51 Upper Cervical and Craniocervical Decompression

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SUMMARY OF KEY POINTS

- A diverse set of pathologies affects the craniocervical junction, including traumatic, congenital, degenerative, vascular, and neoplastic pathologies.
- Traditionally, the craniocervical junction has been treated via transoral, lateral extrapharyngeal, and posterior approaches.
- Improved instrumentation and endoscopic techniques now allow for a broadened number of less invasive approaches to the craniocervical junction.
- Advances in surgical adjuncts, neuroimaging, anesthesia, and postoperative care have improved treatment outcomes of craniocervical junction pathologies.
- Minimization of vascular injury and cerebrospinal fluid fistulae formation may contribute to improved surgical outcomes.

The posterior fossa and craniocervical junction (CCJ) house critical neurovascular structures; their compromise may result in devastating injury. Neural and vascular compression in this region may cause deficits related to the cerebellum, brain stem, and spinal cord. Operative decompression of endangered structures alleviates neurologic dysfunction. Radiographically, the presence of obvious correlative pathology in this region makes the decision to operate straightforward. Discordance between imaging, history, and neurologic examination is more challenging and requires further workup. Complication avoidance and management are paramount to good surgical outcomes. This chapter focuses on indications for surgical intervention, choice of approach, and pearls for complication avoidance when addressing pathologies in the posterior fossa, CCJ and upper cervical spine.

PATHOLOGY OVERVIEW

The CCJ may be the site of complex and diverse pathologies (Table 51-1).[1] In general, most surgical lesions can be approached dorsally through a posterior fossa craniectomy with or without a C1 laminectomy. Abnormalities can be divided into two basic categories: congenital/developmental abnormalities and acquired pathologies. The various pathologies are summarized in Table 51-2.

VENTRAL APPROACHES

There are several accepted routes to the ventral clivus and upper cervical spine. Standard approaches to the ventral cervical spine are limited by the mandible and oropharynx.[2-4] Transoral and endonasal routes to the lower clivus and upper cervical spine are safe for addressing pathologies that cause craniocervical instability and that compress neurovascular structures from the clivus to the C4 vertebral body. In 1917, Kanavel described the first transoral procedure, removal of a bullet lodged between the clivus and ventral atlas.[5] Fang and Ong later reported the first series of six patients who underwent transoral decompression for atlantoaxial instability or congenital anomalies; a high complication rate contributed to slow adoption of this approach.[6]

A resurgent interest in transoral and endonasal approaches led to the refinement of techniques and instrumentation.[7,8] Modern antibiotics and instruments have revolutionized and revitalized this operation. Most notably, improvements in dural repair techniques have lowered the incidence of cerebrospinal fluid (CSF) leaks and infections.[9] Although an extrapharyngeal approach can be used to access the upper cervical region, it is technically difficult.[10] The authors of this chapter prefer the transoral or the endoscopic approach to access extradural pathology and a lateral approach for intradural lesions.

MICROSCOPIC TRANSORAL ODONTOIDECTOMY

Surgical Technique

General anesthesia is used for all ventral cases, including a reinforced endotracheal tube to preserve the airway. Cervical stability is maintained by using fiberoptic endoscopy or awake-intubation techniques if required. The institutional policy of the authors requires intraoperative monitoring of somatosensory evoked potentials (SSEPs) and motor evoked potentials (MEPs). Baseline recordings are obtained before final positioning of the patient.

The patient is placed in the supine position on a standard operating room table with the head fixated in a Mayfield three-pin head holder (Codman, Inc., Randolph, MA) or a halo-ring adapter. A Spetzler-Sonntag transoral retractor (Aesculap, San Francisco, CA) is positioned into the mouth to form a rectangle of exposure. The palate is elevated cephalad, while the tongue and endotracheal tube are retracted caudally. The tonsils and lateral oropharyngeal walls are covered with moist gauze and retracted laterally (Figs. 51-1 and 51-2). Transnasal catheters and palatal retraction sutures can be used in lieu of the Spetzler-Sonntag retractor. If the mouth cannot

TABLE 51-1 Pathology of the Craniocervical Junction

Location	Congenital	Acquired	Primary Neoplastic	Secondary Neoplastic	Intra/Extradural	Neural "Tumors"
Clivus and foramen magnum	Segmentation failure	Basilar invagination Basilar impression (e.g., Paget disease, rickets, osteogenesis imperfecta, acro-osteolysis, rheumatoid arthritis) Paramesial invagination (e.g., achondroplasia)	Eosinophilic granuloma Fibrous dysplasia Chordoma Chondroma Chondrosarcoma Plasmacytoma	Prostate Breast Nasopharyngeal carcinoma Ectopic pituitary	Neurofibroma Meningioma Chordoma glomus tumor Rhabdomyosarcoma	Tumors of brain stem, cerebellum Aneurysms Arachnoid and ependymal cysts Chiari II malformation
Atlas	Assimilation with segmentation failure	Stenosis (e.g., achondroplasia), chronic dislocation (e.g., Morquio syndrome, Down syndrome, rheumatoid arthritis, other arthropathies)	Chordoma Chondroma Giant cell tumor Osteoid osteoma Osteoblastoma	Metastasis Plasmacytoma Local extensions of primary malignancy	Neurofibroma Meningioma Chordoma	Spinal cord glioma Syringohydromyelia Chiari malformation
Axis	Segmentation failure Os odontoideum Neurenteric cysts	Basilar invagination, basilar impression (e.g., Paget disease, rickets plus hyperparathyroid arthropathies, osteogenesis imperfecta, rheumatoid arthritis, skeletal dysplasias) Chronic dislocations Osteomyelitis	Aneurysmal bone cyst Plasmacytoma Chordoma Giant cell tumor Osteoblastoma Chondroma	Metastasis Local extensions of primary malignancy	Meningioma Neurofibroma	Spinal cord glioma Syringohydromyelia

Modified from Menenzes AH: Pathology encountered at the craniocervical junction. Operative Tech Neurosurg *8:116–124, 2005.*

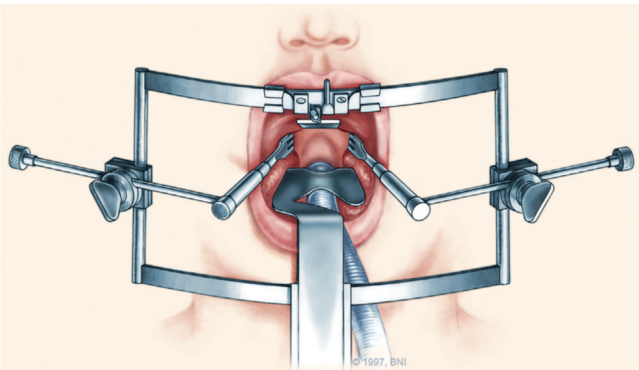

Figure 51-1. Placement of the Spetzler-Sonntag retractor system. The lower blade holds the reinforced endotracheal tube and tongue. The upper blade retracts the soft palate away from the oropharynx. The depth of the lateral "fork" retractors can be adjusted to retract the tonsils first and then the soft tissue of the back wall of the oropharynx as the operation progresses. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

be opened sufficiently, a mandible-splitting approach can be used along with a tracheostomy before the surgical approach begins.[11]

After the retractor is placed in its final position, the oral cavity is cleansed with Betadine. A preoperative dose of antibiotics that covers for oral flora is administered; the authors prefer cefepime and metronidazole. The surgeon stands at the patient's head; adjustments in orientation will be necessary for the duration of the case. An operating microscope is brought into the field. Stereotactic guidance or fluoroscopy may be used to guide the angle of approach and to confirm the location before the palatal incision is made.[12] Electrocautery is used, and the incision proceeds down to bone. Subperiosteal dissection exposes the lower clivus, atlas, and axis. It is seldom necessary to divide the soft palate unless targeting the upper clivus. Self-retaining retractors maintain the exposure.

A high-speed drill is used to remove the ventral arch of C1, which is the load-bearing portion of the bone.[13] A portion of the arch should be preserved to prevent spread of the lateral mass and to maintain orientation relative to midline. Approximately 70% of patients undergoing ventral decompression require supplemental internal fixation, reaching 90% in patients with rheumatoid arthritis.[14] The remaining anterior tubercle of C1 is left as a landmark for posterior fixation (Fig. 51-3A-C). Stereotactic navigation may be useful in cases requiring hardware fixation, especially if a large ventral portion of C1 and surrounding bone must be removed. Long-handled curettes and rongeurs as well as bipolar cautery are useful adjuncts for removing bone and tissue (Figs. 51-4 through 51-6). The average distance between the vertebral arteries is approximately 3 cm. The anatomic "safe zone" is within 1.5 cm on either side of midline (Fig. 51-7). Preoperative radiographs must be evaluated carefully. In many cases, computed tomography (CT) angiography can delineate aberrant or asymmetric vasculature. Further lateral dissection places the

TABLE 51-2 Origin of Pathology at Craniocervical Junction

Congenital
Occipital Bone Malformations
Occipital vertebrae
—Clivus segmentation
—Remnants around foramen magnum
—C1 variants
—Dens segmentation
Basilar invagination
Condylar hypoplasia
Assimilation of atlas
Atlas Malformations
Atlas assimilation
Atlantoaxial fusion
Incomplete arch
Axis Malformations
Segmentation abnormalities
Dens dysplasias
—Os odontoideum
—Ossiculum terminale persistens
—Hypoplasia/aplasia
C2-3 segmentation failure
Developmental or Acquired
Foramen Magnum Abnormalities
Secondary basilar invagination
—Rheumatoid arthritis
—Rickets
—Paget
—Osteomalacia
Foraminal stenosis (e.g., achondroplasia)
Atlantoaxial Instability
Metabolic errors (e.g., Morquio syndrome)
Down syndrome
Infectious
Inflammatory (e.g., rheumatoid arthritis)
Trauma
Os odontoideum
Tumors (e.g., neurofibromatosis, syringomyelia)
Miscellaneous

