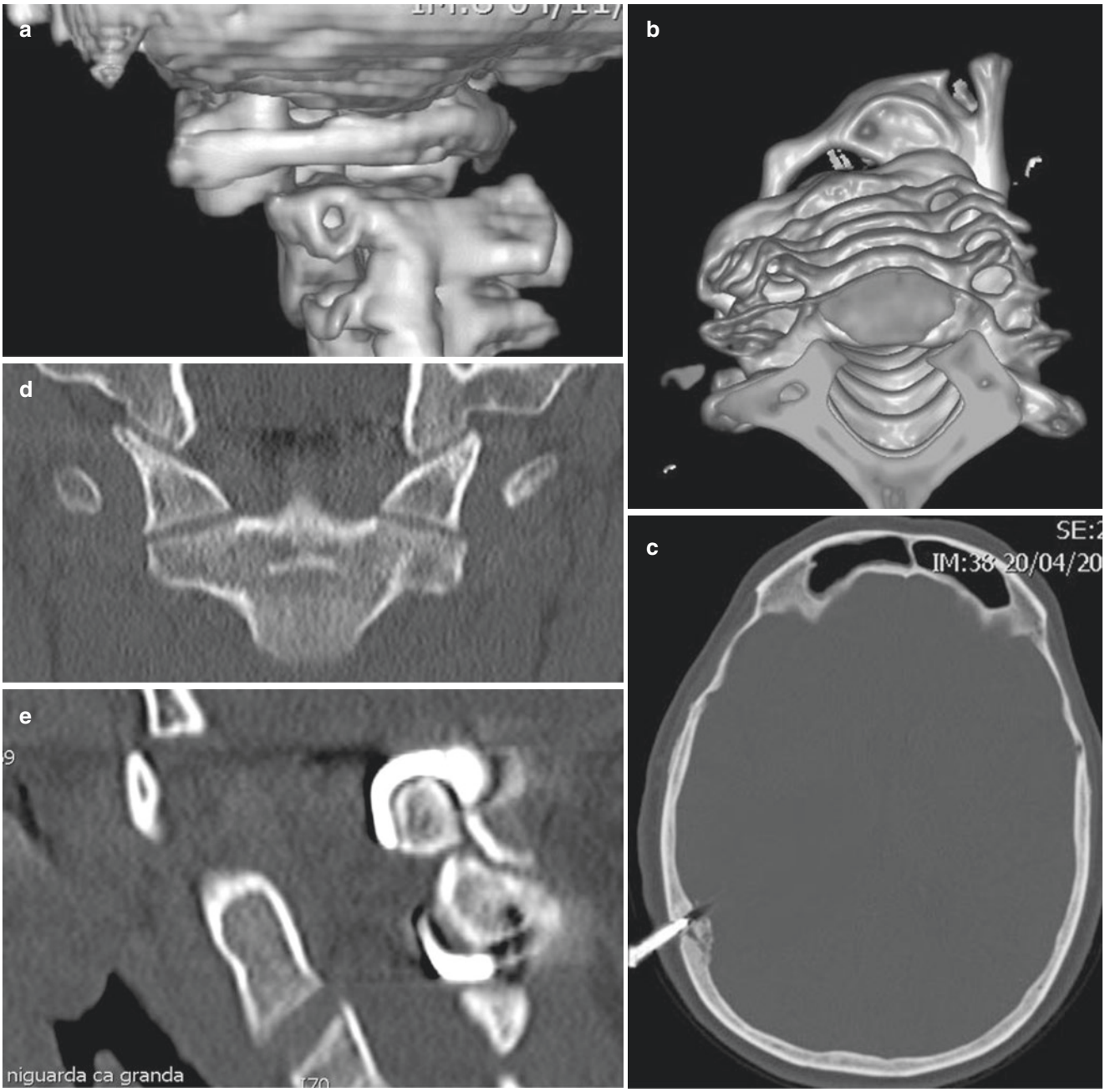

Fig. 1 Initial cervical computed tomography (CT) scans (three-dimensional reconstructions): (**a**) lateral view and (**b**) inferior view showing rotational luxation of the atlas (grade 3 Grisel's syndrome). Cranial CT scan: (**c**) axial view showing halo pin penetration with a corresponding depressed skull fracture. Follow-up cervical CT scans: (**d**) coronal view and (**e**) sagittal view showing restoration of normal atlantoaxial relationships and laminar fusion achieved using an autologous bone graft and sublaminar hooks

Case No. 2

This 11-year-old girl suffered with Costello's syndrome. Therefore, she underwent transoral odontoid resection and occipitocervical screw fixation at another hospital. Unfortunately, an infection of the posterior wound required partial removal of the occipitocervical instrumentation and placement of a halo device. Two months later, two halo pins loosened and the patient was referred to our emergency room. The skin was lacerated by the right posterior pin and a small skull lacuna was evident (Fig. 2a). The halo device was removed and the wound was cleaned and repaired; no local infection was evident. Afterward, the craniocervical fixation was renewed. Three months later, the patient came back to our emergency room because of a headache and opisthotonus.

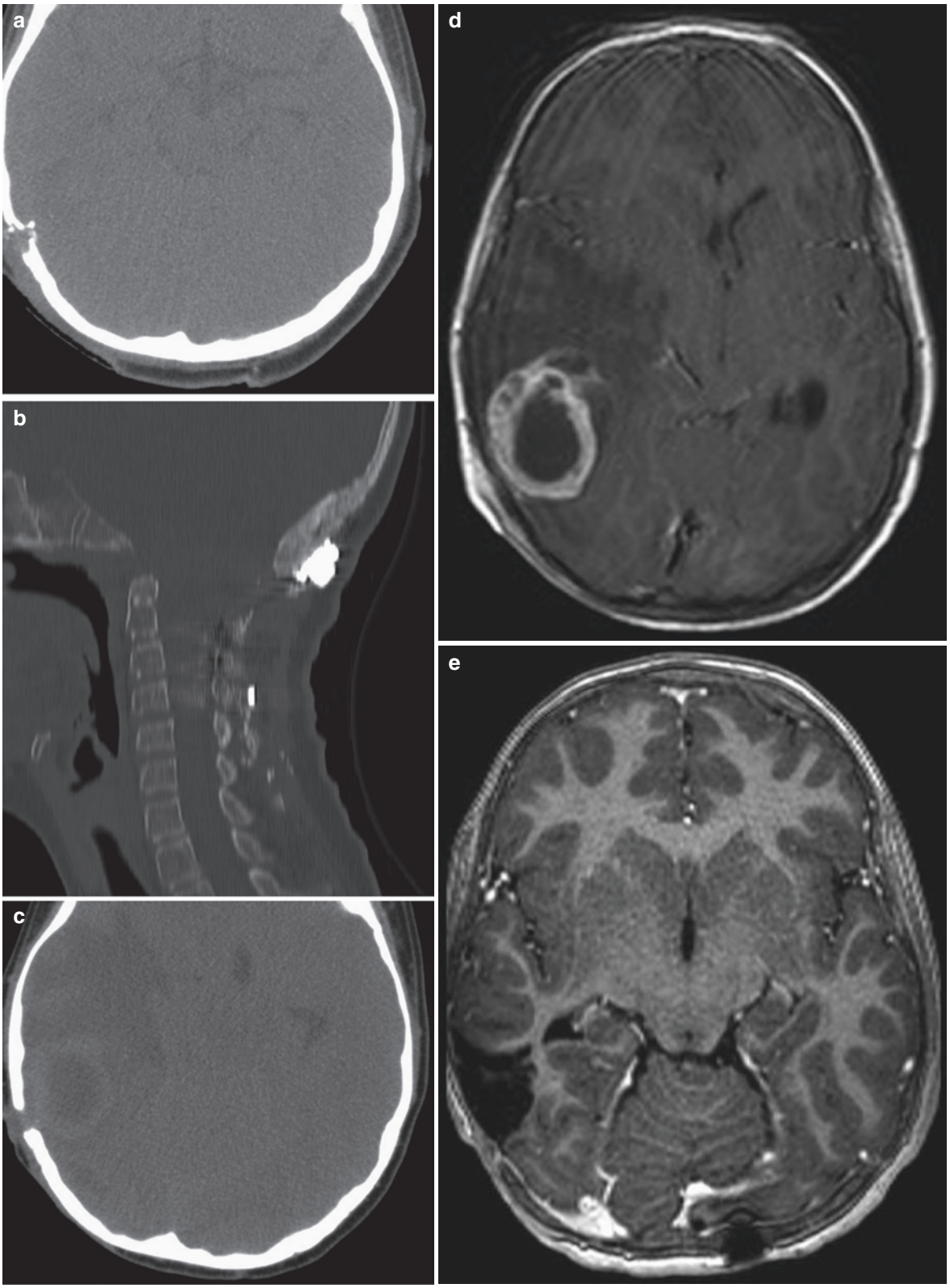

Fig. 2 Cranial computed tomography (CT) scan: (**a**) axial view obtained when two halo pins loosened; a skull defect was evident but there were no sign of infection. Cervical CT scans: (**b**) sagittal view and (**c**) axial view obtained when the patient came back because of a headache and neck stiffness; no cervical misalignment or instrument misplacement was evident, but the upper slices (bone window at cranial level) showed a cystic lesion corresponding to the skull defect. Enhanced cerebral magnetic resonance image (MRI): (**d**) axial view confirming an intracerebral abscess at the right parieto-occipital level. Follow-up enhanced cerebral MRI: (**e**) axial view showing favourable postoperative evolution

There was no fever, and the wounds appeared completely healed. A craniocervical CT scan was immediately obtained. The fixation devices were normally placed, but a cerebral cystic lesion was incidentally partially seen at the top of the scan (Fig. 2b, c). Enhanced CT scanning and magnetic resonance imaging (MRI) of the brain confirmed the presence of a large cerebral abscess under the small skull lacuna, corresponding to the position of the previously loosened pin (Fig. 2d). The girl underwent immediate abscess excision, while the fixation device was left untouched. The results were excellent (Fig. 2e).

Other Cases

Reviewing our series, we found other two old cases: in 1989 an extradural haematoma had to be removed after halo pin placement (Fig. 3), and in 1996 a 12-year-old girl with a craniocervical injury developed an unrecognized chronic local infection around a halo pin, which eventually led to cranial osteomyelitis (Fig. 4a, b) requiring a wide craniectomy and subsequent cranioplasty. Finally, recently, we had to remove a halo vest because it triggered panic attacks in a young doctor with cervical fracture, who preferred to be operated on rather than continue wearing the device.

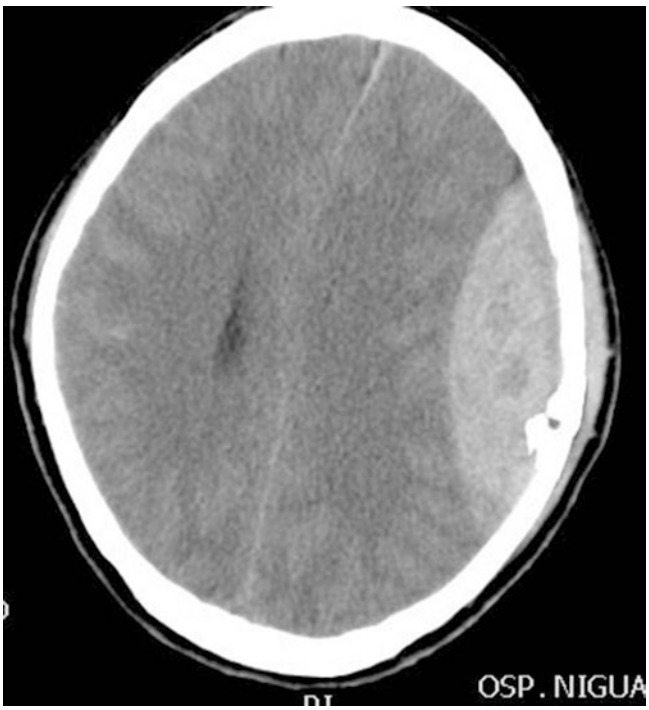

Fig. 3 Cranial computed tomography (CT) scan showing a relatively large extradural haematoma originating from a depressed skull fracture due to halo pin penetration

