Chapter 27
Surgery at the Crossroads: Craniocervical Neoplasms

Arnold H. Menezes, M.D., Vincent C. Traynelis, M.D., Albert J. Fenoy, M.D., Bruce J. Gantz, M.D., Stephen F. Kralik, M.D., and Kathleen A. Donovan, A.R.N.P.

The craniovertebral junction is a funnel comprised of the clivus and foramen magnum and the upper two cervical vertebrae that acts as a biomechanical and anatomic unit. Neoplasms that arise within the craniovertebral junction comprise osseous tumors, extensions from the soft tissue that surround the region and neoplasms originating from nervous system structures contained within.

Tumors of the craniovertebral junction present with a wide constellation of neurological signs and symptoms, and false localizing signs are numerous. Precipitous decline in function can occur with malignant growth, and the potential for rapid progression to irreversible myelopathy demands early recognition and treatment (29, 38, 47, 56, 65). There is no single symptom or neurological finding pathognomonic for a lesion in this location. Symptoms may arise only after significant size has been achieved because of the generous subarachnoid spaces of the cervicomedullary junction. Anatomic complexities of the region and the decussation of the sensory and motor tracts can lead to erroneous diagnosis and fluctuating neurological symptomatology. Elsberg and Strauss (23) were the first to report on a systematic evaluation of foramen magnum tumors. Since then, several authors have reported on extra-axial lesions affecting the region, such as meningioma and neurinomas, and osseous neoplasms, such as chordoma, chondrosarcoma, plasmacytoma, osteoblastoma, fibrous dysplasia, giant cell tumors, and metastatic tumors. Our experience is summarized in this brief presentation. A retrospective analysis of the medical records and radiographs of patients from the University of Iowa Hospitals and Clinics Neurosurgery database and tumor registry was performed. Eight hundred eight patients were evaluated by the senior author (AHM) at the University of Iowa Hospitals and Clinics between the years 1977 and 2003. Three hundred eighty-two patients presented with osseous tumors, and 426 had neural tumors. Of these, 179 patients underwent operative management by the Neurosurgery Service at the University of Iowa Hospitals and Clinics, and inclusion criterion was histopathology consistent with extramedullary origin in the region of the craniovertebral junction. One hundred seventy-nine patients ultimately underwent operative management with the final pathology as follows: chordoma = 49; meningioma = 41; schwannoma = 5; plasmacytoma = 4; osteoblastoma = 3; neurenteric cyst = 5; calcium pyrophosphate masses = 36; and miscellaneous = 36. The miscellaneous category included fibrous dysplasia, multiple myeloma, dermoid masses, metastatic tumor, Ewing's sarcoma, and aneursymal bone cyst.

The location of these tumors was predominantly ventral. One hundred forty patients of 179 had a ventral location, 25 of 179 had a lateral location, and 14 of 179 had a dorsal location. Most of the tumors of ventral location were meningioma and chordoma. These were more frequently located at the foramen magnum with straddle lesions. Purely foramen magnum location was 50 of 179 in the series. A straddle lesion between foramen magnum and the upper most cervical canal was present in 94 of 179 patients. Thirty-eight patients had a lesion corresponding to just below foramen magnum and C1-2 level. The age at presentation spanned 1 to 86 years with an average of 45.7 years. The average time to diagnosis was 13 months.

COMMON CLINICAL MANIFESTATIONS OF CRANIOVERTEBRAL JUNCTION TUMORS

The clinical presentation of craniovertebral junction tumors was divided into those with predominantly intracranial origin and a craniospinal location, with the straddle lesions mainly at foramen magnum and those affecting the high cervical spinal cord (45, 47).

Patients with intracranial lesions presented with involvement of lower cranial nerves, brain stem dysfunction, and occasionally cerebellar symptoms (46). Patients with straddle lesions had lower cranial nerve dysfunction and predominance of high cervical myelopathy. High cervical lesions presented with myelopathy and occasionally with involvement of the spinal accessory nerves and the descending tracts of the trigeminal nerve.

