Transoral-transpharyngeal approach to the anterior craniocervical junction

Ten-year experience with 72 patients

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The anterior transoral-transpharyngeal operation to correct ventral irreducible compression of the cervicomedullary junction was utilized in 72 individuals. The patients' ages ranged from 6 to 82 years, and 29 were children. The pathology encountered was primary basilar invagination, rheumatoid irreducible cranial settling, secondary basilar invagination due to migration of odontoid fracture fragments, dystopic os odontoideum, granulation masses, clivus chordoma, osteoblastoma, and chondroma of the atlas. Fifteen patients had associated Chiari malformation with basilar invagination. Fifty-two patients required subsequent atlantoaxial or occipitocervical fusion. Neurological improvement was the rule. There were two deaths within 30 days of surgery: one from myocardial infarction 4 weeks after surgery and one from Gram-negative septicemia of urinary tract origin. There was one pharyngeal wound infection. The ventral transoral approach provides a safe, rapid, and effective means for decompression of the abnormal craniocervical junction.

KEY WORDS: basilar invagination, rheumatoid arthritis, craniovertebral junction, clivus chordoma, transoral-transpharyngeal approach, surgical approach

The first pathological description of the “abnormal manifestations of occipital vertebrae” was attributed to Meckel in 1815, by Gladstone and Erichsen-Powell. Following this initial description, pathological and autopsy studies were reported in reference to abnormalities of the craniovertebral junction (CVJ). After the classic radiographic study of basilar invagination published by Chamberlain in 1939, abnormalities of the CVJ emerged from the realm of anatomical and pathological curiosity into antemortem recognition and treatment.

The early surgical treatment of CVJ compressive pathology was posterior decompression. The operative procedure consisted of enlargement of the foramen magnum and removal of the posterior arches of the atlas and axis vertebrae. The postoperative mortality and morbidity rates associated with such treatment, when irreducible ventral compressive pathology of the cervicomedullary junction existed, were significant. Subsequently, operative procedures based on an understanding of the craniocervical dynamics, the site of encroachment on the neural elements, and the stability of the CVJ junction have been developed.

Summary of Cases

Patient Population

Between 1977 and 1987, 310 patients underwent surgery in this institution performed by the authors for symptomatic abnormalities of the CVJ. The factors that influence specific treatment are whether the bone abnormality can be reduced to its normal position, the etiology of the lesion, and the direction and mechanics of the compression.

The primary treatment for CVJ lesions that can be reduced is stabilization. An external immobilization is accomplished in those conditions such as ligamentous relaxation in inflammatory states to allow for reconstitution of the ligaments and bone. In other reducible pathological conditions with persistent instability, a dorsal fixation is necessary.

Surgical decompression of the cervicomedullary junction is required in patients with irreducible pathology. This disorder is subdivided into ventral or dorsal categories. In the former, the operative procedure of choice is transoral-transpharyngeal decompression, and the latter requires posterior decompression. If instability is present following either the ventral or dorsal
TABLE 1
Summary of surgical treatment at the craniovertebral junction (1977-1987)*

<table>
<thead>
<tr>
<th>Stability</th>
<th>Compression</th>
<th>Operative Approach</th>
<th>Postoperative Stability</th>
<th>Posterior Fusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>reducible: 160 (54)</td>
<td>none</td>
<td>immobilization: 23 (21)</td>
<td>stable: 20 (5)</td>
<td>137 (33)</td>
</tr>
<tr>
<td></td>
<td>ventral: 72 (29)</td>
<td>anterior.</td>
<td>unstable: 52 (24)</td>
<td>52 (24)</td>
</tr>
<tr>
<td></td>
<td>dorsal: 78 (37)</td>
<td>posterior</td>
<td>stable: 28 (11)</td>
<td>28 (24)</td>
</tr>
<tr>
<td>nonreducible: 150 (62)</td>
<td></td>
<td></td>
<td>unstable: 50 (26)</td>
<td>50 (26)</td>
</tr>
</tbody>
</table>

* Numbers in parentheses indicate children.