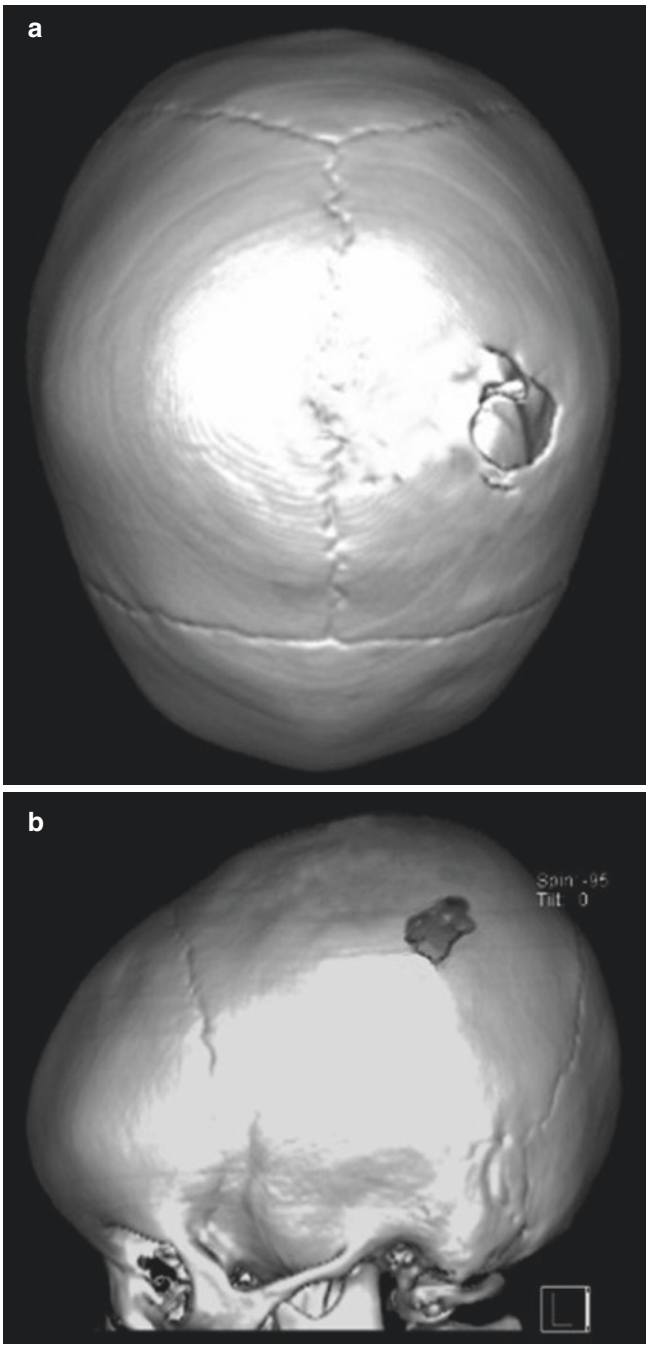

Fig. 4 Cranial computed tomography (CT) scans (three-dimensional reconstructions): (**a**) bird's-eye view and (**b**) lateral view showing a skull defect due to osteomyelitis originating at the site of halo pin insertion. This condition was not recognized and not adequately treated, leading to enlargement of the bone lesion and eventual need for a wide craniectomy and subsequent cranioplasty

Discussion

The halo vest consists of a titanium ring, which is firmly screwed to the skull by means of four pins. Four rods keep the ring attached to a special vest, thus immobilizing the

cervical spine. This device has several advantages: it allows transskeletal traction and may be placed to reduce cervical misalignments [1]; the cervical immobilization is virtually absolute; the mandible is free; the whole neck remains accessible in both its anterior and posterior aspects; the patient can ambulate freely; and the halo can be maintained for months. Accordingly, a common policy consists of halo vest placement to firmly maintain the cervical spine in the correct position for a given period of months; afterward, the healing of the instability is assessed and the halo is either removed or maintained for some further months. Eventually, if cervical stabilization has not been yet achieved, the only remaining option is surgical arthrodesis.

Of course, some limitations exist in the use of halo device. For instance, halo ring placement requires an adequately developed skull, which makes it unsuitable for small infants [4]. Some pinless devices have been developed recently [5], but they do not seem to achieve the same degree of immobilization as the traditional halo system. Moreover, prolonged immobilization (by whatever means) is generally poorly tolerated by children, in whom more aggressive surgical indications may therefore be considered from the outset [6]. Psychiatric patients and even very anxious subjects may not be ideal candidates for a halo vest. Finally, in elderly patients, use of a halo vest results in a low rate of effective and permanent stabilization; thus, direct surgical arthrodesis may be indicated.

Apart from the aforementioned limitations, as with any surgical procedure, several problems may follow halo placement. Poorly outlined side effects are the discomfort of wearing the halo vest for months and the possible psychological problems it may elicit.

However, the most widely reported complications are pin loosening, pin site infection, pressure sores, pin site discomfort, loss of reduction, pneumocephalus, epidural abscess, brain abscess, subdural haematoma and seizures [1, 5, 7–12]. Even a fracture of a ventriculoperitoneal shunt and orbital violation by a pin have been reported [13, 14]. All of these complications mainly result from inadequate techniques of halo placement; for instance, a pin that is placed too deeply can be responsible for intracranial troubles, but pin that is not placed deeply enough may lead to system loosening, misplacement, skin injury and so on. Moreover, infective complications may result from poor pin care, especially in outpatients. Accordingly, adequate education of both physicians and patients is mandatory [9].

In 1986, Glaser et al. [8] reported pin complications in about 7% of cases. In the same year, Garfin et al. [7] published estimates of a 36% incidence of pin loosening, a 20% incidence of pin site infection, an 18–19% incidence of wound problems, a 2% incidence of nerve injuries and a 1% incidence of dural violation. Indeed, halo complications are mainly reported as anecdotal case reports [1, 8–14], and their actual rate of occurrence is quite difficult to assess precisely [7]. Undoubtedly the most frequent problem is pin loosening. Following halo placement, the pins usually need to be retightened. This is quite normal and is related to minimal, nearly physiological bone lysis at the pin tip. However, late loosening is typical of local infection, and this should be stressed to patients and their relatives to avoid management delays [9]. General diseases, osteoporosis, radiation therapy, the presence of foreign material and skull lesions are obvious risk factors for pin placement [9, 10, 12]. Accordingly, preoperative recognition of poor bone density is of tantamount importance to prevent complications [10].

However, cranial penetration of halo pins remains a rare event [1, 9]. It usually results from an incorrect placement technique and/or excessive pin tightening. The degree of torque application and the timing of pin retightening have been calculated to minimize the risk of skull penetration [15, 16]. Other possible causes of pin misplacement are falls, head injuries, prolonged halo use and poor patient compliance [1].

Depressed fractures of the skull are easy to understand but are uncommonly reported [7, 8, 15, 16]. They mainly depend on unexpected bone weakness and/or excessive strength or incongruous depth of pin application. We always use a dynamometric screwdriver to place the halo pins. In our case no. 1, the pins were inserted with a torque of about 4 inches per pound, as is usual in small children in our practice. Probably, the bone strength was less than expected, but we detected no anomalous sensation and remained unaware of the underlying bone complication. The procedure was conducted under local anaesthesia and mild sedation, and the patient presented no particular complaints. We did not notice any anomalous motility of the halo ring, which appeared well held to the skull. When the patient returned because of local pain and swelling at the site of one pin, the device still seemed well positioned. Indeed, we suspected a local infection, and the depressed skull fracture was a surprise. Maybe the pin could have been replaced in another position, but we preferred to repair the skull fracture and to fuse C1–C2 during the same procedure. The postoperative immobilization consisted of 2 weeks' use of a soft cervical collar. There were no complications, and the results were excellent. Now, we wonder whether the direct C1–C2 fusion might have been indicated from the outset.

Intracranial abscess is one of the most serious complications of pin penetration. An abscess may be extradural, subdural or parenchymal [1, 7, 12]. In our case no. 2, the halo had to be removed a couple of months after placement, because of pin loosening and skin laceration. A local infection was suspected, but no infective evidence was found. When the patient came back to our emergency room, her only complaints were headache and nuchal stiffness. Recurrence of the craniocervical instability was the main

suspect, but neuroradiological assessments revealed a large cerebral abscess corresponding to a small skull lacuna where the previously loosened pin was placed. The opisthotonus probably was due to the downward displacement of the cerebellar tonsils, and more severe brainstem signs were absent owing to the enlarged foramen magnum. Anyway, the patient eventually underwent successful abscess excision, but we seriously risked missing the correct diagnosis because initially we did not perform a cerebral CT scan and the abscess was only incidentally included in the upper slices of the craniocervical CT scan, which were performed just to build craniocervical CT reconstructions. The lesson learned from this case was that late pin loosening must always be regarded with a high degree of suspicion, and probably an infective disease cannot be ruled out by simple local inspection and a simple CT scan. In such cases, adequate serial clinical and neuroradiological assessments should be planned to prevent more serious complications.

Conclusion

There are patients in whom halo placement is the only therapeutic option. For instance, we recently reported [4] an 8-month-old infant who required occipitocervical arthrodesis using autologous ribs. The skull immaturity mandated the use of a customized brace for postoperative immobilization, which led to progressive graft reabsorption. Five months later, when the skull maturation allowed halo placement, this infant could be effectively treated because the adequate immobilization enabled bone strut engrafting.

There are patients in whom surgery represents the best option; for instance, patients with severe vertebral disruption and elderly patients generally have poor chances of healing without surgical repair.

There are other patients in whom both halo placement and direct surgery are valuable options.

Both options have their pros and cons. Even rare, possible complications of halo placement exist, while modern surgery is no longer as hazardous as in the past. The halo system may allow healing without open surgery, but its potential risks, the discomfort of wearing the device for months and the possibility of needing eventual open surgery because of failed healing must be pondered. On the other hand, the discomfort and the risks of direct surgery are well known and surely not negligible, but modern techniques and the present sophisticated surgical devices usually permit safe procedures and immediate solution of the cervical instability.

In the last three decades, we used a halo device in dozens of patients and had to manage the four cases of serious complications cited in the present paper. In the last 10 years, we performed about 100 craniocervical arthrodesis procedures and surely we had to face some postoperative complications and failures, but satisfactory results were soon achieved in the vast majority of cases. Accordingly, the lesser invasiveness of the halo system seemed to be counterbalanced by the immediate and low-risk effectiveness of surgery.

We do not deny the importance and usefulness of the halo system, but we think that the option of surgery should be not rejected a priori. The choice of the most suitable therapeutic option should be tailored for each individual case and discussed in depth with the patient and his or her relatives.

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CVJ Neurophysiology

Occipital Nerve Stimulation for Refractory Pain after Occipitocervical Fusion

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Abstract Occipital nerve stimulation (ONS) is electric stimulation of the distal branches of the greater occipital nerve by cylindrical or paddle leads implanted in subcutaneous occipital tissue. This surgical option has emerged as a promising treatment for different types of disabling medical refractory headache and recently also for residual occipital and nuchal pain after previous occipitocervical fusion. The mechanisms of action have not yet been clearly explained: electrical stimulation of the occipital nerve has both peripheral and central effects on the nervous system, which may modulate nociception. ONS is a well-tolerated and safe procedure in comparison with other invasive modalities of treatment. Lead migration/dislodgement is a common complication, but use of new surgical techniques and leads may reduce the rate of this complication.

Introduction

Occipital nerve stimulation (ONS) is used for various medically intractable cranial and neck pain syndromes such as trigeminal autonomic cephalalgia (TAC), migraine, occipital neuralgia, hemicrania continua, posttraumatic headache and transformed migraine [1–5]. More recently, good outcomes have been reported with ONS for occipital and cervical pain after occipitocervical fusion with subcutaneous placement of leads intersecting with the greater occipital nerve at the level of C1 [6]. This pain is characterized by neuropathic features involving the territory of the greater occipital nerve in the upper part of the neck and the back of the head. The pain is often refractory to conventional medical management and may impair the individual's lifestyle, quality of life and ability to work, resulting in a significant economic burden [6, 7]. The major causes of refractory occipital pain after occipitocervical fusion may be lesions of the occipital nerves, secondary to the surgical procedure; entrapment and infection of occipital nerves at the surgical site; and postfusion arthritis of the C1–C2 segment.

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Surgical Procedure

The surgical procedure is structured into two phases: lead placement with subchronic stimulation; and fitting of an implantable pulse generator (IPG) with chronic stimulation.