Pain was a frequent presentation and occurred in 70% of patients. This was attributed to the second cervical dermatome. The head was found to be flexed and at times resembled torticollis, especially in the younger individuals. Pain was described as an aching sensation aggravated by neck and head motion and attributed to the suboccipital region.

Paresthesias and dysesthesias of the face, hands, and limbs occurred in 40% of individuals. A significant percentage had an abnormal cold sensation of the lower extremities, as had been described by Elsberg and Strauss (23) and, subsequently, Beatty (10) as being pathognomonic of lesions of high cervical cord. Pain and temperature sensation was affected followed by loss of joint sensation. A suspended sensory loss with patches of preservation confused the diagnosis in 18% of patients.

Spastic weakness of the extremities was a prominent feature, and a clockwise paresis was seen in 22%. This distinctive progression was previously described (43). Wasting of the intrinsic hand muscles was seen in 8% of the extradural lesions (44, 61). A mixture of upper motor neuron findings in the upper and lower extremities reflected the pyramidal decussation being affected just below the obex of the fourth ventricle. This was explained by the fact that the medial fibers of the pyramidal tract carry impulses to the upper extremities and cross superior to the lateral fibers that serve the lower extremities. A similar situation occurs with the sensory decussation of the medial lemniscus, which could produce varied patterns of sensory abnormality (38). The syndrome of cruciate paralysis was seen in four individuals with ventrally located foramen magnum meningioma (12).

Cranial nerve palsies were seen in 33% and 28% of individuals showing dysphagia or dysarthria. The most common cranial nerves affected were the vagus, hypoglossal, and glossopharyngeal nerves. This involvement led to dysphagia, slurred speech, repeated aspiration pneumonia, and weight loss. Seventeen percent of individuals experienced hearing loss and complained of tinnitus and vertigo. This was common with the meningiomas and chordoma.

NEURODIAGNOSTIC IMAGING

Imaging modalities have changed the ability to produce a rapid diagnosis as compared with the early part of the series when pneumoencephalography, angiography, and myelography were the studies performed. Presently, complimentary multimodality imaging includes magnetic resonance imaging, magnetic resonance angiography, computed tomography, and three-dimensional computed tomography/angiography. In our institution, cerebral angiography is used to understand the dynamics of collateral circulation and tumor vascularity (46). Temporary balloon occlusion is a means of assessing a patient's tolerance of vascular occlusion of either the carotid or the

vertebral circulation before surgery. Tumor embolization is performed routinely in osseous tumors except for chordoma. Tumor encasement is better seen with conventional angiography than with magnetic resonance angiography.

The stability of the craniovertebral junction was assessed with dynamic magnetic resonance imaging, computed tomography scans, and flexion/extension lateral radiographs of the craniocervical region.

SURGICAL APPROACHES AND DECISION MAKING

Our philosophy of treating craniocervical neoplasms was predicated by whether the lesion was considered to be benign or malignant. In benign lesions, a space is created among the neurovascular structures, thereby allowing surgical debulking and resection "from within."(45) Malignant disease required a radical resection with clean margins if possible. In most instances, benign lesions such as chordomas are radio-resistant, and hence, a gross total resection was the aim. Craniovertebral stability, both before and after operative intervention, was considered. The factors that influenced the specific treatment of neoplasms at the craniocervical junction were 1) whether the lesion was benign or malignant, 2) the direction of encroachment, 3) whether the lesion was associated with vascular or intramedullary component such as syringomyelia, 4) craniovertebral stability, and 5) the patient's age.

In children, the potential for growth and concern about stability and the patient's size was thought to be critical. Tumors at the craniovertebral junction affect the pediatric population to a lesser extent than the adult (2, 30, 34, 35, 46). In a child, a transpalatal approach to the clivus and the sella is considered before a sublabial transsphenoidal or maxillary dropdown procedure with the aim of avoiding damage to the growth centers in the nasal septum and the pterygoid plates (46).