Modified from Menenzes AH: Pathology encountered at the craniocervical junction. Operative Tech Neurosurg *8:116–124, 2005.*

hypoglossal nerve, vertebral artery, and cervical neurovascular bundle at risk.[15]

During a transoral odontoidectomy, cautious dissection and a methodical approach are essential to minimize the risk of CSF fistula. The superior ligamentous complex (consisting of the apical and paired alar ligaments) must first be sectioned to remove the dens, which can be done with curved curettes. This region often strongly adheres to the dura, and great care must be taken to avoid durotomy.[16,17] A thin shell of bone can be left to avoid a dural tear. Once the last of the odontoid is removed, the frameless guidance or a lateral radiograph can be checked with a radiopaque instrument.[18]

Hemostasis is achieved using bipolar cautery and thrombin hemostatic matrix. Dural integrity is assessed with a Valsalva maneuver for 10 seconds. The pharynx is closed in a single layer with an absorbable suture. An enteric feeding tube is placed before anesthesia is reversed. Patients are left intubated 24 to 48 hours to avoid the trauma of reintubation should it become necessary. A feeding tube is placed under microscopic vision to avoid violating the mucosal suture line; fluid intake is allowed 1 week postoperatively. A swallow evaluation and modified barium swallow study may be undertaken before oral feedings are resumed. Dorsal stabilization may be

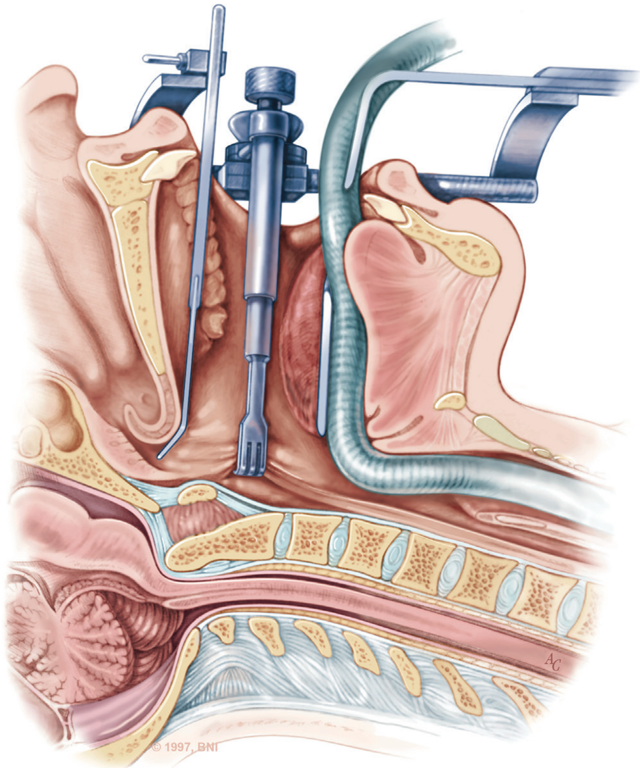

Figure 51-2. Lateral view of the retractor system in place. The lateral retractors lift the soft tissue up and away from the resection bed. Retraction of the palate and tongue provides excellent exposure. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

performed during the same sitting or delayed several days to allow reassessment.

ENDOSCOPIC TRANSNASAL ODONTOIDECTOMY

Surgical Technique

The expansion in endoscopic endonasal approaches allows for novel approaches to the clivus and upper cervical spine.[19,20] Image guidance is of paramount importance in planning and executing endoscopic endonasal approaches to the clivus.[21] Thin-cut computed tomography (CT) and magnetic resonance imaging (MRI) of the cervical spine are useful for assessing the bony anatomy and other critical structures during the approach. If necessary, patients are placed in halo traction for immobilization to prevent risk of injury during positioning.

Intubation is performed using an armored (reinforced) oral endotracheal tube, and the patient is affixed to the table using a standard Mayfield head holder. The patient is typically in a neutral position. Fluoroscopy is then used to obtain a true lateral view, and CT- or MRI-based intraoperative image guidance is registered for optimal trajectory to the clivus or upper spine.

Although both nostrils can be used for access, a right-handed surgeon commonly prefers to access endonasal pathology via the right nostril (Fig. 51-8A). Various endoscopes, including 0-degree, 30-degree, and 45-degree endoscopes, can be used to visualize the surgical field (Fig. 51-8B). A mounted digital camera system is used for most cases (Karl Storz GmbH & Co., Tuttlingen, Germany). The middle turbinate can be resected or outfractured to expose the nasal mucosa, which is then mobilized using cautery in a vertical

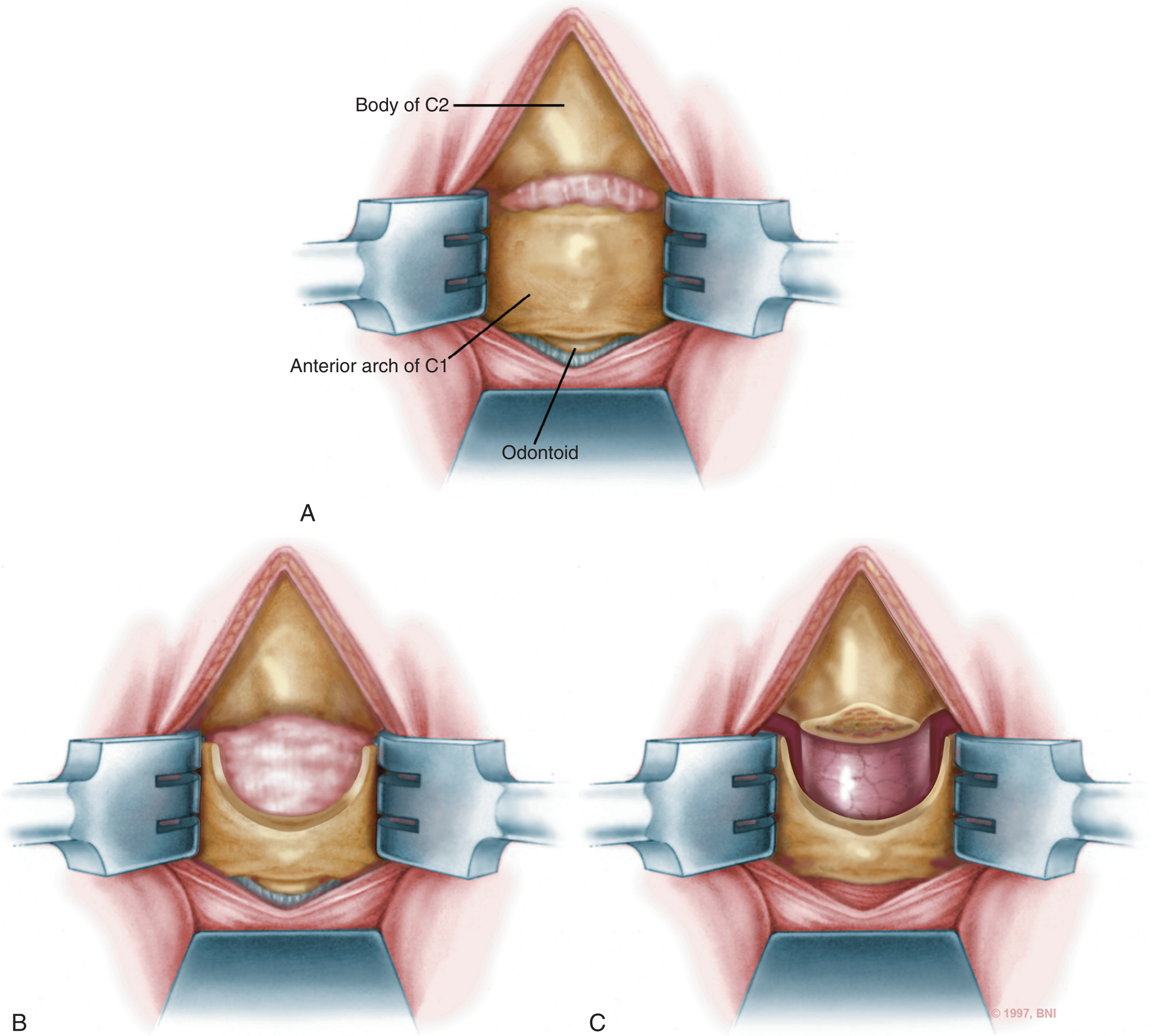

Figure 51-3. Stepwise resection of the odontoid. The back wall of the oropharynx is incised from the top of C1 to the bottom of C2. Subperiosteal dissection is extended laterally. The anterior arch of C1 is identified, and the midline is maintained by preserving part of the anterior arch. Frameless stereotactic guidance is also helpful **(A)**. The anterior arch of C1 is partially drilled away, and the lesion encompassing the odontoid is clearly in view **(B)**. The posterior portion of the odontoid is removed, and the dura is visible. The superior part of the anterior arch of C1 is intact **(C)**. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

incision 1.5 cm from the root of the nasal septum. Periosteal dissection is continued, and the bony nasal septum is fractured to expose the sphenoid rostrum. The optimal trajectory to the clivus can be confirmed using intraoperative navigation.

Once the optimal trajectory is selected, a self-retaining nasal speculum is used to retract the mucosa and further expose the lower clivus and the anterior tubercle of C1. High-speed drills and other microsurgical instruments are used to carry out bony removal of the clivus at the midline, which exposes the transverse and apical ligaments as well as the odontoid process.

In cases of unplanned durotomy or planned intradural work, vascularized pedicles or autologous fat graft can be used to seal areas of CSF leak after primary closure.

Pitfalls and Complication Avoidance

Because many patients exhibit significant debilitation preoperatively, their postoperative needs may include parenteral nutrition, long-term rehabilitation, or placement in a skilled-nursing facility with a long convalescent period. The surgeon must ensure that patients and their families have appropriate and realistic expectations about outcomes and that patients and their families understand the possible risks and complications associated with the procedure.