Lead Placement with Subchronic Stimulation

Under local anaesthesia, and with the patient placed in a prone position, a linear incision of 4 cm is made on the midline from the external occipital protuberance to the posterior cervical region. Blunt dissection of the subcutaneous tissue is performed bilaterally. Under fluoroscopy control, a Tuohy needle is passed transversely in the epifascial plane across the base of the greater occipital nerve at the level of C1, starting from the midline incision, moving toward the mastoid process and following the trajectory of the nuchal line. A quadripolar/octopolar cylindrical lead is introduced into the Tuohy needle, medial to the lateral direction. Then the needle is pulled back and the lead is anchored to the underlying fascia in the midline and connected to an external cable for intraoperative acute testing with an external stimulator. The same procedure is performed on the other side. The stimulation parameters are chosen to obtain paraesthesia in the innervation territory of the greater occipital nerve. The procedure has lead migration rates of up to 24%. To avoid this

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complication, according to the Franzini lead fixation technique [8], the cylindrical lead may be fixed to the lateral portion of the superficial fascia by transfixation of the plastic tip of the lead with a suture needle through a vertical incision of 4 cm, lateral to the external occipital protuberance. Patients are evaluated after a 7- to 15-day trial period of external stimulation, and only those reporting at least a 50% decrease in pain intensity, associated with a decrease in medication use, are selected for permanent implantation of an IPG.

In cases of cylindrical lead migration during subchronic stimulation, it is possible to replace it with a narrow paddle electrode [9].

The Implantable Pulse Generator and Chronic Stimulation

In patients with a positive response to the subchronic stimulation phase, under general anaesthesia a wire cable is tunnelled from the occipital lead to a subcutaneous pocket in the subclavian region, where an IPG is implanted. It is possible to connect both wire cables to a dual-channel IPG or each wire cable to a single-channel IPG. The usual parameters used for chronic stimulation are a frequency of 30–60 Hz, pulse width of 90–120 μs, amplitude of 1.5–9.0 V and bipolar stimulation configuration in the cyclic mode.

Discussion

Occipital nerve stimulation is used with good clinical results in various types of headache involving both the cervical and trigeminal innervation territories, such as occipital neuralgia, cervicogenic headache and TAC [3, 4, 10]. Patients with occipital pain due to degenerative diseases of the cervical spine or pain after surgery on the cervical spine, including ventral discectomy and fusion, have also been reported to benefit from this procedure [6]. Electrical stimulation of the greater occipital nerve, performed through lead implantation by a percutaneous approach, was first described by Weiner and Reed in occipital neuralgia [11], but it reduces pain in both the cervical and trigeminal innervation territories [4].

The greater occipital nerve (GON) arises from fibres (the medial branch) of the dorsal primary ramus of the C2 spinal root and, to a lesser extent, from fibres of the C3 spinal root. The nerve crosses deep to the semispinalis capitis muscle and pierces the fascia just below the superior nuchal line between the semispinalis capitis and trapezius muscles. The GON supplies sensation to the skin in the medial portion of the posterior scalp up to the vertex. Its dorsal root ganglion also innervates the atlantoaxial C1–C2 joints and zygapophyseal C2–C3 joints. The convergence of the C2 dorsal horn with the trigeminal nerve in the 'trigeminal–cervical nucleus' extending from the trigeminal nucleus caudalis to C3 level [10] may explain the effect of ONS on pain located in different territories.

The pathophysiological mechanisms underlying ONS effects on pain have not yet been clearly explained. There are many—and often conflicting—theories, since electrical stimulation of the occipital nerve may modulate nociception by both peripheral and central effects on the nervous system:

(1) Stimulation could lead to depolarization blockade of the small-diameter afferent Aδ and C fibres [12]. Stimulation of the large-diameter afferent Aβ fibres produces an antinociceptive effect due to activity suppression in the C fibres and Aδ fibres of inhibitory interneurons at the level of gate control in the spinal dorsal horn [7].
(2) ONS may stimulate the supraspinal structures involved in central nociceptive control and located in the dorsal pons, with activation of descending antinociceptive pathways, which can modulate nociceptive input at the supraspinal and spinal levels by means of the trigeminal–vascular system at the level of upper cervical pain [13, 14].
(3) ONS may influence the anterior cingulate cortex and the pulvinar nucleus of the thalamus, which are involved in the affective component of pain [12].

Lead migration/dislodgement is the most common complication, occurring in up to 24% of cases. It is due to the wide range of neck movements [2]. Lead and implant techniques have evolved over time to minimize lead displacement. Transfixation of the plastic tip of the cylindrical lead by a needle and suturing of the tip to the superficial fascia with a non-absorbable suture, as proposed by Franzini, is effective in preventing this complication [8]. Replacement of the cylindrical lead with a narrow paddle electrode may be a solution in cases of lead migration [9]. The reported infection rates have ranged from 4% to 30% with varied lengths of follow-up [15, 16].

ONS may require early replacement of the IPG because of power loss/battery failure if high voltage is needed to effectively stimulate a nerve that is not in anatomical contact with the lead. The use of new rechargeable batteries increases IPG longevity and output capability, reducing the size of the device and the need for device replacement surgery.

Conclusion

Subcutaneous occipital nerve stimulation (ONS) for residual and refractory occipital and neck pain after previous occipitocervical fusion surgery is a well-tolerated and effective procedure in comparison with other invasive modalities of treatment. Moreover, the use of drugs can be reduced, with a significant reduction in side effects. The reversibility of the procedure and the absence of side effects during chronic stimulation make this technique ethically acceptable in otherwise untreatable patients, whose quality of life is consistently improved by this neuromodulation technique.

Further development of application of ONS in this field requires continuous accumulation of clinical evidence, further studies on pain pathophysiology, and improvements in implant techniques and equipment technology.

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Intraoperative Neurophysiological Monitoring for Craniovertebral Junction Surgery

Francesco Sala and Pietro Meneghelli

Abstract Craniovertebral junction (CVJ) surgery encompasses a wide spectrum of neurosurgical procedures ranging from transoral approaches for CVJ bone anomalies to surgery for intramedullary tumours. Intraoperative neurophysiological monitoring (IONM) has been increasingly used in recent years because of its ability to prevent neurological complications during surgery. In CVJ surgery the risk of neurological injuries is related first to the positioning of the patient and then to the surgical procedure. Application of IONM during the positioning of the patient permits fast recognition of impending causes of neurological injury. During surgery, continuous IONM permits real-time assessment of the functional integrity of the spinal tracts and provides useful feedback during surgical manoeuvres. The applications of IONM are mainly related to intradural procedures, but wider application of these techniques during surgery for CVJ instability and degenerative disorders has recently been described, leading also to better understanding of the pathophysiology of spinal cord injuries. In this paper we review and discuss the principal IONM techniques used during surgery around the CVJ.

Keywords Intraoperative neurophysiological monitoring (IONM) · Motor evoked potentials (MEPs) · Somatosensory evoked potentials (SSEPs) · Craniovertebral junction (CVJ) · Surgery

Introduction

The craniovertebral junction (CVJ) is a complex transition zone between the skull base and the cervical spine, composed of the occiput, axis and atlas. The structures that must be considered in planning an operative approach to this region include the brainstem and spinal cord, the lower cranial and upper spinal nerves, the vertebral artery and its branches, and the ligaments uniting the atlas, axis and occipital bone. The pathological entities related to this segment of the spine are various: congenital malformation of the CVJ, Chiari malformations, inflammatory diseases (mainly rheumatoid arthritis), traumatic injuries of the CVJ, intramedullary tumours and vascular malformations. Therefore, the anatomical complexity of this region and the different pathologies related to the CVJ region underline the technical challenges of CVJ surgery.

The risk of developing new neurological deficits is related to two principal steps of the surgical procedure: the positioning of the patient and the surgery itself. Positioning, flexion or extension of the head, and the traction used on the shoulders may influence the diameter of the spinal canal, possibly leading to medullary compression and injury [1] or brachial plexus injury [2]. During surgery, a spinal cord injury may occur through either primary or secondary mechanisms: the former category defines injuries caused by surgical manoeuvres performed directly on the spinal cord (intramedullary tumours, above all); the latter defines injuries caused by indirect manoeuvres performed on the spine (e.g. traction and realignment manoeuvres).

Intraoperative neurophysiological monitoring (IONM) is aimed at preventing neurological complications related to the surgical procedure and has been widely adopted during surgery for cervical spine degenerative diseases [3] and for intramedullary tumours [4]. The application of IONM techniques provides continuous assessment of the functional integrity of the spinal tracts and allows us to avoid, or at least limit, neurological injuries during the positioning of the patient and during surgery.

The aim of this paper is to provide a review of current IONM techniques and their applicability to CVJ surgery.

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Intraoperative Neurophysiological Monitoring Techniques and Interpretation

IONM techniques represent a group of different intraoperative evoked potential (EP)–monitoring modalities; during CVJ surgery the techniques most commonly used are somatosensory evoked potentials (SSEPs), muscle motor evoked potentials (mMEPs) and corticospinal motor evoked potentials (D-wave monitoring).

Somatosensory evoked potential monitoring provides continuous assessment of the dorsal column pathways of the spinal cord, from the periphery to the cerebral cortex. The median nerve and the posterior tibial nerve are normally used for SSEP monitoring of the upper extremities and lower extremities, respectively. Upper-extremity monitoring starts with depolarization of the median nerve by electrical stimulation to produce a synchronous action potential volley through the sensory fibres of the dorsal root, which travels through the fasciculus cuneatus to reach the nucleus cuneatus (above T6) and then the contralateral medial lemniscus, and terminates in the ventral posterolateral (VPL) nucleus of the thalamus; third-order neurons reach the somatosensory cortex and parietal association fields and are processed by cortical scalp leads. Lower-extremity monitoring starts with depolarization of the posterior tibial nerve: the action potential travels through the fasciculus gracilis to the nucleus gracilis, then second-order neurons decussate, travel through the contralateral medial lemniscus and terminate in the VPL nucleus of the thalamus. As previously described, third-order neurons project to the somatosensory area and parietal association fields, and are processed by scalp leads. Scalp leads are placed at CP3 and CP4 with a forehead reference (Fpz or Fz). The parameters monitored are the SSEP amplitude and the latency, which refers to the transit time from the point of stimulation along the peripheral nerve to the cortex. Baseline values are recorded prior to incision, before dural opening and at the beginning of intradural manipulation. A standard protocol then has to be used for decision making in the event of a decrease in SSEP amplitude >50% and/or a latency delay >10%.

Motor evoked potential monitoring is used to assess the integrity of motor pathways. These can be assessed with two modalities. Muscle MEPs are recorded via couples of needle electrodes, 2–3 cm apart, inserted into upper and lower-extremity muscles, following activation of the corticospinal (CST) motor tract, as well as other descending motor tracts such as the propriospinal or the rubrospinal pathways, via transcranial electrical stimulation (TES). Muscle MEPs are not specific to the CST. Conversely, D-wave monitoring permits specific evaluation of the fast-conducting fibres of the CST. TES of the motor cortex is usually applied using corkscrew electrodes, which guarantee low impedance [5]. In our daily practice we use short trains of 5–7 square-wave stimuli of 0.5 ms duration and an interstimulus interval of 4 ms, delivered at a repetition rate of up to 2 Hz through corkscrew electrodes placed at the C1 and C2 scalp sites, according to the 10–20 EEG system for electrode placement. A C1–C2 montage preferentially elicits right-extremity mMEPs, whereas a C2–C1 montage favours left-extremity mMEPs. For monitoring of lower-extremity muscles, sometimes a Cz–Fz montage is preferred, producing less intense muscle twitching. The stimulation intensity usually does not exceed 200 mA. We usually monitor mMEPs from the abductor pollicis brevis and the extensor digitorum longus for the arm and from the tibialis anterior and the abductor hallucis for the leg. Muscle MEP amplitude and latency should be evaluated in comparison with baseline values and through a standard step-by-step protocol to exclude anaesthetic or technical abnormalities. Complete loss of mMEPs correlates with permanent paresis; no change in mMEPs usually predicts a good motor outcome from an early stage after surgery. However, because of their polysynaptic pathway, mMEPs are very sensitive to the effects of anaesthesia. Therefore, even when muscle relaxants are not given, wide variations in mMEP amplitude and latency can be observed, and this variability explains the lack of a linear correlation between intraoperative changes in mMEP amplitude and/or latency, and the motor outcome [6].