The advances in microsurgical instrumentation have permitted the development of extensile surgical approaches based on an understanding of the complex anatomy, craniovertebral dynamics, and the site of encroachment. As a result of this development, the entire circumference of foramen magnum is within the neurosurgeon's reach (Fig. 27.1). Surgical limitations and considerations must be appreciated when one is designing an approach to the craniocervical junction. Table 27.1 describes the surgical approaches and the extent of the exposure obtained with each procedure. The pathology that is often encountered for each procedure is likewise listed in Table 27.1, along with the advantages, the limitations, the risks, and long-term side effects. A combination of procedures is possible, such as midline mandibulotomy and glossotomy, to aid with the transoral-transpalatal procedure (19, 33, 37, 44, 50). Similarly, a posterolateral procedure can be combined with a midline posterior fossa exposure or a presigmoid transpetrosal one (3, 22, 32, 43, 52, 57–59). As a general rule, midline extra-axial tumors are best approached in an extra-axial fashion without retracting the brain or violating the dura. A lateral avenue should be used for tumors that are situated laterally (16, 41, 44). The transsphenoethmoidal approach is well tolerated as a route to the upper two thirds of the clivus (43). A better exposure is obtained with access to the contralateral side, and hence, a bilateral approach may be required.

A gross total resection occurred in 83% of the patients and a partial resection in 17%. Surgical approaches were dictated by the manner of encroachment: a transpalatopharyngeal approach was the primary procedure in 48% of patients, a posterolateral-far lateral approach in 38%, dorsal procedure in 28%, and the remainder was performed with the other techniques described. Twenty-five percent of patients required craniocervical stabilization.

Complications occurred in 6% of patients, and a recurrence was seen in 13%. Our average follow-up was 43 months and survival 93%.

SPECIFIC TUMORS OF THE CRANIOCERVICAL JUNCTION

Clival and Craniovertebral Junction Chordomas

Chordomas are locally destructive rare tumors that are of presumed notochordal origin. They arise along the vertebral axis with a propensity for the spheno-occipital region and the sacrum (7). Surgery should be attempted for radical removal because incompletely resected tumors recur uniformly and more extensive tumor removal is associated with a longer patient survival (3, 45, 53, 60). The proximity of clival and craniovertebral chordomas to the brain stem, cervicomedullary junction, and the high cervical spinal cord, as well as the cranial nerves, makes complete resection difficult. In addition, aggressive surgical treatment has shown frequent morbidity (3,28). Advances in neuroimaging, neurosurgical instrumentation, and cranial based techniques have permitted extensive resection and are better tolerated.

Proton beam therapy has emerged as a valuable adjunct to surgery. Forsyth et al. (26), in 1993, reported an overall survival of 51% at 5 years and 35% at 10 years among 39 patients with clival chordomas treated with surgery and conventional photon radiation (4). The results from the Harvard cyclotron and Berkley's Lawrence Livermore Laboratory with proton radiation of clival chordomas are impressive (8, 9, 13, 18). The actual control rates are 82% at 5 years and 58% at 10 years. Relapses were mainly local, and metastasis occurred in approximately 20% of patients. They felt that tumor volume of more than 75 mL, tumors that were at least 10% necrotic, and tumors with involvement of the cervical spine carried a poor prognosis. There was no histological correlation regarding prognosis. These data strongly suggested that surgery, when combined with radiation therapy, particularly proton beam therapy, offered substantial improvement over the natural history of chordoma.

Forty-nine patients with chordomas of the clivus and upper cervical spine have been treated since 1985 at the University of Iowa Hospitals and Clinics. There were 27 men and 22 women. Fourteen had undergone previous surgery, and 11 had received radiation therapy, 5 of whom were treated with proton beam therapy. These 49 patients underwent 56 skull base surgical procedures. Ten of the 49 patients required a separate operation for stabilization (Fig. 27.2). Nineteen patients underwent a transoral-transpalatopharyngeal approach, 8 underwent the transmaxillary approach, and 6 underwent the transsphenoethmoidal approach. The infratemporal fossa approach was used in four patients, the lateral extrapharyngeal approach in four patients, the far lateral transcondylar approach in nine patients, and the transfacial-transbasal in six patients. Fifty-seven approaches were made (Fig. 27.3). A gross total resection was accomplished in 14 patients, a subtotal resection (removal of more than 90% of tumor documented on a postoperative magnetic resonance image) was accomplished in 16 patients, and a partial resection was accomplished in 19 patients.