Intraoperatively, checking and rechecking midline orientation and depth of resection is key to safety. Dissection planes may be obscured by local pathology and may never emerge during the course of a case. As the resection proceeds deeper, meticulous dissection is crucial. A clean and blood-free field

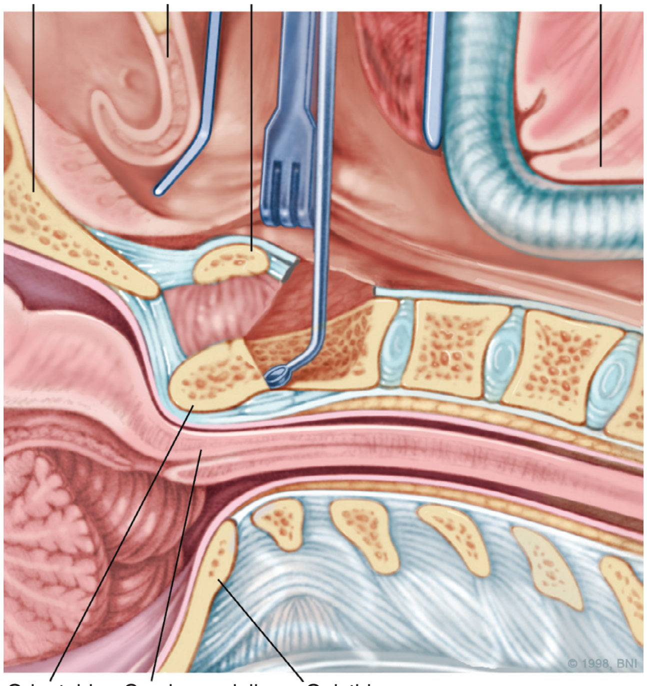

Figure 51-4. Decompression of the cervicomedullary junction by resection of the odontoid. A midline incision is made in the oropharynx, and a long, curved curette is used to resect the bone or mass. Orientation to midline is crucial during the compressive stage. A portion of C1 should be left to maintain the appropriate trajectory. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

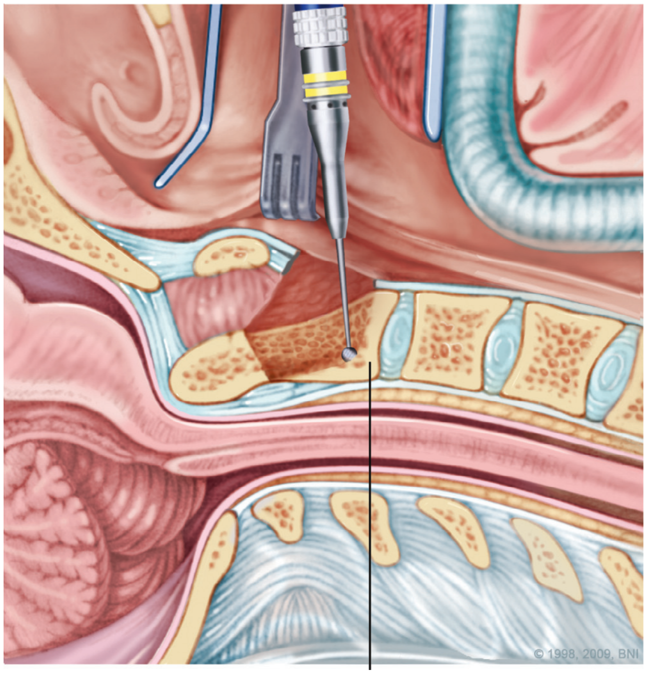

Figure 51-5. Further decompression is achieved with a high-speed drill. A handpiece with an angled bur works well in the depths of the resection bed. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

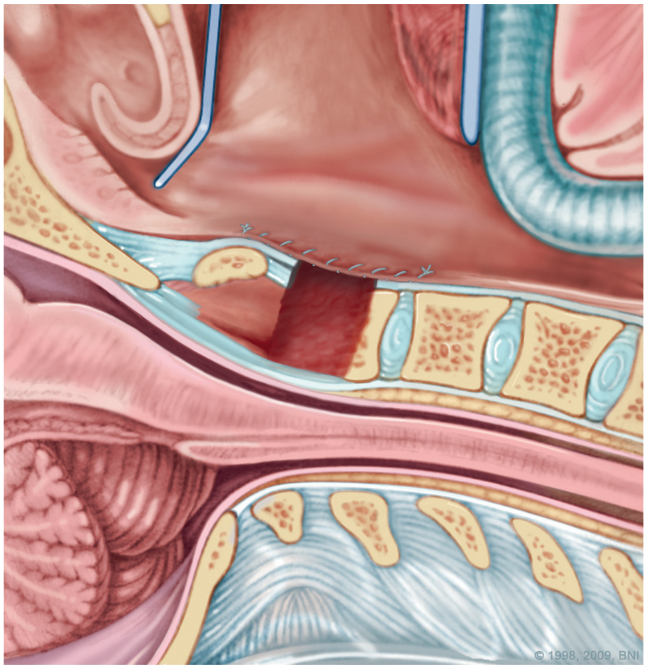

Figure 51-6. A watertight closure of the oropharyngeal incision is important to prevent postoperative cerebrospinal fluid leakage. The brain stem is well decompressed and in anatomic position. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

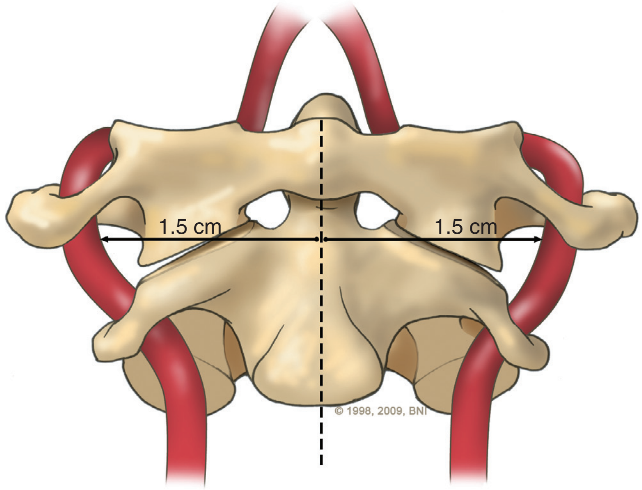

Figure 51-7. Illustration showing the lateral "safe zone" for resection of the odontoid, which is within 1.5 cm of midline. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

must be maintained. Should the dura become lacerated, the surgeon should make every effort to repair it primarily; fibrin glue or a fascial graft should be considered for adjuncts. A lumbar subarachnoid drain should be placed and left for several days. The patient should be under constant surveillance. CSF cultures should be surveyed for signs of meningitis, and preoperative antibiotics should be maintained postoperatively if contamination is suspected. Before extubation, upper airway and tongue swelling should have abated enough to avoid obstruction and the need for reintubation. Should this situation develop, an emergent tracheostomy or

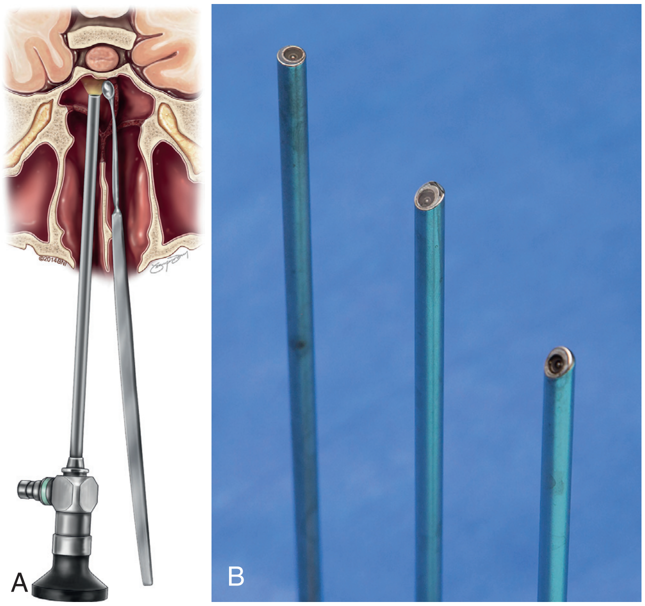

Figure 51-8. A, Endonasal endoscopic setup used to access the clivus and upper cervical spine. **B,** Photograph of the various endoscopes used for visualization of the upper craniocervical junction. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

cricothyrotomy may reestablish the airway. With the endonasal endoscopic approach, care must be taken to minimize injury to the eustachian tube and vidian nerve.

LATERAL APPROACHES

The lateral craniocervical region is rarely involved with bony compressive lesions. Most of these lesions are strongly associated with assimilation of the atlas into the occiput when segmentation fails between the last (fourth) occipital and first cervical sclerotomes. The sclerotomes fail to separate and remain fused, resulting in an incompletely separated atlas and sometimes a bifid lamina. This defect is associated with various syndromes (e.g., achondroplasia and Morquio syndrome) and usually occurs in conjunction with other occipitocervical bony anomalies. Astute clinicians should be wary of entities such as Chiari I malformations and Klippel-Feil syndrome, which may be common. Rarer entities such as basilar invagination and congenital fusion of C2 and C3 also occur.[22,23] Thus, lateral approaches may need to be a part of a larger operative plan and used in conjunction with other techniques to completely address the pathologies.

Lateral compression can be unilateral or bilateral. Stenosis of the foramen magnum leads to pain (usually occipital or high cervical) and possibly myelopathy. Instability may be present, and a fusion procedure may be needed in addition to decompression. Other lesions located ventrally in the foramen magnum, such as vertebrobasilar aneurysms and meningiomas, can be approached laterally. A lateral approach allows adequate exposure while protecting delicate neural structures.