D-wave monitoring permits direct evaluation of the integrity of the CST along the spinal cord. A single TES stimulus is applied, using the same montage as for mMEPs, and the D-wave is recorded by an electrode placed in the epidural or subdural space of the spinal canal, caudal to the lesion. Signals are amplified 10,000 times, and the bandwidth is amplified from 1.5 Hz to 1700 Hz. Baseline D-waves are recorded after exposure of the spinal cord. According to the literature, a decrease of more than 50% of the baseline amplitude is considered significant and is related to long-lasting neurological deficits, whereas decrements of the D-wave amplitude between 30% and 50% of the baseline are considered minor warning signs [7]. The D-wave has proven to be the strongest predictor of long-term motor outcome, and its preservation above 50% of the baseline amplitude typically correlates with only a transient deficit. Therefore, with regard to intramedullary spinal cord tumour surgery, in consideration of the wide variability in mMEP amplitude, whenever the D-wave is monitorable, we use an ‘all or nothing’ criterion for mMEPs. However, this criterion should not be applied to other spinal procedures and scenarios when the D-wave is not monitorable, because motor deficits can occur in the presence of a significant amplitude drop. In general it is advisable to inform the surgeon of any ‘significant’ (50–75%) drop in mMEP amplitude [7–9].

Lower cranial nerve monitoring assesses the functional integrity of the IX, X, XI and XII cranial nerves. For recording, needle or wire electrodes are inserted into the following

muscles: the posterior wall of the pharynx (IX–X), the vocal cords or cricothyroid muscle (X), the trapezius (XI) and the tongue (XII). Monitoring of lower cranial nerves can be performed by means of free-running electromyography (EMG) with identification of different discharge patterns, related to either irritative or injury activity [10]; the triggered EMG mapping technique can also be used during surgery. However, the interpretation and value of free-running EMG for the lower cranial nerves have been rather disappointing, as both false positive and false negative results are possible [11]. Therefore, to improve the reliability of cranial nerve monitoring, the principles of mMEP monitoring for limb muscles have been extended to monitoring of muscles that are innervated by cranial motor nerves. Muscle MEPs are recorded, after TES, by wire electrodes inserted into the muscles innervated by the lower motor cranial nerves. The main advantage of this technique is that the functional integrity of the corticobulbar tracts is assessed continuously from the motor cortex down to the neuromuscular junction. A reproducible mMEP can be continuously recorded from the pharyngeal and tongue muscles while the lower brainstem is being surgically manipulated. In our department, we use TES with a train of four stimuli at a rate of 1–2 Hz and an intensity of 50–150 mA. The electrode montage is usually C3–Cz for right-side muscles and C4–Cz for left-side muscles. Additional details on this technique have been described elsewhere [12].

Anaesthesia Protocol

IONM can be deeply influenced by the nature and depth of anaesthesia. Halogenated anaesthetics elevate the synaptic threshold for activation and transmission through the motor pathways, thus affecting the reliability of mMEPs. The recommended regimen for general anaesthesia is based on the total intravenous anaesthesia, which is based on the use of propofol and opioids in continued infusion. The use of muscle relaxants should be limited to the induction of general anaesthesia because of their ability to abolish mMEPs. Conversely, D-wave monitoring is not affected by muscle relaxation and, in general, is very little affected by anaesthesia because of its monosynaptic pathway.

Discussion

CVJ surgery is highly demanding because of the eloquence of the regions involved and the bony–ligamentous complexity. The multiplicity of risky steps and the evolution of surgical techniques have rapidly increased the need for IONM to avoid iatrogenic injury during surgery. The risk of new neurological injury or worsening of previous deficits during CVJ surgery is related to two principal steps: the positioning of the patient and the surgical procedure itself.

During the positioning of the patient, various degrees of flexion or extension of the cervical spine are related to changes in the spinal canal and thus also the potential risk of neurological injury. During surgery for degenerative cervical spondylosis, changes in SSEPs [13] and mMEPs [14] have been reported in relation to the position of the cervical spine. Plata Bello et al. reported that IONM changes were more common during positioning than during surgery. The warning criteria were an amplitude reduction of more than 50% and/or a latency increase of more than 10% on SSEP monitoring. On mMEP monitoring, an amplitude reduction of more than 80% and/or a latency increase of more than 10%, uni- or bilaterally, were considered significant. IONM changes were recorded in five patients, all during positioning. There was complete mMEP loss in all five patients and in two cases it was associated with SSEP loss; restoration of baseline IONM values after repositioning was achieved in all but one patient. This latter case showed a transient tetraparesis, which disappeared within 2 weeks after surgery. The other four patients did not show any postoperative deficits [15]. This report may further explain the presence of 'false negative' results reported in other surgical series, which were probably related to the fact that at the beginning of the surgical procedure, neurological injuries were already present, having been induced by the patient positioning [16, 17]. It should also be considered that surgical access to the CVJ sometimes requires moderate traction on the shoulders to achieve adequate visualization of the region; however, traction on the shoulders has been related to SSEP changes during surgery for cervical myelopathy [1]. Labrom et al. [18] reported an ulnar nerve SSEP amplitude decrease greater than 30% in 27 patients prior to scoliosis surgery in the prone position; clinically identifiable brachial plexus palsy occurred in only one case, in which all attempts to regain electrical function by repositioning failed. Furthermore, Schwartz et al. [19] reported impending injury to the brachial plexus as the most frequent cause of SSEP and/or mMEP changes in patients undergoing anterior cervical spine surgery in the supine position: 65% of the patients with neuromonitoring changes (1.1% of a total of 3806 patients) showed position-induced injury to the brachial plexus. Impending brachial nerve injury was most commonly noted immediately following shoulder taping; neck extension for optimal surgical access was the second most frequent cause. The American Society of Anesthesiology has identified male sex as a prominent risk factor for position-related nerve injuries; other patient characteristics that have been reported to carry a greater risk of position-related injury include obesity, preexisting spinal cord disease and diabetes mellitus [20].

CVJ malformation requires instrumented surgery to restore the normal realignment of the bony–ligamentous structure and thus enlarge the spinal canal. In most cases, the main principle of the surgical procedure is distraction to make the odontoid process move away from the CVJ [21]. The surgical approaches and techniques used to accomplish the target are various but are mainly represented by transoral decompression followed by posterior fusion, and posterior decompression and fusion. Kim et al. [22] investigated the role of distraction during surgery for congenital CVJ anomalies with the aid of SSEP and mMEP monitoring; the reported warning criteria were more than a 50% decrease in amplitude or more than a 10% increase in latency (in comparison with the baseline values) for SSEP, and disappearance of the wave for mMEP monitoring. Two patients experienced significant changes in mMEPs during distraction, with subsequent normalization of the potentials after the release of the tension; these two patients also presented with Klippel–Feil syndrome (KFS). The authors stated that there was no univocal relationship between the length of the distraction and the risk of a new postoperative neurological deficit; conversely, the preoperative condition of the spinal cord (e.g. the presence of syringomyelia or oedema) could influence its sensitivity to external manipulations. Patients with KFS may be at higher risk of spinal cord injury because of a narrower spinal canal, hypermobility and an anomalous course of the vertebral artery, in comparison with non-syndromic patients with atlantoaxial dislocation [23]. Overstretching of the spinal cord and the related decrease in blood perfusion are potential causes of neurological injury [24].

During surgery for CVJ malformation, especially basilar invagination, the presence of Chiari malformation type I (CM) is a frequent feature (37–94%), which develops because of progressive herniation of the odontoid process into the posterior fossa, forcing the tonsils into the spinal canal [21]. Apart from CVJ anomalies, surgery for CM itself is considered to be low risk, and there have been few reports of application of IONM in this context. Danto et al. [25] reported that significant SSEP changes (>10% latency prolongation and/or >50% amplitude reduction) occurred in 32% of 500 patients operated on for CM over a 2-year span at the Chiari Institute. Notably, 75% of these changes were related to positioning and 80% of these patients were repositioned following neurophysiological alerts. Such a high incidence of position-related neurophysiological changes was likely due to a unique pattern of patient referral to that institution, including very complex malformations of the craniocervical junction and/or second-look surgeries.

In general, surgery for CM carries a very low risk of neurological injury, to the point that reports of positive IONM findings are merely anecdotal in the literature [26, 27]. Conversely, IONM has played a part in investigating the role of different surgical steps (bony versus dural decompression) in achieving decompression of the lower brainstem (Fig. 1). Some studies [27, 28] have suggested that the most effective improvements in SSEPs and brainstem auditory evoked potentials (BAEPs) are achieved following bony decompression rather than following opening of the dura. Although there is no evidence that these electrophysiological findings have clinical relevance, it is conceivable that a more conservative approach with bony decompression and dural 'tearing' alone, as is common practice in paediatric neurosurgery, may still be effective in achieving brainstem decompression.

More recently, the role of IONM in the positioning of paediatric patients with CM for foramen magnum decompression has been highlighted by Barzilai et al. [29], who reported three cases of position-related attenuation of potentials (SSEPs and mMEPs) without any postoperative deficit. Finally, Skinner described use of accessory nerve (cranial nerve XI) motor evoked potential monitoring during CVJ surgery to augment the sensitivity to spinal cord injury, especially at the level of the C1–C5 ventral horn columns [30].

Tumours at the level of the CVJ include extramedullary tumours (meningiomas, schwannomas, chordomas) and intramedullary tumours. In lesions located within the brainstem or in its vicinity, the roles of BAEPs [31] and mapping of cranial nerve function have proven useful [32]. More recently, monitoring of corticobulbar function with corticobulbar motor evoked potentials was introduced and has proven reliable with regard to neurological outcomes in cerebellopontine angle surgery [33]. Kodama et al. recently reviewed the intraoperative course of long-tract monitoring during infratentorial surgery to evaluate the correlation of mMEP and SEP changes with the clinical outcome, and pointed out that long-tract mMEP and SEP behaviour follows the 'all or nothing' criterion applied to intramedullary spinal cord procedures and is therefore different from the scenario in supratentorial surgery, where minor changes such as an increase in the motor threshold or an amplitude decrement can be followed by long-lasting neurological sequelae. The negative predictive value in their series was very high (0.989), demonstrating the efficacy of long-tract monitoring to predict the postoperative outcome. Conversely, the positive predictive value was 0.467; the authors stated that this might have been explained by the fact that they did not distinguish between permanent and transient alterations in monitoring [34]. In the same study the authors reported a 35% incidence of mMEP and SEP deterioration during posterior fossa surgery, and they pointed out that the risk of such deterioration occurs mainly at the final stage of tumour removal; at this point the higher risk is due to manoeuvres around the brainstem and to the manipulation of perforating vessels. Thus, brainstem ischaemia and vascular damage are the main putative causes of mMEP deterioration during surgery around the lower brainstem [35].