Within the 19-year follow-up range, seven patients had died. Five deaths occurred in patients who had undergone transpalatopharyngeal approaches. Three of these occurred in 2 years of operation at our institution, and each of these had previously been subjected to at least four operative procedures for tumor resection elsewhere and proton beam therapy. The interval between proton beam radiation and consultation at our institution with recurrent tumor was less than 3 years in each case. In a fourth patient, a transoral biopsy had been followed by 66 Gray conventional

radiation therapies. Progressive tumor enlargement during radiation prompted referral to our institution. The tumor was completely resected with a transpalatopharyngeal route and followed by a dorsal occipitocervical fixation mandated by tumor destruction of the occipital condyles. However, 3 weeks later, the patient suffered fatal rupture of the vertebral artery between C1 and C2. The fifth patient died after a lateral skull base approach performed at another institution for tumor recurring 2 years after the transoropharyngeal operation.

Of the 14 patients with a gross total resection and no radiation, 84% were tumor-free at follow-up. There was one death in this gross total removal procedure that has been previously described. Of the 16 patients with a subtotal resection, all underwent radiation at our institution using a larger dose more than 66 Gray, and presently, this is using the LINAC accelerator. Seventy-five percent of patients were without tumor progression. There were two deaths in this with recurrence at 2 years. Sixteen patients underwent partial resections. Five had previous resections and proton beam therapy, as described, and the deaths were in this group.

Postoperative deficits occurred with one patient who was clinically worsened and came in with a large retroclival and C1-2 spinal canal mass with quadriparesis. This patient was worse after surgery on the unaffected side but regained function in the affected side. One child presented with hindbrain herniation syndrome, and at operation, was found to have in additional tumor metastasis of chordoma without a primary focus. This patient is now at 1 year after the surgery.

Conclusions and Recommendations for Craniocervical Chordoma

Gross total or near total resection of chordomas of the cranial base should be attempted whenever possible (3, 7, 20, 27, 38, 46, 53). No one approach can be standardized. More than one approach may be required to achieve maximum safe resection. Instability at the craniocervical junction may be due to occipital condyle involvement or the surgical approach (22, 52, 59). Radical resection with functional preservation is the goal. A subtotal resection may be required, with radiation therapy to follow (36, 39). This is particularly true with clinically and histologically aggressive tumors, especially those in children and those that have been partially resected.

Foramen Magnum Meningiomas

Meningiomas are common at the foramen magnum and account for approximately 3% of all meningiomas (2, 5, 47). Women comprise between 66 and 73% of cases (29, 56). Typically, patients become symptomatic between 35 and 60 years of age, although meningiomas occasionally occur at the foramen magnum in children and may occur in the setting of neurofibromatosis (30, 45, 46).

The lesions typically are attached at the anterior rim of foramen magnum and frequently invade the region of the entrance around the vertebral artery and the exit of cervical nerve roots (5, 56). Lesions may extend above or below the foramen magnum equally. Numerous subtypes have been identified at the foramen magnum including the meningothelial, the fibrous, xanthomatous, clear cell, transitional, and the lymphoplasmacytic-ridge meningiomas (17, 64). In this present series of 41 patients, 4 occurred in the pediatric population. However, the average age of diagnosis was 50.1 years. Seventy-eight percent were women. The symptom duration was an average of 12.4 months. Pain was a major presenter in 68%, cranial nerve deficits in 68%, paresthesias in 29%, and lower cranial nerve deficits in 32% of patients. Motor deficit was seen in 26% and a coordination deficit in 32% of patients. Gross

involvement of the vertebrobasilar arterial system was recognized in 32% of patients.