Lateral approaches to the upper cervical spine have evolved for decades, resulting in techniques commonly performed today. In 1973, Henry described a technique for gaining access to the V2 and V3 segments of the vertebral artery through a similar incision.[24] Similarly, Whitesides and Kelly developed lateral approaches for spinal fusion.[25] Their technique was based on dividing the sternocleidomastoid muscle at its origin in the mastoid and reflecting it dorsally. Since then, numerous authors have modified, refined, and expanded this approach to the lateral upper cervical spine and foramen magnum for both extradural and intradural lesions.[26-29] Some have reported more aggressive approaches involving mobilization of the vertebral artery and sectioning of the sigmoid sinus when greater exposure is required.[30] Approaches have been modified to allow resection of lesions traversing the foramen magnum and into the upper cervical spine—the so-called extreme lateral approaches.[31] Either a lateral or a medial incision can be used with the muscle flap directed medially or laterally, respectively. The authors' preferred method is based on a dorsal midline approach.

FAR-LATERAL APPROACH

Surgical Technique

Intraoperative stereotactic navigation can be very helpful with the far-lateral approach. In tumor resection, it can be useful to monitor the lower cranial nerves in addition to SSEPs. The patient is placed in the three-pin head holder with one pin on the forehead in the supraorbital area of the ipsilateral forehead. The two-pin portion is placed low on the contralateral occipital bone, above the level inion, leaving the retroauricular area to the midline open (Fig. 51-9A).

The patient is placed in a modified park-bench position, and the lower arm is slung off the table and cradled with the arm padded and the sling taped to the Mayfield adapter (Fig. 51-9B).[32] The head is flexed and rotated contralaterally to bring the lesion to the highest point in the field. The surgeon should guard against the tendency for the patient's head to translate forward during positioning. Bending the head away from the ipsilateral shoulder opens the angle between the upper cervical spine and the occiput. The upper shoulder is pulled caudally and taped. Tape should not go over the clavicle because torsion on the upper brachial plexus can cause Erb's palsy. The axilla is well padded using beanbags and generous foam padding. The body, especially the upper hip, should be well taped to the bed, permitting full vertical and lateral rotation of the operating table. While the patient is positioned, the evoked potentials are monitored closely for changes.

A satisfactory stereotactic film and registration are critical. Either a curvilinear "hockey stick" or paramedian incision can be used. A paramedian incision is simpler, but stereotactic guidance should be used to avoid injury to the vertebral artery (Fig. 51-10). If used, a curvilinear incision is curved rostromedially below the superior nuchal line. The inferior part of the incision extends caudally to about C4; however, in heavier patients C4 may be difficult to identify. A cuff of muscle and fascia can be left near the superior nuchal line to aid with closure. With subperiosteal dissection, the remaining muscle and fascia are elevated in one flap and retracted laterally. This maneuver exposes the occiput down to the spinous process of C2. Fishhooks or heavy "0" sutures can be used to hold the flap away with a Leyla bar or other retractor system. The C2 ganglion, which exits the lamina of the axis, is preserved throughout its course.

The ring of C1 can be palpated deeper to and above the spinous process of C2. Subperiosteal dissection proceeds laterally onto the lateral mass of C1. The suboccipital triangle is composed of the superior oblique, inferior oblique, and splenius capitis muscle. Deep to the triangle lays the vertebral artery. The lateral point of the triangle is the C1 tubercle. The vertebral artery usually courses along the upper border of the C1 lamina (Fig. 51-11). Familiarity with the particular patient's anatomy, especially that of the vertebral artery and its segments (Fig. 51-12), is important.

During operative exposure, the surrounding venous plexus can cause troublesome bleeding but may be a harbinger for

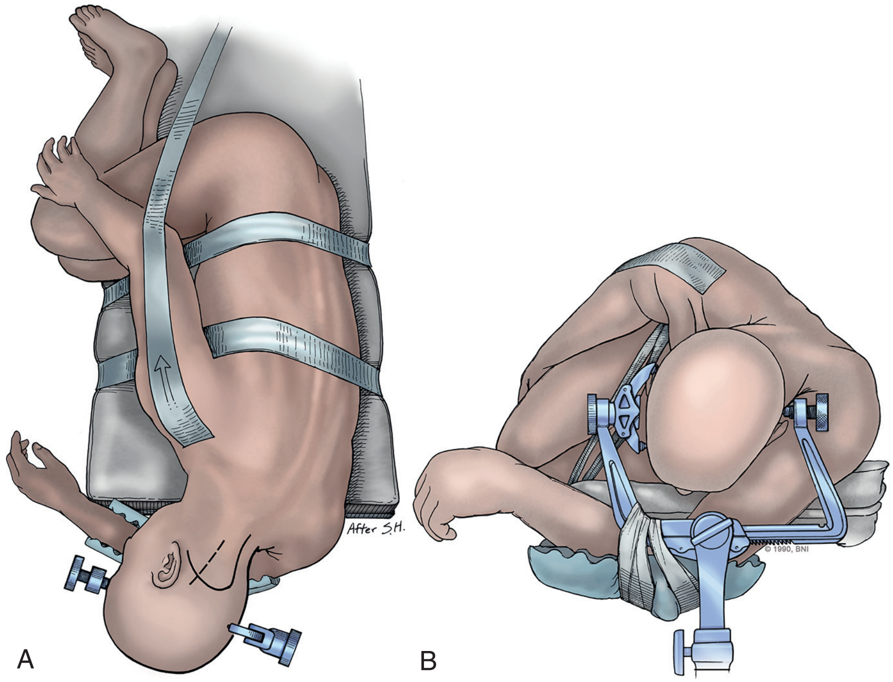

Figure 51-9. Patient position for a far-lateral craniotomy. Both the standard and paramedian linear (dashed line) incisions are shown. Note the position of the lower arm, which is slung in an egg crate from the Mayfield head holder. The upper shoulder is taped on the lateral side to open the working space and to avoid injury to the brachial plexus (**A**). Another view of patient position. The Mayfield pins are placed to maximize exposure. Two pins are placed anteriorly. The third is placed far on the side opposite the operative side. The neck is flexed slightly. Care should be taken to avoid too much traction on the shoulder. The lower arm is slung from the head holder (**B**). *(**A,** Modified and used with permission from Baldwin HZ, Miller CG, van Loveren HR, et al: The far lateral/combined supra- and infratentorial approach: a human cadaveric prosection model for routes of access to the petroclival region and ventral brain stem.* J Neurosurg *81:60–68, 1994. **B,** Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

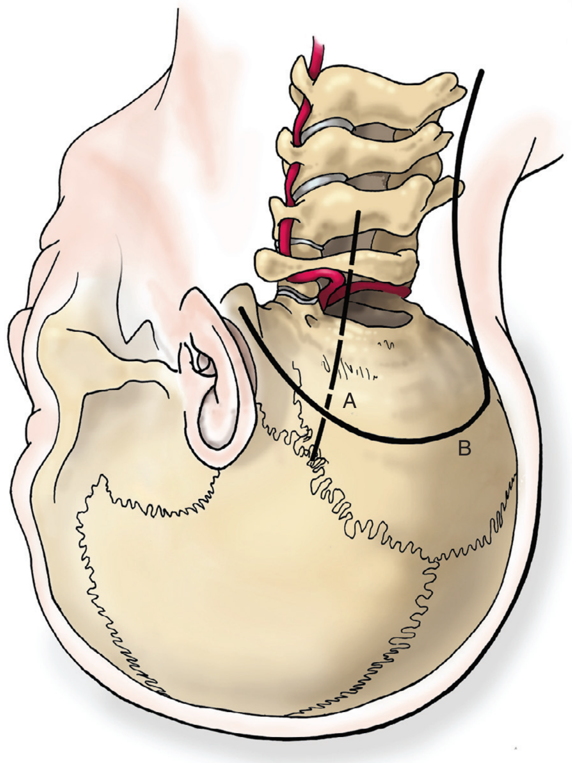

Figure 51-10. Linear incision for a far-lateral craniotomy, which is the simplest method for exposure. Great care should be taken to avoid the vertebral artery, which is near the deep portion of the incision, just inferior to the foramen magnum (**A**). The classic "hockey stick" or "inverted J" incision (**B**). The lateral portion begins at the mastoid tip and is directed medially to the midline. The long arm of the incision is at midline, ideally in the avascular midline (the nuchal line). *(Used with permission and modified from Baldwin HZ, Miller CG, van Loveren HR, et al: The far lateral/combined supra- and infratentorial approach: a human cadaveric prosection model for routes of access to the petroclival region and ventral brain stem*. J Neurosurg *81:60–68, 1994.)*

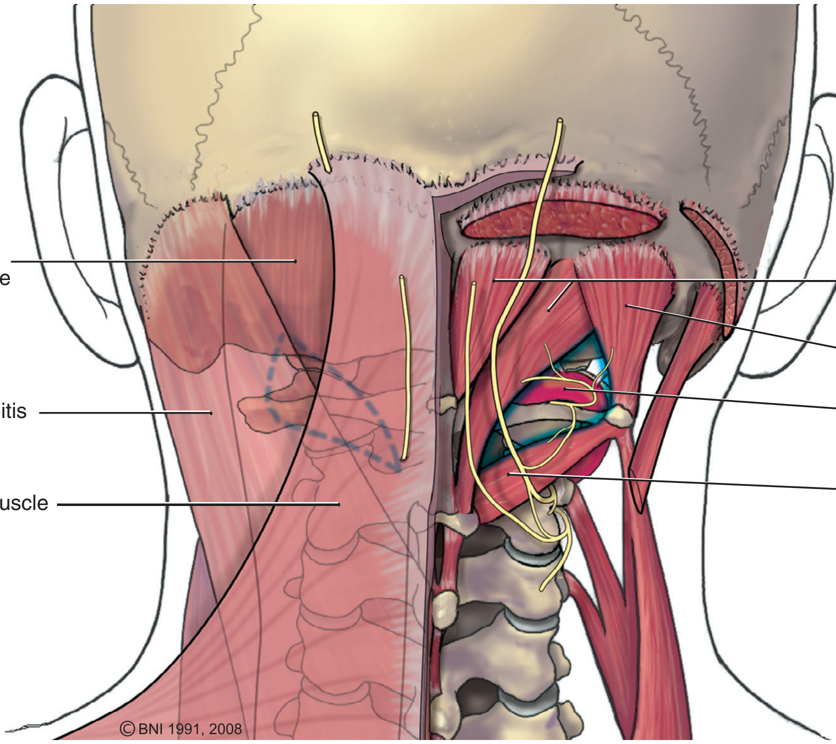

Figure 51-11. Regional anatomy of the suboccipital triangle. The vertebral artery lies in the deep portion of the triangle formed by the superior oblique, inferior oblique, and rectus capitis muscles. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