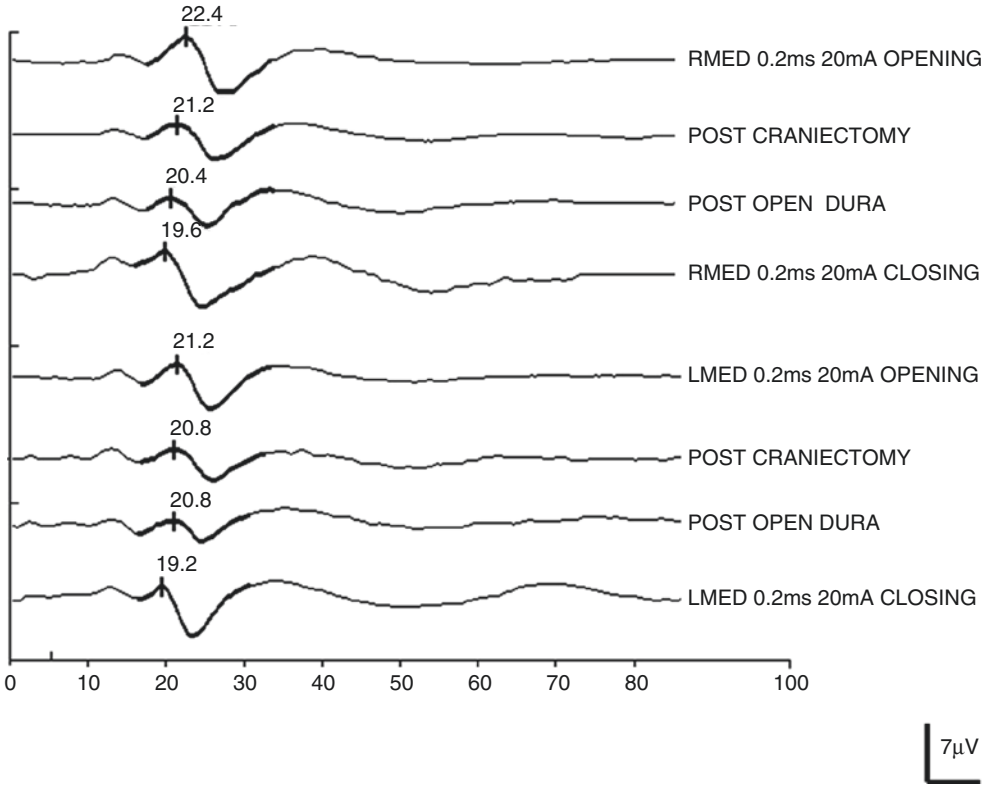

Fig. 1 An 18-year-old woman presented with a long history of headache exacerbated by Valsalva manoeuvres and four-limb paraesthesia. *Upper panels:* Preoperative magnetic resonance imaging (MRI) (**a**) showed a typical Chiari malformation type I with tonsillar herniation >5 mm inferior to the foramen magnum. The patient underwent foramen magnum decompression and duroplasty; postoperative MRI (**b**) showed the results of surgery with enlargement of the cisterna magna and foramen magnum decompression (*white arrowhead*). *Lower panels:* Intraoperative neurophysiological monitoring of left median nerve somatosensory evoked potentials (SSEPs) (*upper four traces*) and right median nerve SSEPs (*lower four traces*) during the stages of surgery: at the beginning of the procedure, following craniectomy and foramen decompression, after dural opening and at closure. There is a progressive reduction in both the left and median nerve SSEP latency following each step of decompression, suggesting an improvement in conduction velocity through the lemniscal pathway

One of the most challenging surgical procedures at the level of the CVJ is surgery for cervicomedullary tumours. True cervicomedullary tumours, per se, behave like true intramedullary spinal cord tumours (ISCTs) and tend to displace the lower cranial nerve nuclei upward. In these cases, surgery should proceed as for a classical ISCT. The tumour is approached through the dorsal longitudinal raphe, initially focusing on SSEPs. As soon as the dorsal column has been displaced laterally and the tumour is exposed, mMEPs and D-wave monitoring becomes critical. At this stage, surgery should proceed in a stop-and-go fashion, based on the neurophysiological feedback, and according to the warning criteria described in Table 1. Whenever the mMEPs and/or the D-wave decrease in amplitude, corrective measures should be taken, which are summarized by the acronym 'TIP': 'T' stands for 'time', 'I' stands for 'irrigation' and 'P' stands for 'blood pressure' or 'papaverine' (Table 2).

The time variable is a critical one. We have consistently observed that if surgery is transiently stopped immediately after mMEPs have disappeared or the D-wave has significantly deteriorated, these potentials often spontaneously recover. Conversely, to ignore these events and continue any cord manipulation may transform a reversible injury into an irreversible one. IONM allows us to tailor the surgical strategy to the spinal cord's level of tolerance of surgical manipulation (traction, compression, coagulation, etc.) during the procedure.

With regard to irrigation, while the mechanism behind the beneficial effect of warm irrigation of the surgical field has not yet been explained, it is a common observation that this facilitates evoked potential recovery. A possible explanation is that irrigation affects the washout and dilution of extracel-

Table 1 Motor evoked potential (MEP) interpretation during surgery for intramedullary spinal cord tumours

D-wave	Muscle MEPs	Motor outcome
Unchanged or decreased by 30–50%	Preserved	Unchanged
Unchanged or decreased by 30–50%	Lost unilaterally or bilaterally	Transient motor deficit
Decreased by >50%		Long-term motor deficit

Table 2 Intraoperative management of motor evoked potential (MEP) changes during intramedullary spinal cord surgery (from [4])

D-wave	Muscle MEPs	Corrective measures	Predicted outcome
Unchanged	Present	None	Unchanged
Unchanged or above 50%	Present with minor changes (decreased amplitude)	Transiently move surgical manipulation to a different area and apply warm irrigation (37 °C)	Unchanged
Unchanged or above 50%	Lost unilaterally or bilaterally	Transiently move surgical manipulation to a different area and apply warm irrigation (37 °C), then transiently stop the surgery and/or improve spinal cord blood flow (using local irrigation with papaverine, slight hypertension). If the MEPs do not recover, surgery can still proceed, since the D-wave is preserved	Transient motor deficit (affecting the involved extremity)
Decreased by >50%	Lost bilaterally	Stop surgery immediately. If the D-wave does not recover, abandon the surgery	Permanent motor deficit
Unmonitorable	Lost bilaterally	Transiently move surgical manipulation to a different area and apply warm irrigation (37 °C), then transiently stop the surgery and/or improve spinal cord blood flow (using local irrigation with papaverine, slight hypertension). If the MEPs do not recover, abandon the surgery	Inability to differentiate between transient and permanent deficit

lular potassium, which may accumulate with the disruption of cell membranes and as a result of depolarization [36, 37]—or, more simply, the effect of the higher temperature may be beneficial to evoked potential recovery.

Local application of papaverine and an increase in the mean arterial pressure are both methods used to improve local perfusion to counteract incipient ischaemia. Sometimes, mMEPs are dramatically correlated with blood pressure values, and sustained hypotension may affect mMEPs and unfavourably affect the outcome [38].

Much care should be taken during the last part of the surgery when the tumour is devascularized from the perforating branches of the anterior spinal artery axis. It is of paramount importance to avoid any coagulation in these vessels and to keep both the mMEPs and D-wave under continuous monitoring.

Corticospinal MEP monitoring (D-wave) during intramedullary tumour surgery provides the most reliable information about the functional integrity of the CST and demonstrates a clear relationship between intraoperative modification and postoperative outcome (Figs. 2, 3, 4, and 5) [6]. The combined use of D-wave and mMEPs may be the most specific and sensitive measure of motor tract integrity during ISCT resection [39], but careful analysis of intraoperative data is needed [7]. Sudden disappearance of both the D-wave and mMEPs is rare and is likely related to a vascular insult caused by bipolar coagulation. D-wave deterioration is usually gradual and thus permits to take corrective measures. Disappearance of mMEPs usually precedes changes in the D-wave, although the D-wave may remain stable or may drop insignificantly despite complete mMEP loss. It should be stressed that these presence and absence criteria for mMEPs should be applied only to spinal cord surgery, not to supratentorial surgery, in which permanent and even transient alterations in mMEPs may result in permanent new deficits [40]. When judging mMEP loss, it should be taken into consideration that the specificity of mMEP monitoring and therefore its predictive value increase with the number of muscles that are monitored [4]. SSEP monitoring in ISCT surgery has proven useful. Kearse et al. [41] prospectively evaluated the relation between intraoperative SSEP changes and clinical outcome, and concluded that deficits were always predicted by SSEP changes; however, a high rate of false positive results was observed. Nevertheless, preservation of SSEPs during ISCT surgery facilitates postoperative functional sensorimotor integration [4]. Although our historical control study on the value of IONM in spinal cord tumour surgery did not focus solely on cervicomedullary tumours, it suggested that the outcome was improved in the monitored patient group as compared with the non-monitored group [4], and there is a broad consensus on the use of IONM as a standard of care for ISCTs [42].

Unlike cervicomedullary spinal cord tumours, true intrinsic, focal medullary tumours tend to displace the lower cranial nerve nuclei ventrally rather than upward [32]. Therefore, it is valuable to perform neurophysiological mapping of the obex to localize the nuclei and the intramedullary roots of the lower cranial motor nerves IX–XII. To do so, a handheld monopolar (or bipolar concentric) stimulating probe can be used. A single stimulus of 0.2-ms duration is delivered at a repetitive rate of 1–2 Hz. By moving the tip of the stimulator 1 mm away, it is then possible to explore the floor of the fourth ventricle and identify the areas with the lowest threshold (which is the one closer to either the nucleus or the

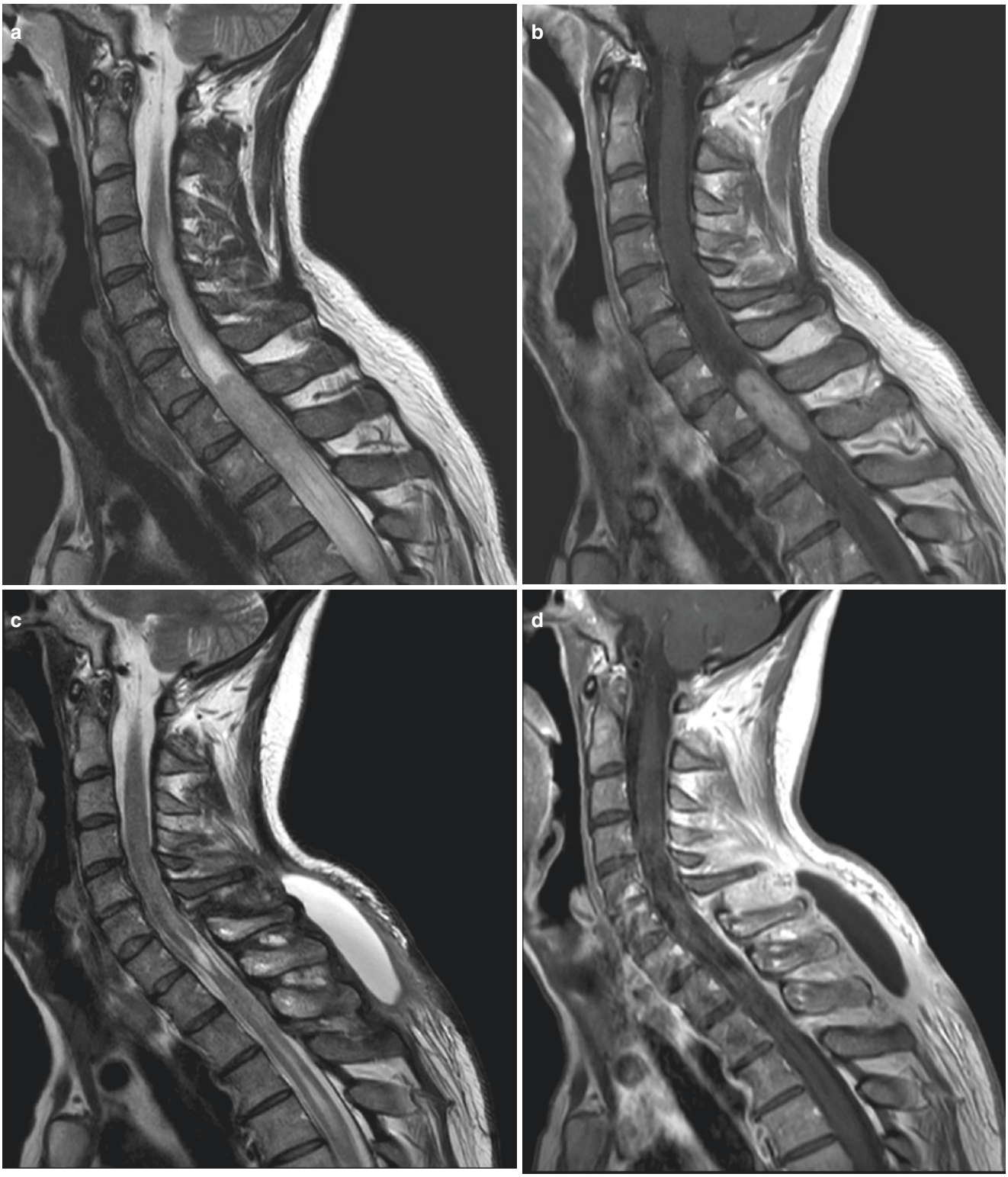

Fig. 2 A 46-year-old woman presented with an 8-month history of bilateral numbness in the inferior limbs. *Upper panels:* Preoperative magnetic resonance imaging (MRI) showed a D1–D2 intramedullary spinal cord tumour with cystic components above and below the tumour (**a**), and homogeneous enhancement by contrast medium (**b**). *Lower panels:* Postoperative MRI 2 months after surgery showed complete removal of the tumour and a reduction in the sizes of the satellite cysts (**c**, **d**); a small pseudomeningocele developed after surgery and disappeared a few months later without further intervention