Surgical treatment consisted of a posterior or posterolateral approach to foramen magnum and the clivus. In five of the individuals, a combined presigmoid and retrosigmoid craniotomy allowed for gross total removal. A gross total resection was accomplished in 87% of foramen magnum meningiomas (Fig. 27.4). Partial resection was made in 10% of patients, and a repeat surgical procedure was required in 3% of the patients. Surgery was combined with radiation therapy in 13% of individuals with a diagnosis of a clear cell or angioblastic meningioma. However, surgical complications with new deficit were seen in 32% of individuals. Of these, 50% had resolution of symptoms at their last follow-up. As previously mentioned, recurrence was seen in 13%, arterial involvement in 32%, and a fusion was required in two individuals after the posterolateral transcondylar approach. A tracheostomy was required in two individuals with new hypoglossal and vagal nerve dysfunction, which was not present before surgery.

Thus, the main stay treatment for foramen magnum meningiomas is surgical resection. Samii et al. (56) reported complete removal in 63% of patients and subtotal removal in 30%. The patient's mean Karnofsky performance score increased from 63 to 73. In the series by Arnautovic et al. (5), 67% had a gross total resection, subtotal resection in 22%, and a near total resection in 11% of their patients. These incomplete resections were mainly in patients who harbored a ventral foramen magnum meningioma.

Conclusions and Recommendations for Craniocervical-Foramen Magnum Meningiomas

Postoperative lower cranial nerve palsies are the most frequent surgical complication (45, 56) and are significant because they are associated with longer periods of hospitalization. They may be associated with repeated-aspiration pneumonia. Multiple-regression analysis has indicated that recurrent tumor, arachnoid scarring, craniovertebral meningioma, and no preoperative cranial nerve deficits but a new postoperative deficit were predictors for postoperative aspiration. Thus, aggressive comprehensive treatment is recommended once these deficits occur. Weakness, poor mobility, and aspiration pneumonia with deep venous thrombosis accounts for up to 50% of postoperative mortality (5, 29, 46, 56).

Other postoperative complications that require attention are cerebrospinal fluid leaks and meningitis after a far lateral-transcondylar approach, and may require cerebrospinal fluid diversion or ventriculoperitoneal shunt. Occipitocervical instability after the posterolateral-far lateral approach to the foramen magnum can be eliminated with judicious resection of no more than one third to one half of the occipital condyle (11, 16, 45, 59). Incomplete tumor removal and aggressive recurrence of invasive meningiomas will respond well to conventional radiation (35, 48, 51).

Our experience with foramen magnum meningiomas in the pediatric population comprises five children. Neurofibromatosis was seen in two. A clear cell meningioma at foramen magnum recurred 12 years later with metastasis in the sacrum and with complete destruction of the S2 and S3 vertebral bodies. This has responded to radical resection with follow-up radiation therapy. Stereotactic radiosurgery represents a valuable adjunct to therapy for incomplete tumor removal and aggressive tumors.

Calcium Pyrophosphate Masses or Pseudogout

There is an increasing geriatric population that presents with neurological deficits secondary to a large mass at the

foramen magnum and C1 (6, 45). Thirty-six patients are present in our series with an average age of 75 years. The male to female ratio is nearly equal. The duration of symptoms was 21 months, and pain was a major symptom in 90% of patients. Paresthesias occurred in 44%, cranial nerve deficits were seen in 44%, and the lower cranial nerve deficit was seen in 34% of individuals.

A calcium deposit was seen on high-resolution thin sliced computed tomography scans of the craniocervical region. This usually was present as specks within the tumor itself or around the periphery (46). The mass extended laterally into the joints between the occipital condyle and the lateral mass of C1 and C2.

A gross total resection was accomplished in every patient using the transoral-transpharyngeal route. There were no recurrences, and improvement or resolution of preoperative neurological deficit occurred in 75% of the patients. A fusion between the occiput and C1 was required in 78% of individuals.

>Conclusions and Recommendations for Craniocervical Pseudogout

This is a diagnosis that can be suspected before surgery and confirmed at frozen section. The recognition of abnormal birefringence is well documented. Unfortunately, these are older individuals who have significant co-morbidities that must be taken into consideration before planning operative intervention.