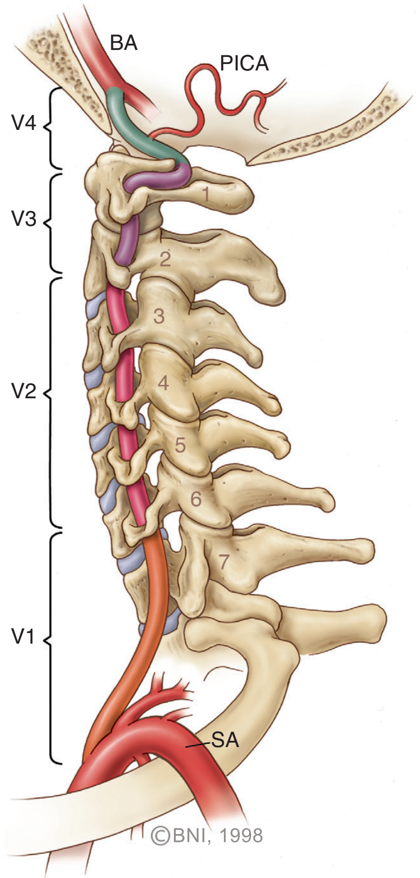

Figure 51-12. Segmental anatomy of the vertebral artery. The four segments are V1, ostial; V2, transverse; V3, suboccipital; and V4, intracranial. BA, basilar artery; PICA, posterior inferior cerebellar artery; SA, subclavian artery. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

the location of the vertebral artery. Bipolar cautery, surgical cellulose, and liquid hemostatic agents usually control any encountered bleeding.

A C1 hemilaminectomy is performed from just beyond midline to the ipsilateral sulcus arteriosus. The artery is unroofed from the foramen and mobilized. A lateral suboccipital craniotomy is performed (Fig. 51-13). The craniotomy is enlarged using rongeurs until the lateral mass of C1 and the occipital condyle are exposed. The vertebral artery is kept moist and protected. A high-speed air drill is used to remove the lesion extradurally. A diamond bur may be used, but care should be taken to avoid heating the bit near the artery. Upbiting curettes are used for lesions that are difficult to reach. If a dural breach is seen, primary closure or a patch graft should be used with fibrin glue. If deemed necessary, an instrumented stabilization is performed. The wound is closed in anatomic layers, and a lumbar drain is placed if the dura is violated.

The occipital condyle can be removed to facilitate further decompression of the foramen magnum (Fig. 51-14). The hypoglossal foramen runs in a posteromedial to anterolateral direction with the occipital condyle (Fig. 51-15). The posterolateral portion of the condyle can be removed to increase access to the anterior portion of the foramen magnum. A wide surgical corridor can be achieved with access to the anterior portion of the foramen magnum (Figs. 51-16 and 51-17). The relation of the occipital condyles can vary greatly from patient to patient. Some patients have anteriorly or posteriorly displaced configurations (Fig. 51-18A-C). Careful and gradual drilling can achieve the desired access to the anterior foramen magnum. If brisk bleeding from the dorsal condylar emissary vein is encountered, it can be controlled with Surgicel Nu-Knit absorbable hemostat (Ethicon US, LLC) or bone. The anatomy of the hypoglossal canal must be understood to avoid injuring cranial nerve XII. The canal runs anterolaterally to posteromedially. The dorsal portion of the condyle can be removed with a high-speed drill. Using

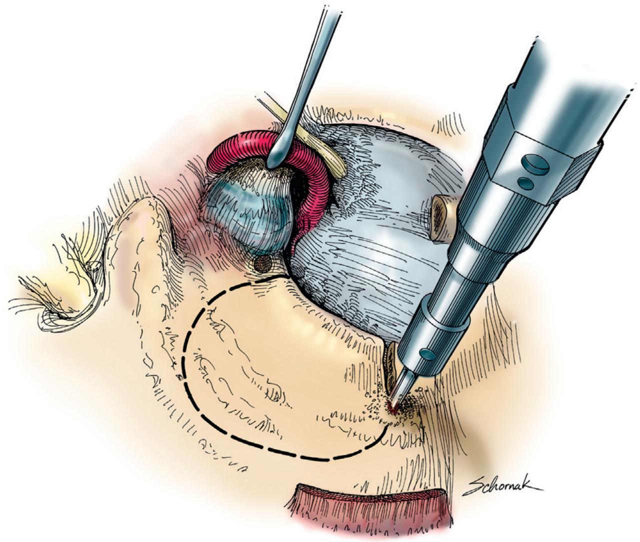

Figure 51-13. **Craniotomy for a far-lateral approach.** The C1 hemilaminectomy is completed first and provides decompression and a useful working space. The footplate of the drill is guided from the midline to the area behind the occipital condyle. The vertebral artery is gently retracted. Its normal position is directly behind the occiput-C1 facet. *(Used with permission and modified from Baldwin HZ, Miller CG, van Loveren HR, et al: The far lateral/combined supra- and infratentorial approach: a human cadaveric prosection model for routes of access to the petroclival region and ventral brain stem.* J Neurosurg *81:60–68, 1994.)*

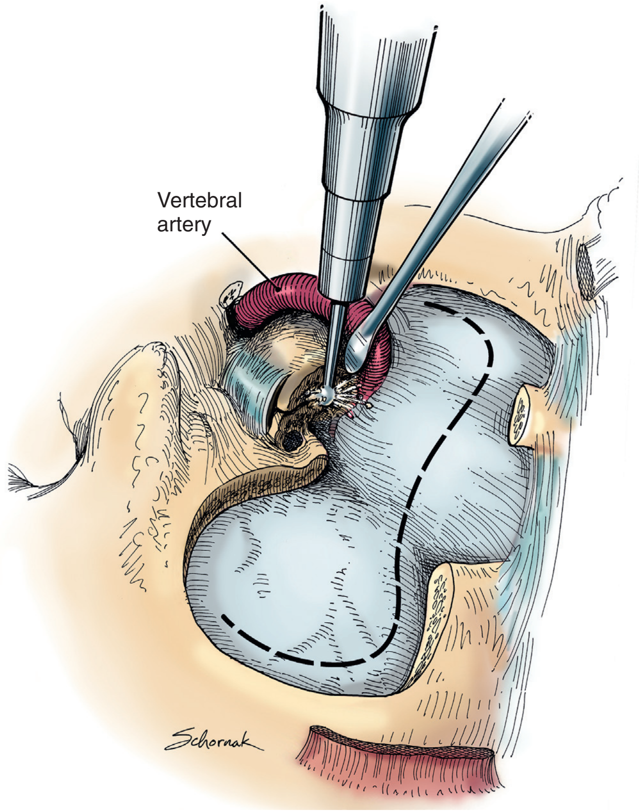

Figure 51-14. Condylar drilling. A powered drill is used to gradually remove the posterolateral portion of the occipital condyle to maximize access to the anterior foramen magnum. *(Used with permission and modified from Baldwin HZ, Miller CG, van Loveren HR, et al: The far lateral/combined supra- and infratentorial approach: a human cadaveric prosection model for routes of access to the petroclival region and ventral brain stem.* J Neurosurg *81:60–68, 1994.)*

navigation at this depth is difficult. In select cases, occipitocervical stabilization should be considered. Biomechanical stability is altered when more than 50% of the condyle is resected.[33] Stability is also significantly reduced if the ventral portion of the condyle is removed. Options for fusion include occipitocervical plating with lateral mass screws, Steinmann pin fixation, and sublaminar wires.[34,35]

LATERAL RETROPHARYNGEAL APPROACH

Surgical Technique

Lateral retropharyngeal approaches have been used as alternatives to transoral ventral approaches to the clivus and upper spine.[36-38] The patient is placed supine upon the table, and the head is extended and rotated 30 to 45 degrees away from the side of the lesion. A parallel incision is prepared 4 cm inferior to the mandible, and the marginal mandibular nerve is protected. The platysma muscle is identified, divided, and elevated superiorly. Next, the submandibular gland is elevated rostrally to expose the digastric muscle, facial artery, and facial vein. The digastric muscle is mobilized by detaching the tendons and elevating the muscle superiorly; this exposes the hypoglossal nerve (Fig. 51-19A). The hypoglossal nerve is then retracted rostrally, and the pharyngeal muscles are medially mobilized to expose the anterior tubercle of C1 (Fig. 51-19B and C). Image guidance can be used to identify the lateral mass of C1 and C2 as needed. A high-speed drill can be used to perform bony removal.