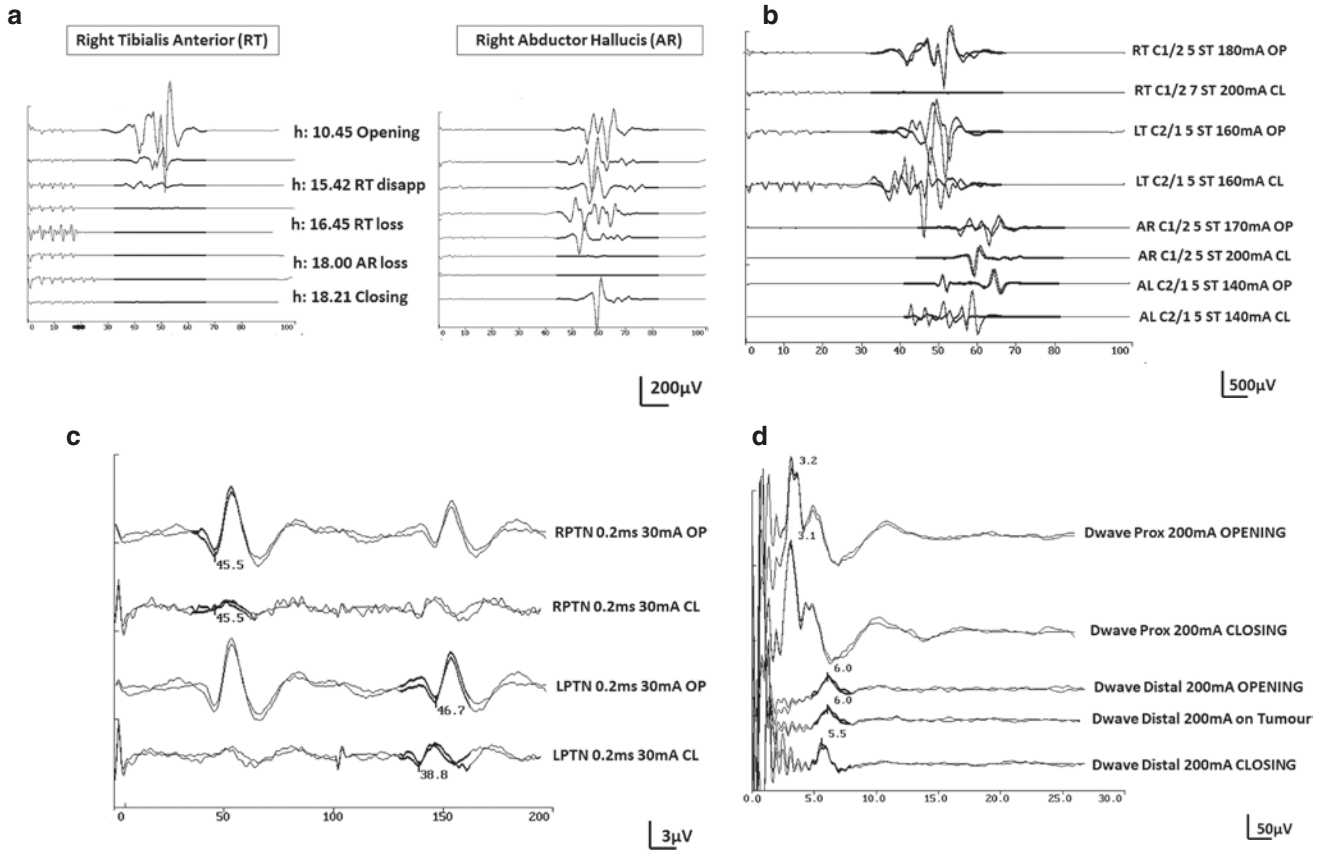

Fig. 3 (**a**) Intraoperative neurophysiological monitoring (IONM) of tibialis anterior and abductor hallucis motor evoked potentials (MEPs) during different stages of surgery. Disappearance of the right tibialis anterior (RT) MEP was reported, and it did not recover despite corrective measures (a transient halt in the surgery, warm irrigation, moderate hypertension); later on, the right abductor hallucis (AR) MEP also disappeared but partially recovered at the end of surgery. (**b**) RT MEPs (*first two traces*) and left tibialis anterior (LT) MEPs (*third and fourth traces*) at opening and closing, showing loss of the RT MEP. (**c**) Somatosensory evoked potentials (SSEPs) of the right posterior tibial nerve (RPTN) (*first two traces on the left*) and left posterior tibial nerve (LPTN) (*last two traces on the right*), showing a bilateral drop in amplitude at the end of the surgery, likely related to the incision of the dorsal median raphe. (**d**) D-wave monitoring with catheter electrodes placed both rostrally (*upper two traces*) and caudally to the lesion (*lower three traces*) during the different stages of surgery; the distal D-wave did not present any relevant amplitude change during surgery. The patient experienced transient paresis of the right inferior limb and gradually recovered within 3 weeks after the surgery; the stability of the D-wave, despite the loss of the RT MEP, was predictive of motor recovery

intramedullary root of the nerve) and with the highest threshold or no response at all. These latter areas are likely to be the safer entry zones, as the nuclei or tracts are far from the tip of the stimulator. To avoid any cardiovascular derangement, no stimulation intensity higher than 2 mA should be used at the level of the medulla. To record the responses from cranial motor nerves IX/X and XII, wire electrodes are inserted into the innervated muscles.

With regard to glial tumours of the medulla, these are usually focal, low-grade astrocytomas, especially in children, and there is no reason to be aggressive from a surgical standpoint, as violation of the ependymal layer with an injury to the underlying nuclei may expose the patient to unacceptable morbidity in terms of dysphagia, loss of the coughing reflex and other life-threatening conditions. With this in mind, however, most tumours in this region can be surgically resected with good results following appropriate perioperative management (airway and respiratory function). Furthermore, in well-demarcated tumours the extent of surgical resection clearly correlates with the long-term outcome; radiation therapy as a primary modality of treatment is inappropriate in such conditions, thus elevating surgery to the main treatment modality. Conversely, surgical treatment of diffusely infiltrative or malignant tumours offers little benefit and should be discouraged [43].

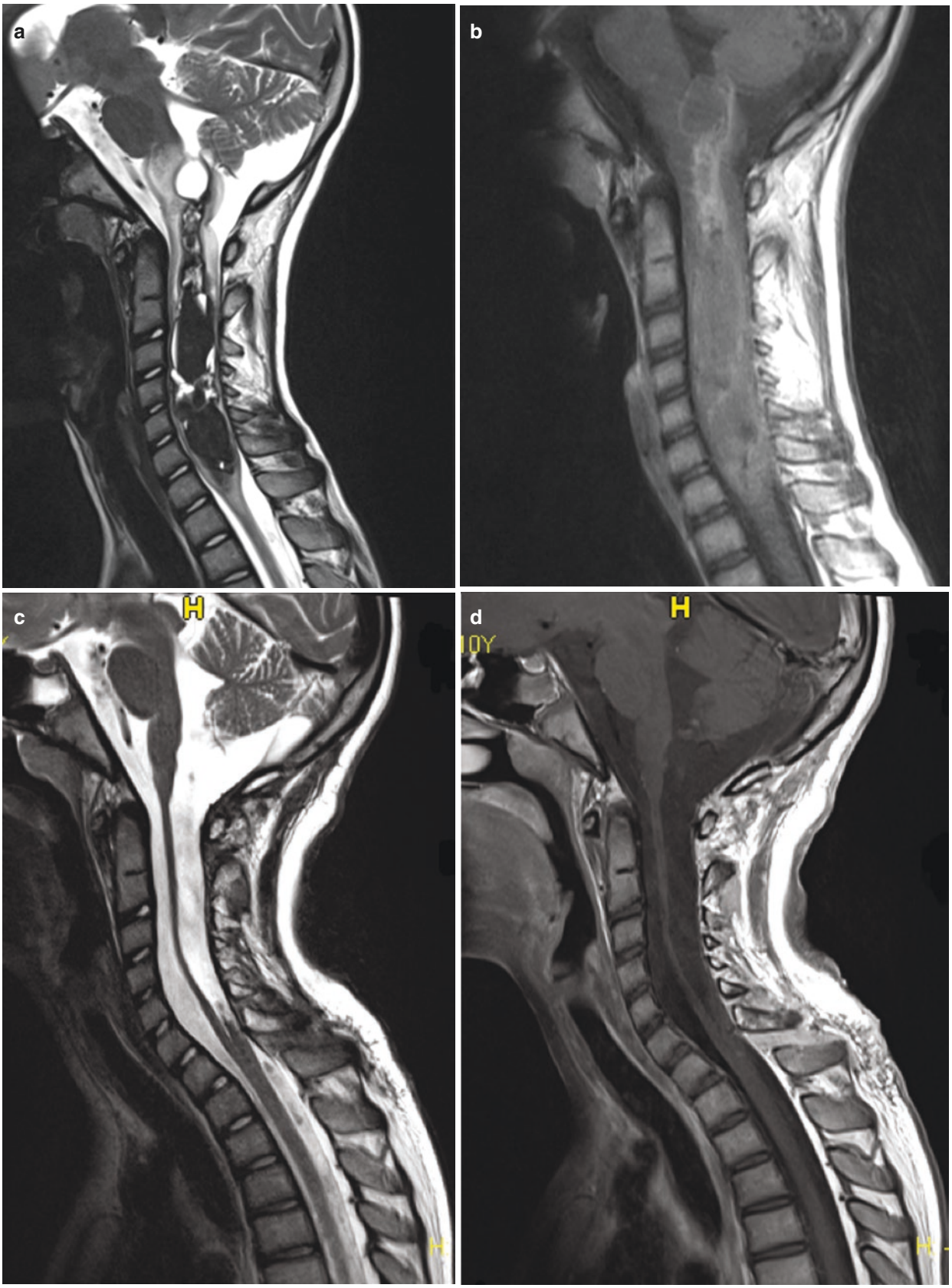

Fig. 4 A 10-year-old boy presented with a short history of neck pain, which progressively worsened. *Upper panels:* (**a**, **b**) Spinal magnetic resonance imaging (MRI) revealed a large cervicomedullary spinal cord tumour extending from the medulla to C7, with heterogeneous enhancement and with cystic components. At surgery, both lower cranial nerves and upper- and lower-extremity muscle motor evoked potentials (mMEPs) were monitored. Postoperative MRI documented total tumour removal (*lower panels*; (**c**, **d**))