Miscellaneous Tumors

Plasmacytoma

There were five patients in this series. The average age was 60 years with a female to male ratio of 3:2 (21, 31, 42, 46, 49, 55, 62, 63). Average symptoms were very short in duration (4 mo). Pain was present in all individuals, as was cranial nerve deficit. A gross total removal was accomplished in two individuals and two others a partial removal. In one individual, a biopsy was obtained followed by radiation therapy. All patients improved after treatment. A disseminated plasmacytoma was not seen in our series, with an average follow-up of 8 years. The conversion to multiple myeloma was thus not seen in our series. However, this latter situation bodes a poor outcome.

Foramen Magnum Schwannomas

The average age of presentation was 38 years, with a male predominance in our series. Pain again was the major symptom in 63% of patients and paresthesias 25%. A lower cranial nerve deficit was seen in 38% and myelopathy in 50% of patients. A total gross removal was accomplished in all patients, and no recurrences were seen.

Patients with neurofibromatosis type I tend to become symptomatic at a younger age and have multiple or bilateral tumors. Fortunately, a significant number of neurinomas arise in a dorsal location, but in a rare situation, the schwannomas of the craniovertebral junction may be ventral to the dentate ligament (2, 10, 15, 40, 65). In such cases, section of the ligament or even a spinal accessory nerve may be required. Dumbbell neurofibromas can cause craniovertebral instability, and this possibility must be kept in mind at the time of reconstruction. The surgical outcome, as shown in our series, is excellent. This series, however, is skewed and has very few patients that were operated upon with schwannomas.

Bone Tumors

Eosinophilic granulomas were seen at C1 and C2 as solitary lesions (24, 54, 55). Osteoid osteoma and osteoblastomas occur at C1 and C2 and have a propensity to the posterior half of the vertebra (Fig. 27.5) (45, 46, 55). However, aneursymal bone cysts do have a predilection to the vertebral body. Fibrous dysplasia has not been encountered in the cervical spine but rather the skull base affecting the clivus, as well as the occipital condyles and exoccipital bone (1, 46). In our series, we have encountered several with involvement of the hypoglossal, vagal, and the spinal accessory nerves in decreasing frequency (Fig. 27.6). The tumor removal is always followed by consideration of stability and possible fusion. With osteoblastomas and aneursymal bone cysts, preoperative embolization of the tumor is always performed before embarking on operative intervention. Neurenteric cysts at the craniocervical borders are intradural and always ventrally situated. A complete resection is essential (14, 46).

CONCLUSIONS OF SURGERY AT THE CROSSROADS FOR CRANIOCERVICAL NEOPLASMS

The goal of the surgical approach is to maximize exposure of the tumor affecting the craniocervical region and involving the brain stem and spinal cord. One tends to minimize bone removal to maintain the stability at the craniocervical junction. Neurologically devastated individuals are less likely to obtain gainful resolution of their deficits. Thus, an early diagnosis when the tumor is small and the deficits are minimal tends to lead to better surgical outcomes and prognosis.

Co-morbidities such as cardiopulmonary involvement make it essential to have these assessed before surgery and also the involvement of the swallowing mechanism. Peculiarly, patients with a preexisting 9th and 10th nerve dysfunction do fair better than those who incur a neurological deficit after surgery. In any case, the possibility of prolonged postoperative tracheal intubation or tracheostomy must be presented before the operative intervention. Large lesions involving the spinal canal may require preoperative evaluation of cervical motion to assess stability and determine whether neurological deficits develop during flexion because of compression of the brain stem. This must be presented to the neuroanesthesiologist when intubation is made and to allow for appropriate positioning.

Most of the patients are approached via a posterolateral, far-lateral, or posterior midline route. We feel that compulsory use of the operating microscope, early tumor debulking during exposure, and gentle traction maintained away from the neural structures is required. Electrophysiological monitoring of the trigeminal, facial, glossopharyngeal, vagal, and hypoglossal nerves is performed routinely at our skull base center. Motor- and sensory-evoked responses have been used by the authors, but it is not though to be a required as part of the surgical procedures.

The transoral and anterolateral exposures have been used in primarily extradural lesions such as chordoma and pseudogout. The ventrally located schwannoma and meningioma have been approached via the far lateral-posterolateral route. Similarly, neurenteric cysts can be resected completely via this route.