Pitfalls and Complication Avoidance

Patients usually present with cervical pain and various degrees of neurologic compromise. Potential complications are closely

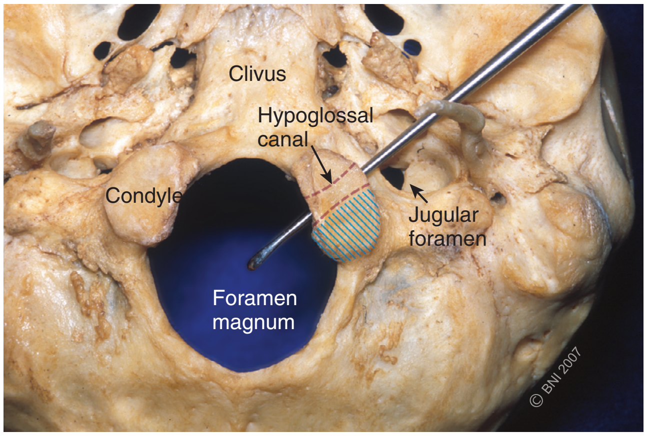

Figure 51-15. The hypoglossal canal and occipital condyle. The posterolateral portion of the condyle can be drilled to increase anterior exposure. The metal probe indicates the location of the hypoglossal canal within the condyle. *(Acknowledgment: Anatomic dissection and dissection photo by Mauro Ferreira, MD. Figure used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

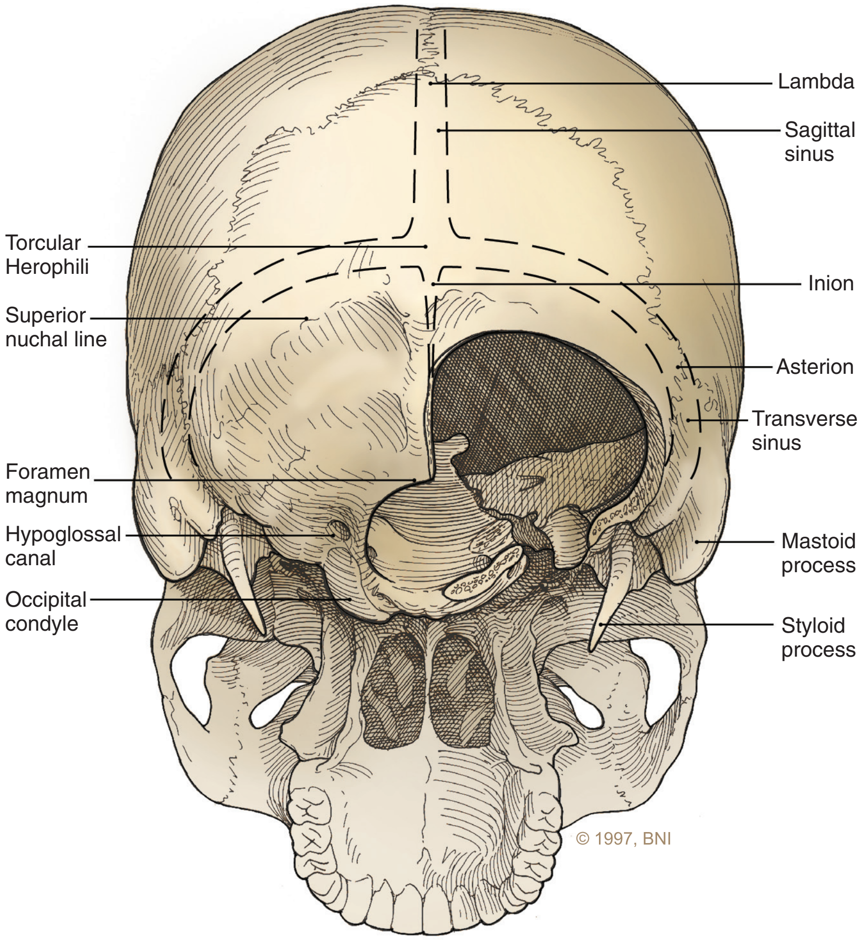

Figure 51-16. Detailed bony anatomy of the access provided by a far-lateral craniotomy. Much of the anterior foramen magnum can be accessed through this corridor. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

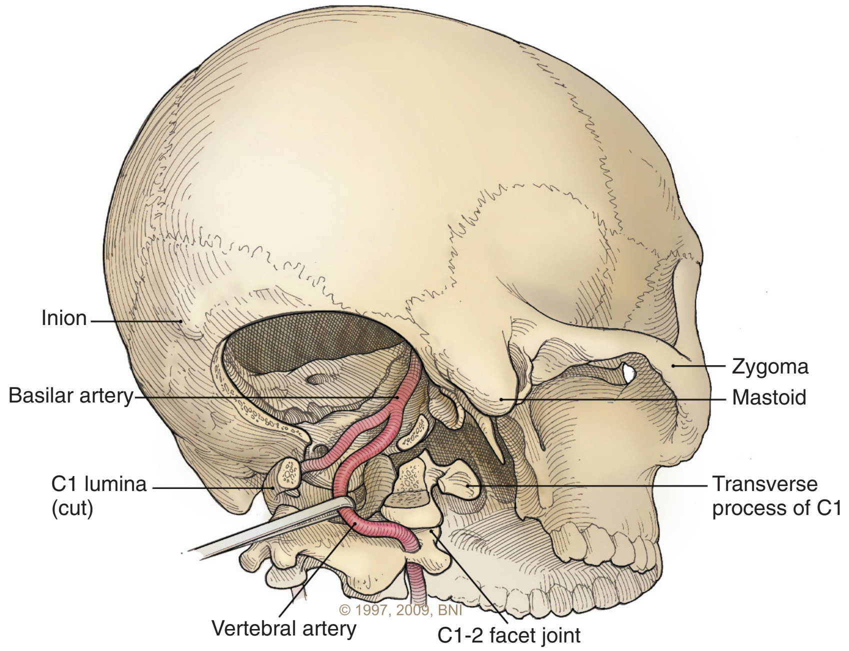

Figure 51-17. Oblique view of anterolateral access provided by a far-lateral craniotomy. The vertebrobasilar system is included to show the relevant regional arterial anatomy. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

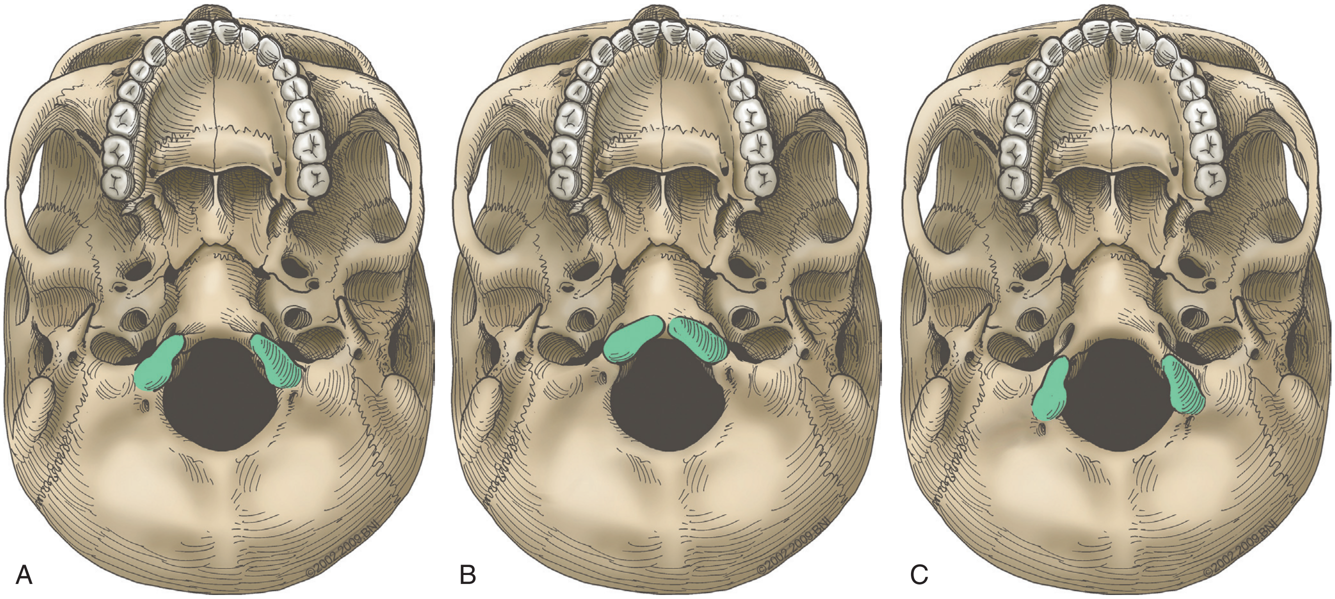

Figure 51-18. Normal configuration of the occipital condyles *(green)* at the 10 o'clock and 2 o'clock positions **(A)**. Anterior **(B)** and posterior **(C)** variants of the occipital condyles *(green)*. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