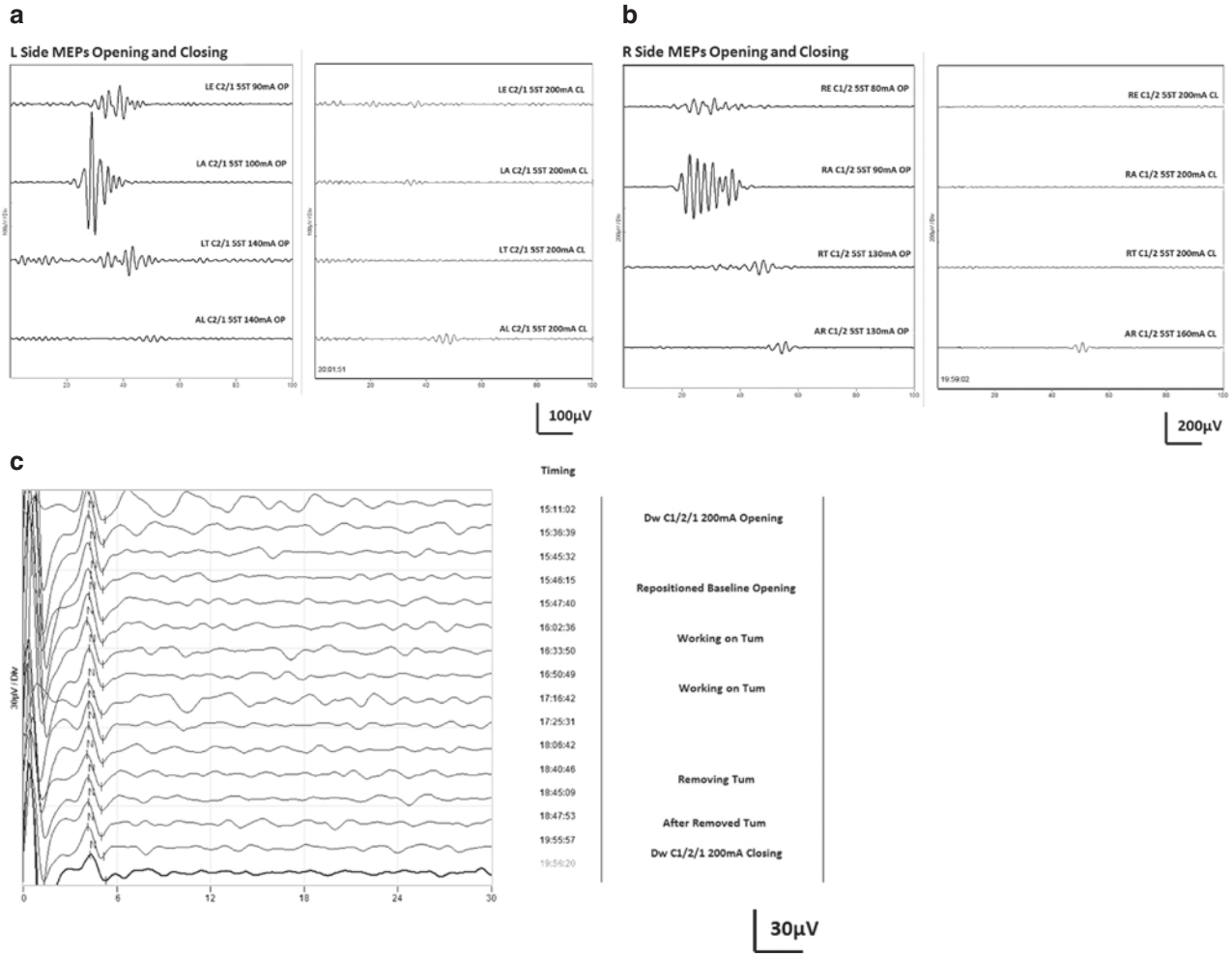

Fig. 5 During surgery, muscle motor evoked potentials (MEPs) from both left- and right-side muscles progressively decreased in amplitude. The worsening was more marked on the right side. The surgery was repeatedly halted to allow for muscle MEP recovery, and the usual corrective measures were taken (warm irrigation, mild hypertension). Despite the pauses and corrective measures, at the end of the surgery a small response was still present for all left-side muscles except for the tibialis anterior (LT) response (**a**), while all right-side muscle MEPs were lost except for the abductor hallucis (AR) MEP (**b**). Nevertheless, D-wave monitoring remained stable throughout the procedure and the amplitude never decreased by more than 20–30%. So, in consideration of the D-wave stability, the decision was made to carry on with the surgery (**c**). Postoperatively the child experienced tetraparesis, which was initially severe on the right side, but at 3-month follow-up he was able to walk independently (McCormick grade II) and he made a full recovery within about 9 months. Cranial nerve muscle MEPs (not shown) also showed various fluctuations during the surgery and an amplitude drop of about 50% at the end of the surgery. The child experienced a transient swallowing problem early after surgery but did not require a tracheostomy or a gastrostomy, and this problem completely resolved within about 1.5 months

Conclusion

Intraoperative neurophysiological monitoring (IONM) is a valuable aid for craniovertebral junction (CVJ) surgery and permits safer surgical management. Application of IONM during positioning, especially in patients with an unstable CVJ, reduces the risk of presurgical neurological injury, and continuous monitoring during surgery provides data on the functional status of the tracts during intramedullary surgery and during instrumented surgery for CVJ instability.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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Management of Anaesthesia

Federica Tosi, Orazio Genovese, Tamara Jovanovic, and Massimiliano Visocchi

Abstract Surgical treatment of the craniovertebral junction (CVJ) requires excellent management by the anaesthetist. Patients undergoing this type of surgery have a wide range of concomitant diseases. Therefore, before proceeding to CVJ surgery, it is recommended to analyse the clinical aspects of the patient that could complicate the outcome of the surgical procedure.

In this paper we aim to establish what constitutes the best surgical and anaesthesia management of these patients. We consider airway management, trying to identify the gold standard for the patient. We also consider the most appropriate intraoperative approach to guarantee the best management of the patient.

Introduction

Anaesthesia and intensive care for the treatment of craniovertebral junction (CVJ) anomalies are difficult. The anaesthetist has to deal with different kinds of pathological conditions ranging from neurosurgical and vascular malformations to acquired disorders (traumas, infections, tumours and metabolic disorders) and congenital bone lesions (Table 1). During childhood the main pathological conditions are congenital. Adult patients present mainly for surgery with one of four pathologies: trauma, infections, congenital lesions or degenerative lesions. The most important factor to consider is the patient's age, since the surgical procedure exposes the patient to potential blood loss. Other important factors to be considered are the extent of the surgical procedure, especially if it involves multiple levels; the position of the patient on the table (for example, prone); and the necessity for a combined approach, both anterior and posterior. The intubation manoeuvre and the position of the patient are made dangerous by the presence of an anomaly in the cervical spinal cord and atlantoaxial instability. Kyphoscoliosis can lead to pulmonary and corpulonal hypertension may be the consequence of kyphoscoliosis. In many cases, patients are subjected to repeated interventions with repeated risks related to multiple anaesthesia. Therefore, in this type of surgery, a comprehensive approach to patients is recommended, including complete metabolic, neurophysiological and cardiorespiratory assessment.

Table 1 American Society of Anesthesiologists (ASA) physical status classification system

Classification	Definition
ASA I	A normal healthy patient
ASA II	A patient with mild systemic disease
ASA III	A patient with severe systemic disease
ASA IV	A patient with severe systemic disease that is a constant threat to life
ASA V	A moribund patient who is not expected to survive without the operation
ASA VI	A declared brain-dead patient whose organs are being removed for donor purposes

Preoperative Anaesthesia Assessment

A general assessment of the patient's physical state is done using the classification system established by the American Society of Anaesthesiologists (Table 1), which considers the patient's physical state regardless of the surgical event he will undergo and the possible coexistence of malformations or physiological problems associated with pathogenic conditions that need to be dealt with. Anaesthetists should also not

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underestimate the importance of the patient's psychological background, especially in patients whose interventions are being repeated over and over again.

Intubation Versus Tracheostomy

Whether intubation or tracheotomy is the best choice for optimal airway management is agreed between the anaesthetist and the surgeon. This decision must be made considering, first of all, anatomical characteristics that could limit the intubation manoeuvres and/or an in-depth evaluation of the risk of execution of the surgical procedure itself, as well as the risk of upper airway oedema as a dangerous postoperative complication.

A careful assessment should be made to identify previous difficult intubation, restriction of neck movement or instability of the cervical spine.

The stability of the spine must be checked preoperatively with both clinical assessments (of both pain and neurological deficit) and computed tomography (CT) scanning of the vertebral column.

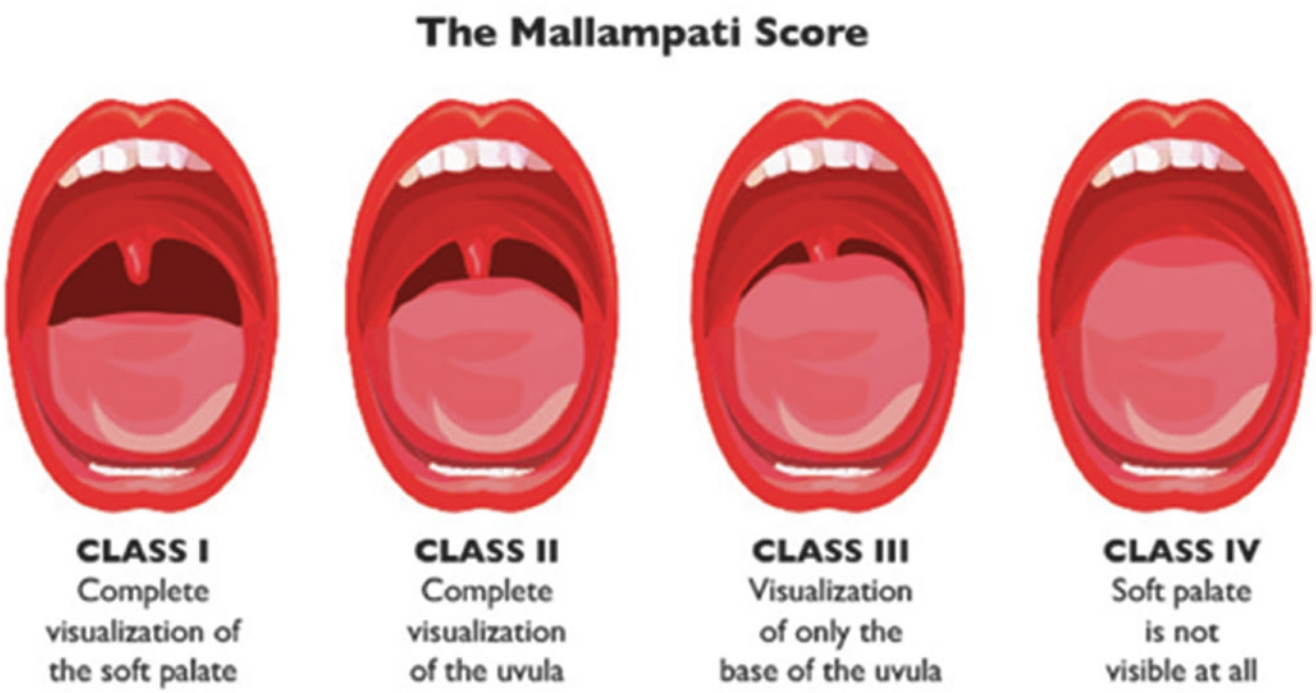
For this goal, either plain lateral films or CT scans of the spine are useful. For example, above-C2 unstable injuries include a Jefferson burst fracture, lesions of the transverse atlas ligament, condylar fractures, and lesions of the alar and tectorial ligaments [1].

During preoperative assessment the anaesthetist must decide whether to awake intubate the patient or not. Indications for awake intubation include the presence of a full stomach (as can happen in an emergency setting), the necessity to evaluate the neurological status after intubation in the presence of spine instability or the presence of a neck stabilization device such as a halo device. Direct laryngoscopy can contribute to vocal cord injury, but, if team members help each other with manual stabilization of the head, it is considered an accepted means of intubation provided that one can ensure absence of neck movement. A further aspect to be taken into account is the choice between an anterior or posterior surgical approach [2, 3].

Micrognathy, mandibular hypoplasia and macroglossia are the main causes of difficult intubation as the mobility of the soft palate during laryngoscopy is made difficult because the space between the hyoid bone and the jaw is limited. Goldenhar syndrome and Klippel-Feil syndrome are characterized by difficult intubation due to hypomotility between the cervical vertebrae and the atlanto-occipital joint. Compression of the cervical cord requires a clinical examination and an accurate patient history [4, 5]. In mucopolysaccharidosis type IV (i.e. Morquio syndrome), choosing orotracheal intubation with a laryngoscope can expose the anaesthetist to major problems with airway management given the macroglossia and the reduction in the ventilatory space. Difficulty in airway management pertains to masked ventilation and/or laryngoscopy and/or tracheal tube placement.

There are some evaluation scales that can be used to try to prevent difficulty in managing the airway. The Mallampati classification, which is based on anatomical structures when the patient opens his mouth (Table 2), is one of these. A

Table 2



Mallampati score of III or IV may be suggestive of a potentially difficult intubation or ventilation [6]. In addition to the Mallampati test, there is also the upper lip bite test (ULBT), which is used to evaluate the airway in older children (Table 3). A higher ULBT classification may be predictive of difficulty in managing the airway [7]. During laryngoscopy, we use the Cormack and Lehane classification (Table 4). Intubation is likely to be difficult in patients with a Cormack grading of 3 or 4 during laryngoscopy manoeuvres.

It is important to know that atlanto-occipital and atlanto-axial rotation movements performed during laryngoscopy can endanger the patient. There are now various devices (videolaryngoscopes) available that allow us to minimize flexion–extension movements of the cervical spine. For example, studies have shown that with an airway scope there was 39% less extension of the cervical spine between the occiput and C4, and 42% less movement of the atlanto-occipital region [8, 9].

Difficulty in management of the airways can, as previously mentioned, involve difficulty in ventilation or intubation. Intubation is considered difficult when an expert anaesthetist is not able to intubate the patient after three attempts. In this case, fibre-optic intubation is strongly recommended in the case of a surgical procedure on the CVJ, but tracheotomy should also be considered.