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TABLE 27.1. Surgical approaches to the craniovertebral junction

PROCEDURE	EXTENT OF EXPOSURE	COMMON PATHOLOGY	ADVANTAGES	LIMITATIONS	RISKS AND LONG-TERM SIDE EFFECTS
Transsphenoethmoid	Good exposure of clivus and opposite side if ethmoid approach used; transsphenoidal route is midline only	Extradural clival and sellar disease	Well-tolerated midline approach to clivus only; short operative depth	Lower clivus poorly accessible and none below; cavernous sinus limits exposure	Injury to optic nerves and cavernous sinus
Transbasal	Anterior skull base; exposure of clivus is limited by two optic nerves going below sella	Lesions of sphenoethmoid bones and anterior skull base, (e.g. meningioma, fibrous dysplasia, chordoma)	May be combined with pterional approach to access middle fossa; Exposure of anterior fossa, clivus, C1	Optic nerves, sella and cavernous carotid artery	Limited exposure of clivus; careful reconstruction of base with bone, fascia and pericranium
Transoral Transpalato-pharyngeal	20-30 mm width of ventral midline, lower clivus, atlas and axis	Congenital development and degenerative diseases; extradural tumors	Simple direct; may be combined with palatal split, mandibular split	Pterygoid plate, hypoglossal nerves, Eustachian tubes, vertebral arteries	CSF leakage, inability to achieve clival closure, and possible instability. Hence, intradural surgery to be avoided

Transfacial	Utilizes LeForte osteotomies of maxilla; exposure from anterior fossae to sphenoid and entire clivus; lateral extent is into sphenomaxillary spaces and sinuses.	Extradural disease; chordoma, angiofibromas, fibrous dysplasia	Wide exposure	Poor dural coverage for CSF leak; miniplates needed to secure maxilla	Tracheostomy required; facial incisions; closure and coverage of dura and pharynx poor
Lateral extra-pharyngeal transcervical	Retropharyngeal midline CVJ and C1-C2; laterally to carotid canal and inferior petrosal sinus; exposure widened by facial nerve dissection and removal of submandibular salivary gland	Chordoma, metastasis, plasmacytoma, and basilar invagination	No communication with oral cavity; may be able to stabilize with bone or methylmethacrylate	Deep field with retraction of cranial nerves IX and XII and pharynx; Midline and contralateral exposure better	Pharyngeal dysfunction and palsies of cranial nerves IX and XII; lower facial nerve palsies
Lateral Transcondylar	Lower clivus and jugular bulb area below internal auditory canal; retrosigmoid craniectomy made	Extradural and intradural disease; meningioma, neurinoma, chordoma	No oral penetration; can be combined with infratemporal fossa procedures and may descend to expose vertebral artery for control; lateral brain stem easily visualized	Sigmoid sinus and condyle with hypoglossal nerve	CSF leakage versus plexus bleeding; stabilization may be needed if more than half of condyle removed
Lateral basal with infratemporal fossa exposure	Petrous bone, upper clivus, ventrolateral brain stem, and middle cranial fossa	Intradural tumors; meningioma, epidermoid, aneurysm, neurinoma	Control of carotid; no brain retraction	Combined approach needed to access lower clivus and foramen magnum; visibility across midline inadequate	Hearing loss; cuts across external ear canal and temporomandibular joint; facial nerve palsy; trismus
Dorsolateral Far lateral Lateral cerebellar	Midline posterior fossa to mastoid air cells, lateral half of condyle, and lateral atlantal masses; posterolateral spinal canal; inferior clivus, CP angle and lateral CVJ and cervical canal	Extradural and intradural disease at lower clivus and CVJ; aneurysms of proximal basilar artery	Rapid exposure, familiar field; good control of vertebral artery and also CP angle, ventral brain stem and cord may be combined with fusion; no brain stem retraction	May be combined with presigmoid posterior fossa exposure; basilar artery, anterior inferior cerebellar artery, O-C1 joints are limits	CSF leakage; communicating hydrocephalus; cranial nerves XI and XII at risk

| Posterior midline suboccipital C1-C2 laminectomy | Covers 120 degrees of CVJ dorsal circumference; access to both sides of midline | Dorsolateral and dorsal tumors; bony decompression of foramen magnum | May be combined with dorsal fusion | Cannot be used for lateral or ventral lesions | Very few disadvantages when approach is indicated |

CVJ = craniovertebral junction; CSF = cerebrospinal fluid; CP = cerebellopontine

FIG. 27.1   *A*, artist's illustration of the extent of exposure to the clivus and upper cervical spine in the lateral view. *B*, illustration of circumferential approach to the foramen magnum, clivus, and upper cervical spine with different operative techniques.