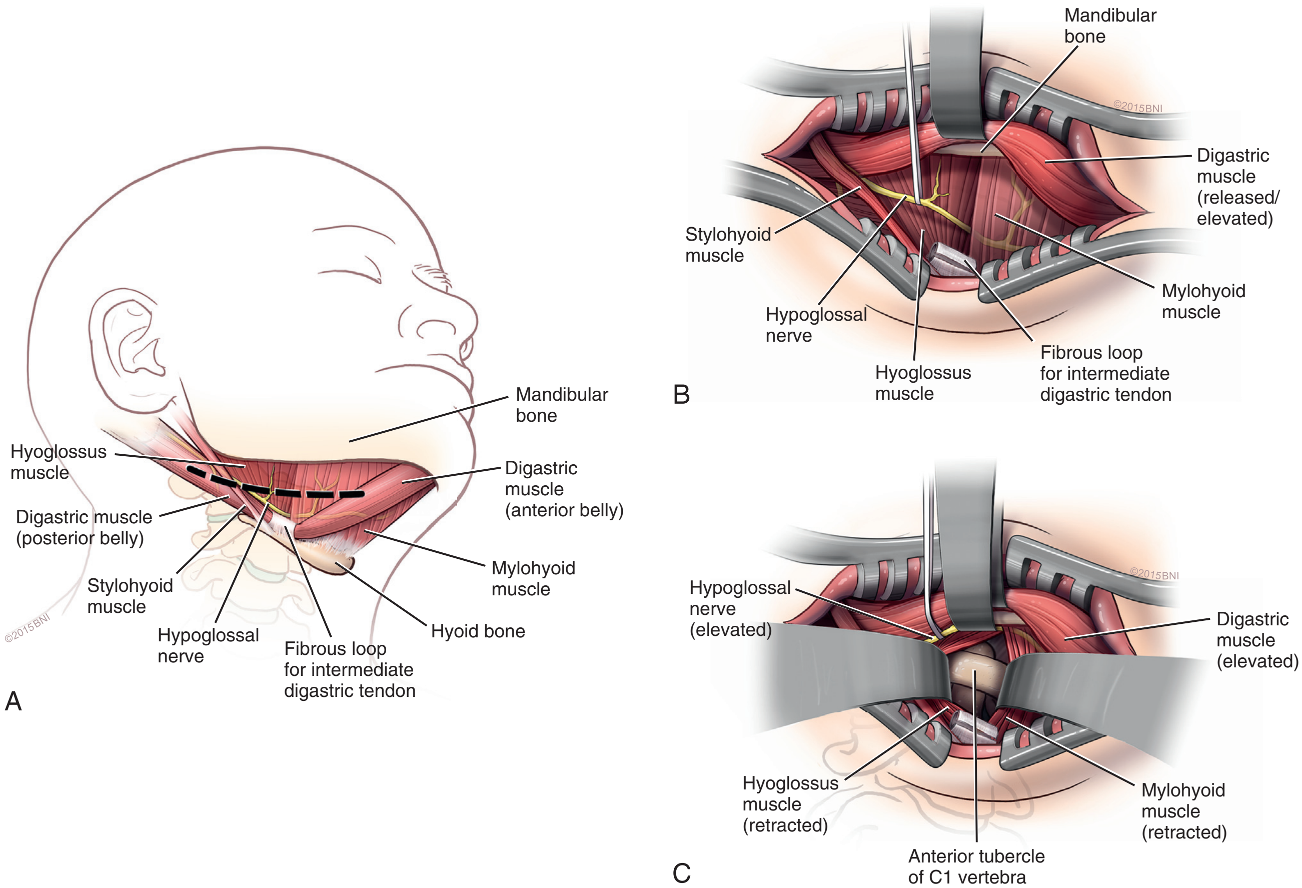

Figure 51-19. Diagram illustrating the steps of the retropharyngeal approach to the upper cervical spine. **A,** Skin incision (*dashed line*) and underlying structures. **B,** Once the skin incision is made, the digastric muscle is mobilized and retracted to expose the hypoglossal nerve. The hypoglossal nerve is retracted with vessel loops. **C,** Next, the hypoglossus and mylohyoid muscles are retracted to expose the C1 tubercle. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

associated with the surgical angle of approach. The vertebral artery is at risk from surgical manipulation. Arterial lacerations should be repaired primarily. If a rent is too large, an end-to-end anastomosis should be performed with nonabsorbable monofilament suture (usually 6-0 to 10-0). Although most patients may tolerate occlusion of the vertebral artery, the procedure should be avoided. Other possible vascular complications include thromboembolic stroke or ischemia from an arterial dissection. If available, endovascular therapies may be considered. Postoperatively, CT angiography or vessel-specific angiography should be completed expeditiously if vascular injury occurs.

Other complications reported with this technique and its variants include venous air embolism, lower cranial nerve palsies, CSF leakage, hydrocephalus, brain stem edema, and death.[28,30,31] Complications are most common with ventral and lateral tumors that require some brain retraction and cranial nerve dissection. If hydrocephalus is present preoperatively, it may be prudent to place an external ventricular or lumbar drain before surgery. Theoretically, doing so may reduce the need for retraction and lessen the likelihood of a CSF fistula.

DORSAL APPROACHES

Early on, nearly all decompressions for the CCJ were performed dorsally. Posterior fossa craniectomy with upper cervical laminectomy for the treatment of ventral pathology was soon found to be inadequate, and, in some instances, the patient's neurologic status worsened. With the newer, more direct approaches, the surgical strategy now focuses on the location of the pathology and local anatomy. Dorsal decompression still holds a prominent place in the surgical methods of contemporary neurosurgeons.

Described by Hans Chiari, Chiari I malformations result from descent of the cerebellar tonsils below the level of the foramen magnum (Fig. 51-20AB).[39] Others have also been involved in the elucidation of this neurologic abnormality.[40,41] Patients usually suffer from headaches but may also exhibit cervical myelopathy and hydrocephalus. Myelopathy may result from direct compression from tonsillar herniation or syringomyelia, whereas hydrocephalus reflects alterations in CSF flow patterns. Both conditions usually resolve after posterior fossa decompression and duraplasty.[42] Syringomyelia most often resolves 3 to 6 months after surgery.[43] Type II Chiari malformations are often associated with spina bifida and may necessitate decompression if symptoms occur (Fig. 51-20C).

Achondroplasia is the most common form of dwarfism and is often associated with craniocervical deformities.[44] Most cases are sporadic. The foramen magnum becomes narrowed due to craniovertebral abnormalities related to thickening of the occipital bone. A thick epidural band can accompany the bony changes.[45] Spinal stenosis leads to spinal cord compression and hydrocephalus.[45] The major threat is respiratory compromise and the risk of sudden death.[46] Newborns with achondroplasia may benefit from early neurologic and respiratory screening, MR imaging of the CCJ, and early decompression for severe cases.[46,47] Rheumatologic and degenerative diseases can also cause chronic subluxation and basilar settling. Irreducible cervicomedullary compression often results. Dorsal decompression and C1 laminectomy with dorsal stabilization are often used to correct the deformity, to decompress neurologic structures, and to halt progression.

SUBOCCIPITAL CRANIECTOMY

Surgical Technique

Monitoring of SSEPs is a useful adjunct. Baseline readings should be obtained before the patient is positioned. Lower cranial nerve monitoring is not routinely necessary. Preoperative halo traction or Gardner-Wells tongs may be placed to reduce the deformity or to maintain stability. Often patients undergo further reduction when under general anesthesia. Before the incision is made in patients in traction, a lateral radiograph should be obtained to assure optimal alignment.

The patient is turned prone onto chest rolls, and the arms are tucked at the sides. The chin is tucked forward against the neck in a military tuck position (Fig. 51-21). The planned craniectomy is bordered on all sides by the venous sinuses. Overaggressive lateral decompression should be avoided to prevent cerebellar sagging (Fig. 51-22). A midline incision is

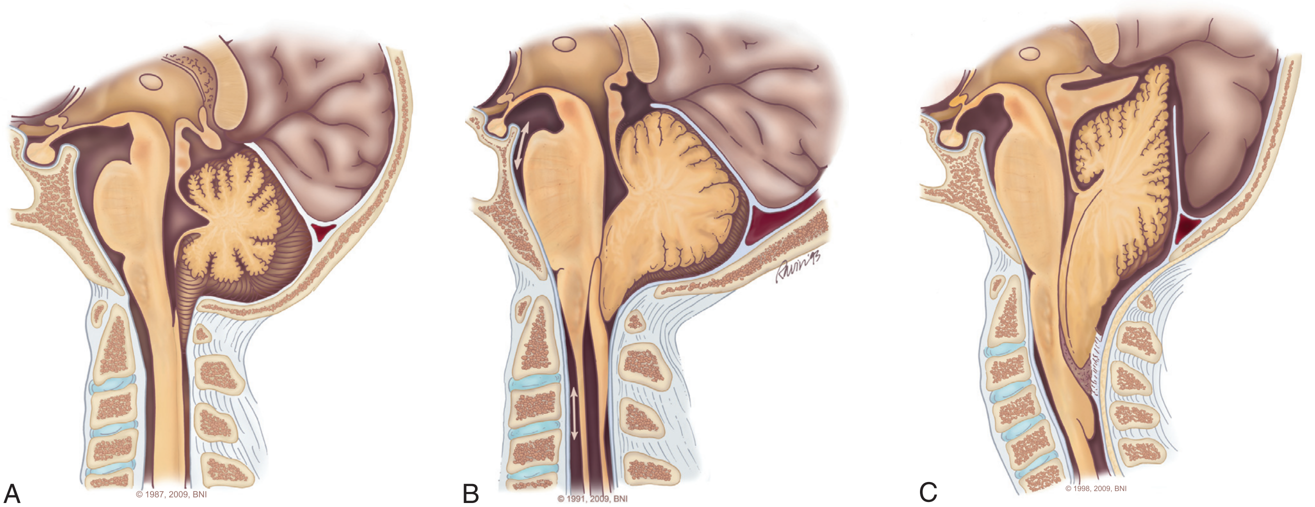

Figure 51-20. Type I Chiari malformation with cerebellar tonsillar herniation only **(A)**. Type I Chiari with an associated syrinx **(B)**. The bidirectional arrows indicate an altered pattern of cerebrospinal fluid flow. Type II Chiari malformation associated with various types of spina bifida **(C)**. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

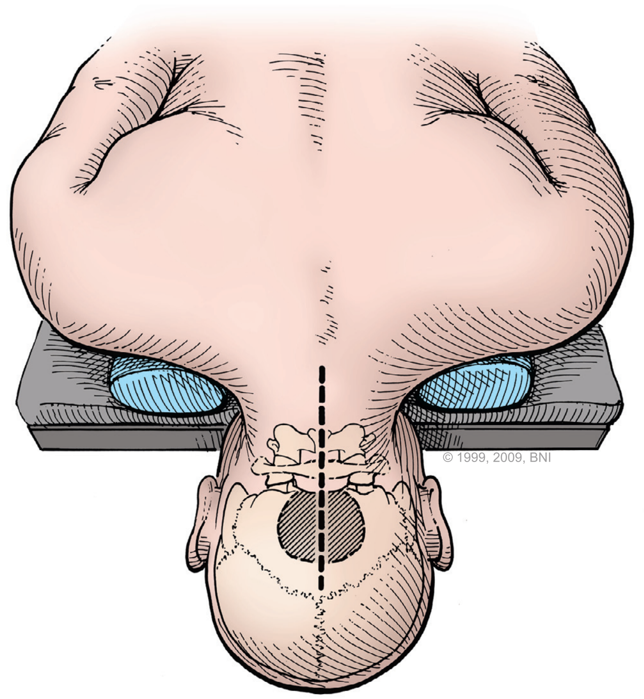

Figure 51-21. Patient position for suboccipital craniectomy. The patient is placed prone on soft gel rolls with the chin in a "military tuck." The incision is made from just above the inion to the spinous process of C2 in the midline. The proposed craniectomy (cross-hatched) incorporates the posterior foramen magnum. Cervical laminectomies of C1 and C2 may be necessary to achieve adequate decompression. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