Use of fibre-optic technology requires an expert operator. The last few years have seen the introduction of optical fibres that are suitable for very young children and therefore are also suitable for anatomically altered airways or compromised air spaces. This technology offers a safe and effective method of airway management (which can otherwise be very difficult to perform in uncooperative patients and children), consequently overcoming the difficulties encountered in the past when only an adult-sized fibre-optic apparatus 6 mm in diameter was available. There are intubation support devices available, such as the aforemen-

Table 3

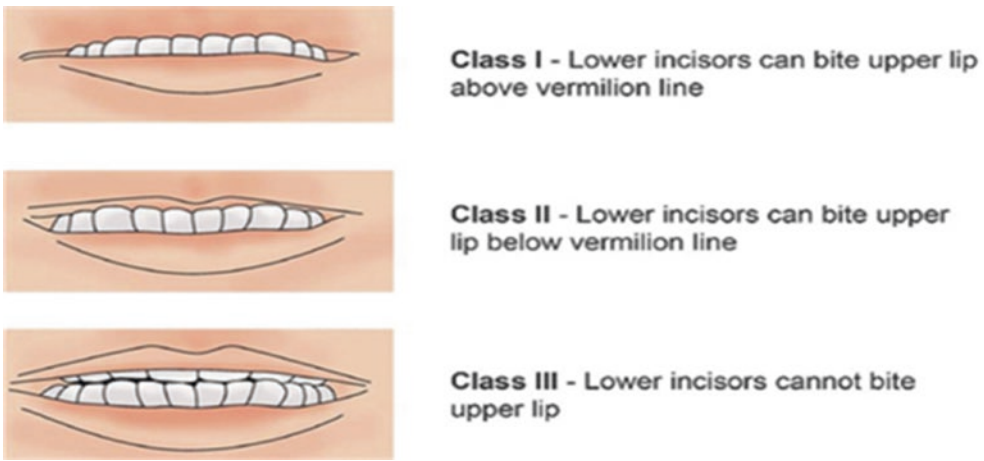
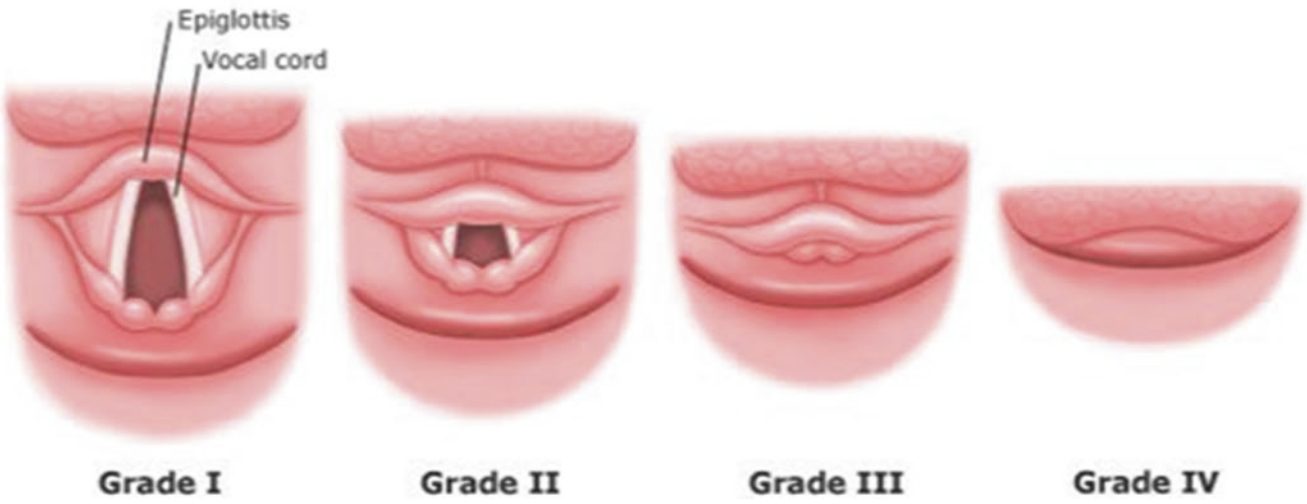


Table 4



tioned videolaryngoscopes or laryngeal masks that allow intubation [4, 5].

Should we intubate or tracheostomize? With agreement between the anaesthetist and the surgeon, if the postoperative course involves massive oedema, then it is clear that a decision to perform a preventive tracheostomy is certainly the wisest choice. In addition, this choice allows quicker reawakening of the patient than intubation does, with all of the advantages resulting from this choice rather than prolonged intubation [10].

This is the reason why better intubating devices such as video laryngoscopes are able to provide a better laryngeal view than direct laryngoscopy [11].

Although the terms 'tracheostomy' and 'tracheotomy' are often used interchangeably, it should be remembered that 'tracheostomy' (which comes from the Greek word *stomoun*) refers to the act of creating an opening, whereas 'tracheotomy' (which comes from the Greek word *tome*, meaning 'to cut') specifically refers to the result of a surgical cut. The need for an external cervical orthosis, such as a halo vest, suggests that a tracheostomy should be performed to avoid the need for an emergency procedure to treat overwhelming respiratory distress in the postoperative phase.

Choice of Surgical Route

Two surgical routes can be chosen by the paediatric neurosurgeon: the anterior route, to deal with direct decompression of the cervical spinal cord; and the posterior route, to deal with neurological decompression, and for instrumentation and fusion of the cervical spine [12].

The Anterior Route

Orotracheal intubation can be chosen in cases of an endoscopic transnasal approach, since no conflict with the surgeon's activity is provoked. The choice of orotracheal intubation may be more debatable in cases of transoral, transmandibular or transmaxillary approaches, as some surgeons do not worry about possible conflict between the pharyngeal spreaders of the oral distractor and the orotracheal tube, whereas others prefer nasotracheal intubation to be performed by the anaesthetist to reduce the chance of possible conflict with the oral distractor, and others prefer to keep the surgical channel free of any foreign body by using a tracheostomy.

The Posterior Route

In cases of a posterior surgical approach (for decompression and for instrumentation and fusion techniques), a tracheostomy is not used for surgical requirements, since the airways are far from the surgical target. Nevertheless, in patients with pre-existing brainstem damage, which could increase the likelihood of postoperative respiratory disturbance and necessitate use of a halo vest, a tracheostomy could be advocated [2, 3].

Sedation and the Drug Delivery System

Sedation during spontaneous breathing before laryngoscopy is necessary to evaluate adequate oxygenation and ventilation. Anaesthetic induction is carried out with non-irritating agents (such as sevoflurane) or intravenous drugs (such as propofol and remifentanil). It is useful to consider the use of a fibre-optic bronchoscope in cases of difficult intubation.

The use of optical fibre techniques has increased in selected paediatric patients, thanks to the combined techniques of topical anaesthesia with local airway blocks and intravenous sedation, or general anaesthesia with use of a mask with maintenance of spontaneous ventilation [13].

The ideal dimension of the fibrescope is modified in relation to the diameter of the endotracheal tube (ETT) to be crossed. Intubation requires an ETT dimension only slightly larger than the diameter of the fibrescope. Modern flexible fibrescopes reach a diameter of 2 French and allow passage of a 2.5-French endotracheal tube.

Dexmedetomidine produces moderate sedation without respiratory distress, apnoea or haemodynamic instability, so it can be successfully used in patients who need awake fibre-optic intubation for cervical spine surgery [14]. Electrocardiographic monitoring with pulse oximetry and pressor monitoring are required throughout the procedure in all patients. Maintenance of anaesthesia is achieved with sevoflurane (minimum alveolar concentration (MAC) 1, 1.5) in O_2 plus air, with a target partial pressure of carbon dioxide ($paCO_2$) of 32–35 mmHg. Muscle relaxation is obtained with cisatracurium besilate.

Induction

Intravenous or inhalation induction depends on the difficulty of managing the airway and the physical condition of the patient [15, 16]. The use of succinylcholine in such patients is no longer recommended, as use of this drug has been completely abandoned by many centres because of the numerous complications caused by its use [17].

Maintenance

During maintenance we need to use a technique that is compatible with somatosensory evoked potential (SSEP) and motor evoked potential (MEP) monitoring. A technique involving nitrous oxide 60% and isoflurane <0.5 MAC is acceptable [17], but an intravenous technique using propofol is recommended. Neurophysiologists should be aware of any decrease in arterial pressure or the need to administer a bolus of an opioid or another agent.

Intraoperative Monitoring

Respiratory monitoring should include end-tidal CO_2, peak airway pressure and serial measurement of arterial oxygen tension. The temperature should be monitored, intravenous fluid should be warmed and a warm air mattress device is recommended.

Invasive arterial pressure assessment is mandatory. Prolonged anaesthesia, combined with blood loss, necessitates detailed monitoring of the cardiovascular system.

Intraoperative Neurophysiological Monitoring

Assessment of the functionality and control of the spinal cord during surgery is now possible through the use of intraoperative monitoring techniques [18]. The integrity of the spinal cord can be evaluated with different techniques, which include:

1. The alarm test
2. Somatosensory evoked potentials
3. Motor evoked potentials
4. Dermatomal responses

The Wake-Up Test

One of the first spinal cord function tests to be used is an evaluation of the patient's intraoperative motility by awakening him for a period of time to evaluate his movements. This test has been used during corrective procedures on the spine, but the complications of the test have not been underestimated: accidental extubation, detachment of fixers and gaseous embolism on deep inspiration. The main limitation of this test is that the spinal cord is evaluated only in a small phase without taking into account the phases in which the patient is subjected to deep narcosis. Another limitation of this test is the fact that it cannot be applied to infants and young children. Alternative tests such as tetanic stimulation of the lower leg and observation of the clinical response can be used in those age groups.

For the awakening test, all possible anaesthetic choices can be used.

To perform this test, many anaesthetic techniques have been advocated. Koscielniak-Nielsen suggested use of midazolam, instead of propofol, for the maintenance of these patients. The presence of a midazolam antagonist could make this intraoperative awakening test much easier.

Use of midazolam has been associated with a shorter intraoperative wake-up time, a better quality of wake-up and a shorter postoperative wake-up time. Even remifentanil has a pharmacokinetic profile suitable for this test, with a delay of only 5 min between the request for the test and adequate conditions for neurological assessment.

Somatosensory and Motor Evoked Potentials

The use of SSEPs is now the most widely used method for study of the spinal cord [19]. Through stimulation of peripheral nerves, the responses at the level of the cerebral cortex are observed by using superficial electrodes placed on the scalp and deeper electrodes at the epidural site. Basal SSEPs are recorded in order to exclude neurological dysfunction.

The stimulus is applied to the peripheral nerve on both sides as a square wave at 3–7 Hz, and the response is recorded with electrodes over the somatosensory cortex on the scalp. The intensity of the stimulus is typically in the range of 25–40 mA. Baseline data are obtained after skin incision to assess the effect of the anaesthetic agents on the amplitude of the recording. During surgery, responses are recorded repeatedly, comparing the amplitude and the latency of the response to evaluate the integrity of the sensor spinal pathways. A 0.50% decrease in the SSEP amplitude or a 10% increase in the latency are deemed significant. It should be stressed than the blood supply of the motor pathways differs from that of the sensory tracts. It is therefore possibly to have normal recordings from the SSEPs but a tetraplegic patient postoperatively. Numerous anaesthetic agents, the level of surgical stimulation and hypothermia may alter the SSEP results, while other factors such as hypothermia, inhalation anaesthetics, hypercapnia and hypoxia suppress both SSEPs and MEPs. As for the muscular artefacts that are recorded on SSEPs, they are reduced by a neuromuscular block induced by curarization.

It is essential to use both SSEPs and MEPs in spine surgery. It should be stressed, however, that MEPs are the most effective instrument available to evaluate the motor integrity of the spinal cord [20].

Electrical or magnetic means can stimulate the motor cortex, and myogenic or neurogenic responses are recorded. The myogenic response of a stimulated muscle is evaluated by electromyography (EMG) and abolished by neuromuscular blockade.

Even with a neuromuscular block, it is possible to obtain neurogenic responses in a peripheral nerve or in the spinal cord.

Anaesthetic agents have a greater impact on cortical evoked responses than on spinal responses. Propofol is a potent suppressor of cortical evoked responses, midazolam has a lesser effect and opioids do not seem to cause effects.

Dermatomal Potentials

Dermatomal somatosensory evoked potentials (DSEPs) give us information about the brain response to a certain stimulated dermatomal. It is a pure sensory input into any level of the spinal cord. Unfortunately, the literature concerning this monitoring is not sufficient, and this makes this procedure difficult to apply.

Competing Interests The authors declare that they have no competing interests.

Compliance with Ethical Standards No financial support was received for this work.

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