FIG. 27.2   *A*, gadolinium-enhanced sagittal magnetic resonance image of anterior skull base and craniocervical junction in a patient with chordoma replacing the clivus from the cribriform plate to the anterior arch of the atlas. *B*, gadolinium-enhanced magnetic resonance image axial view of chordoma replacing the anterior clivus with encasement of the carotid vessels. *C*, postoperative mid-sagittal T1-weighted magnetic resonance image after resection of clivus chordoma via the transmaxillary route. *D*, axial T1-weighted magnetic resonance image through the plane of mid-clivus demonstrating complete removal of the tumor via the transmaxillary route.

FIG. 27.3   *A*, composite of coronal magnetic resonance image with gadolinium enhancement through the plane of the anterior arch of the atlas and the odontoid process (*right*). Note the tumor replacing the clivus and in the ventral aspect of the posterior fossa. There is distortion of the carotid artery. This individual had a recurrent craniocervical-clivus chordoma. *B*, composite of axial magnetic resonance image with gadolinium enhancement through the plane of the midbrain (*left*) and pons (*right*). The recurrent chordoma involves the cavernous sinus, petrous bone, and ventral aspect of the left posterior fossa with involvement of the midbrain and pons. *C*, composite of parasagittal and mid-sagittal magnetic resonance image of the brain and upper cervical spine. Note the extent of recurrent chordoma involving the pterygopalatine fossa, the clivus, and ventral prepontine space. *D*, postoperative magnetic resonance image with gadolinium enhancement in the parasagittal and near mid-sagittal planes. The tumor has been removed using preauricular infratemporal approach. *E*, composite of axial magnetic resonance image with gadolinium enhancement through the plane of the midbrain and pons. Tumor resection was accomplished by a preauricular infratemporal fossa approach with replacement of the dura and filling of the resection cavity with fat. He has had no recurrence.

FIG. 27.4   *A*, composite of T2-weighted parasagittal magnetic resonance images through the craniocervical junction. Note the straddle foramen magnum meningioma with encasement of the left vertebral artery. *B*, composite of T2-weighted axial magnetic resonance image through the plane of the medulla. A large ventrally situated foramen magnum meningioma indents into the left ventrolateral medulla encasing the vertebral artery. *C*, operative photomicrograph of patient; *A* and *B*. left posterolateral-far lateral transcondylar approach was made to the foramen magnum ventral meningioma. The tumor is visualized ventral to the cervicomedullary junction. *D*, operative photomicrograph of patient. The ventral foramen magnum meningioma has been grossly excised.

FIG. 27.5   *A*, composite of computed tomography two-dimensional reconstruction of craniocervical junction to demonstrate the extent of atlantal osteoblastoma involving both the lateral masses and the occipital condyle. This patient presented with neck pain and quadriparesis. *B*, preoperative axial computed tomography scan demonstrating the erosive change of the entire anterior portion of the atlas vertebra by osteoblastoma.

FIG. 27.6   *A*, axial computed tomography scan through plane of foramen magnum and the condylar process of the mandible. This patient presented with neck pain and isolated left hypoglossopharyngeal, vagal, and hypoglossal paralysis with hearing loss. Fibrous dysplasia had caused expansion of the foramen magnum and the occipital condyle on the left with destruction of the lateral clivus. *B*, coronal computed tomography scan through the plane of the occipital condyles and the odontoid process. The fibrous dysplasia has destroyed the occipital condyle and involves the hypoglossal nerve. Decompression and dorsal occipitocervical fusion was made.