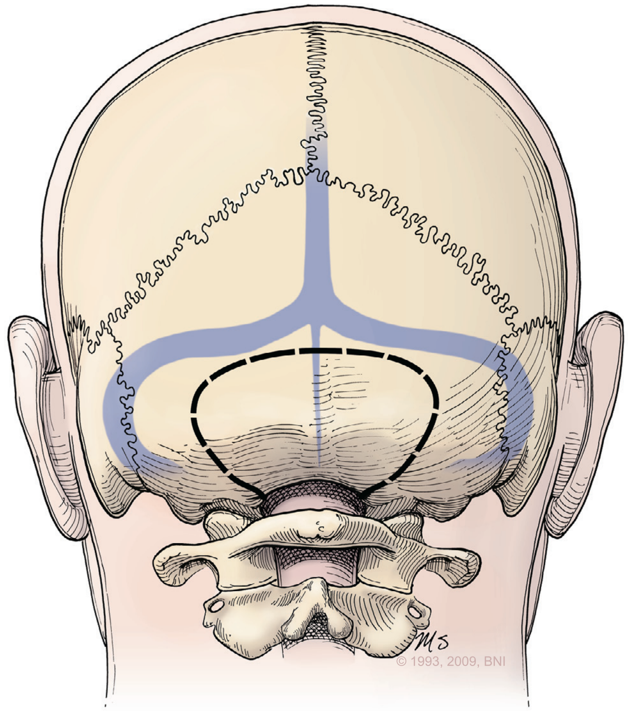

Figure 51-22. Suboccipital craniectomy. The venous sinuses border the proposed craniectomy. Care should be taken to avoid overzealous bone removal. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

planned from the inion to C4 using the nuchal line as the avascular midline plane. Dissection proceeds in this plane to the occiput and spinous process of C2. The muscle attachments of C2 are left intact, but the top of C2 should be exposed (Fig. 51-23). The region around C1 and the foramen magnum is often the deepest part of the exposure. The occiput is exposed laterally and inferiorly in a subperiosteal fashion. Near the foramen magnum, curettes are used to expose the foramen magnum carefully. A sharp ventral curve of the occipital bone indicates the margin of the foramen magnum. The C1 lamina is identified. A bifid C1 is often associated with a Chiari I malformation, and the lamina should be cautiously identified. On both sides of midline, the exposure is continued laterally onto the ring of C1 using periosteal elevators and curettes. Laterally, the paravertebral venous complex can cause brisk bleeding. Such bleeding can be avoided by the use of bipolar forceps as a periosteal dissector on the lateral portion of the atlas and by the use of Metzenbaum scissors to cut the coagulated muscle. Hemostatic agents can be helpful adjuncts. Small curved curettes are used to palpate under the ring of C1 and foramen magnum.

The bone of the occiput is removed with a high-speed drill. Two bur holes can be placed on either side of the midline "keel" that is sometimes present in the posterior fossa. The holes can be used both as depth gauges for drilling the bone and as access to strip the dura from a particularly deep keel with Penfield dissectors. The degree of bony removal depends on the anatomy of the pathology. In some cases, the foramen magnum only needs to be opened minimally. In the case of a Chiari malformation, a generous dorsolateral decompression allows adequate room for the cerebellum. At this time, a C1 laminectomy should be performed before the dura is opened.

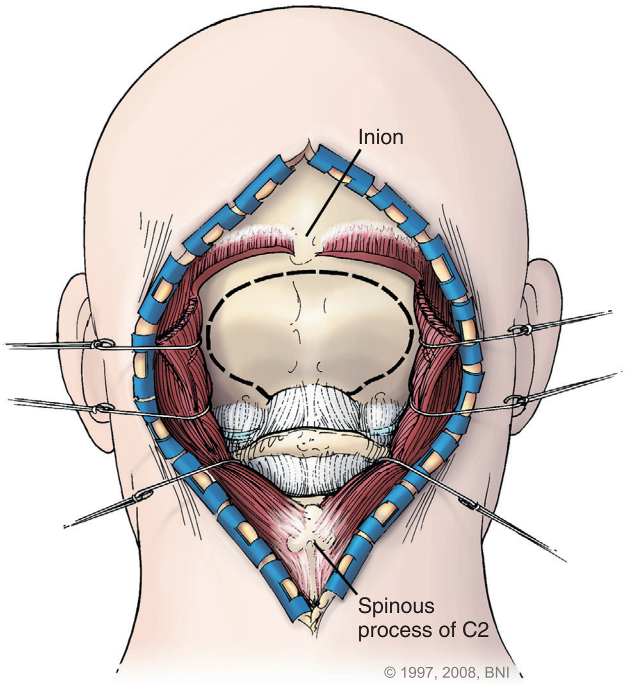

Figure 51-23. Midline incision for decompression. The avascular midline plane is followed and exposure extends to C2. Bone is removed with a drill, Kerrison punches, or both. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

If instrumentation is anticipated, it can be placed before duraplasty.

A thick fibrous band, which can be quite vascular, is often present at the foramen magnum, especially in achondroplastic patients. If a duraplasty is planned, the dura is opened in a Y-shaped fashion. This configuration allows hemostatic clipping of the occipital sinus in the posterior fossa midline. Clips are placed sequentially along the dural incision. Arachnoid adhesions obstructing the foramen of Magendie should be lysed. The authors do not use bipolar cautery near the cerebellar tonsils. The practice of plugging the obex has also fallen from favor. A chevron-shaped patch is grafted. Historically, autologous fascia lata and skull periosteum are the best-tolerated grafts. Bovine pericardium, cadaveric dura, or paravertebral fascia are all reasonable substitutes.

A watertight closure is critical, and special attention should be paid to the inferior corner when the graft is sewn in place (Fig. 51-24). At this stage, CSF leaks can occur from a small defect in dural closure. More critical is the cervical fascial closure. The junction of the cervical fascia and galea at the inion should be closed meticulously. Here, most CSF leaks occur due to inadequate closure, especially in the corners of the durotomy. If allograft is used, some advocate a short course of low-dose steroids to reduce the rate of postoperative chemical meningitis.

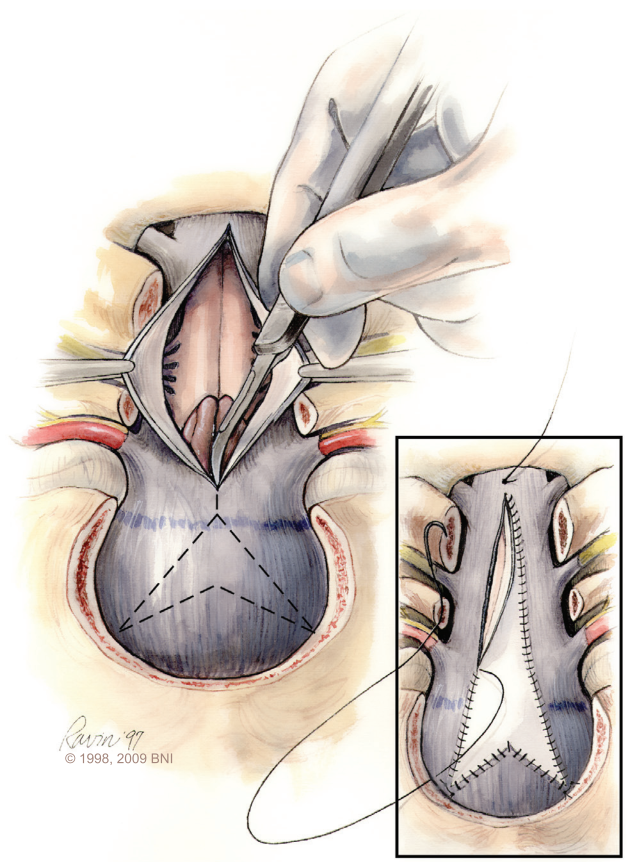

Figure 51-24. A midline dural incision is used, and a patch graft of autologous fascia lata or other suitable substitute is sewn in place. Watertight closure is critical to prevent postoperative complications. *(Used with permission from Barrow Neurological Institute, Phoenix, AZ.)*

Pitfalls and Complication Avoidance

The most commonly reported complications associated with dorsal approaches are CSF leaks, hydrocephalus, and pseudomeningocele. Dural closure is therefore paramount to prevent complications. There have been no prospective, randomized studies on preoperative placement of a ventriculostomy, and only class III data are available. Many have reported their experiences with achondroplasia. Aryanpur and colleagues[48] reported 4 documented CSF leaks in 15 cases; 1 patient developed meningitis. Three of their patients required postoperative shunting. Other studies on this entity have reported low rates of CSF leakage.[42,45,46,49]

Neurologic outcomes are also limited to class III data. Lower cranial nerve dysfunction, respiratory difficulties, weakness, sensory changes, and chemical meningitis are all possible. Poor preoperative neurologic status tempers hopes for a good outcome. The diligent clinician should ensure respiratory and alimentary function in the postoperative period and during the patient's convalescence. Often, a swallowing evaluation is necessary both pre- and postoperatively to assess the patient's recovery. Increased weakness and sensory changes should prompt the surgeon to obtain postoperative imaging or, if severe enough, should warrant immediate reoperation. Patients with delayed signs should be screened for iatrogenic instability with dynamic films. Chemical meningitis, which is usually a diagnosis of exclusion, should be entertained if a spinal fluid assay is negative for infection. Usually, low-dose dexamethasone taken orally brings relief to the patient.

SUMMARY

Advances in instrumentation, imaging, and intraoperative monitoring have opened new horizons and afforded increasingly effective and safe methods of decompression and stabilization of this vital region. A clinical presentation well correlated with a neurologic examination and radiographic findings is the basis for surgical evaluation. Choosing the appropriate approach, effectively executing the surgery, and having an astute knowledge of the regional anatomy and pathology are all critical factors for achieving satisfactory patient outcomes.

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