TABLE 2
Pathology in 72 patients with transoral-transpalatine clivus-odontoid resection, 1977-1987

<table>
<thead>
<tr>
<th>Pathology</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>primary basilar invagination</td>
<td>39</td>
</tr>
<tr>
<td>rheumatoid irreducible cranial settling</td>
<td>14</td>
</tr>
<tr>
<td>basilar invagination after malunion, odontoid-clival dislocation</td>
<td>4</td>
</tr>
<tr>
<td>upward migration of nonfused odontoid fracture (C1-C2 posterior fusion)</td>
<td>3</td>
</tr>
<tr>
<td>dystopic os odontoideum</td>
<td>5</td>
</tr>
<tr>
<td>granulation mass</td>
<td>3</td>
</tr>
<tr>
<td>cancer, pyrophosphate mass</td>
<td>1</td>
</tr>
<tr>
<td>osteoblastoma at C-1</td>
<td>1</td>
</tr>
<tr>
<td>chordoma lower clivus</td>
<td>1</td>
</tr>
<tr>
<td>chordoma at clivus-C-1</td>
<td>1</td>
</tr>
</tbody>
</table>

decompression, posterior fixation is required. All patients can be classified into one nonoperative and five operative categories for treatment purposes (Table 1). This paper focuses on the 72 patients between the ages of 6 and 82 years who underwent ventral transoral-transpharyngeal decompression of the craniovertebral junction (Table 2).

Investigative Studies
Preoperative neuroradiological investigations consisted of plain radiographs of the craniovertebral area, followed by dynamic pluridirectional polytomography in the flexed and extended positions. These studies define in detail the bone abnormalities, and demonstrate abnormal biomechanical and translation forces and effects of cervical traction for reduction. Early in this series, gas myelography was used to study soft-tissue abnormalities. This study has been subsequently replaced by computerized tomography (CT) myelography and magnetic resonance (MR) imaging.

Operative Procedure
Nasopharyngeal cultures were obtained 3 days prior to the proposed surgery. If no pathological flora were found, 2 million units of penicillin-G was given intravenously during the operation, and every 4 hours for the next 24 hours. The patient was placed supine on the operating table with 5 to 8 lbs of skeletal traction applied in an MR-compatible halo ring. Fiberoptic endotracheal intubation was accomplished, during which the patient was repositioned and examined while awake to insure no change in neurological status from positioning. A tracheostomy was performed under anesthesia to provide for better operative exposure and to insure an adequate airway. Following tracheostomy, a gauze packing was placed to occlude the laryngopharynx and to prevent blood from draining into the stomach.

A Dingman self-retaining mouth-retractor allowed for tongue depression and lateral retraction of the cheeks. A midline incision in the soft palate was made, extending from the hard palate to the base of the uvula, then deviating away from the midline. Stay sutures were used to provide retraction. The operating microscope is used at this point to provide magnification and a concentrated light source.

A midline incision was made in the posterior pharyngeal median raphe through the pharyngeal mucosa, the constrictor muscles, and the buccopharyngeal fascia. The prevertebral fascia and longus colli muscles were dissected from their ligamentous osseous attachment to expose the caudal clivus, atlas, and axis vertebrae. The anterior longitudinal ligament and occipital ligaments were coagulated and dissected free of the anterior arch of the atlas, the caudal anterior clivus, and the anterior aspect of the axis body. The anterior arch of the atlas was removed for a width of 3 cm in the midline to visualize the proximal odontoid process. When resection of the caudal clivus was necessary, the dura was separated from the posterior aspect of the clivus to prevent tearing and subsequent bleeding from the marginal sinus. All soft tissue ventral to the odontoid process was excised, after which the odontoid was resected in a rostral-caudal direction using a high-speed drill with a diamond burr. After removal of the tectorial membrane and/or additional granulation tissue, the surgeon was assured that adequate decompression had been accomplished when the pulsatile dura protruded ventrally into the decompression site.
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If the intradural exposure was required, a cruciate incision was made in the dura caudal to the foramen magnum and extended cranially. The marginal sinus was cauterized and the cerebrospinal fluid (CSF) was drained by a previously placed lumbar subarachnoid drain. Dural closure with 4-0 Vicryl sutures (polygalactin) was as complete as possible and facilitated by placing fascia harvested from the external oblique aponeurosis adjacent to the dural closure. A fat pad was then used to reinforce the fascia.

The longus colli muscles, the pharyngeal musculature, and the mucosa were approximated in individual layers using dyed 3-0 Vicryl sutures. The soft palate was closed in two layers and the pharyngeal pack was removed. Intraoperative somatosensory evoked potentials and brain-stem auditory evoked responses were recorded preoperatively and intraoperatively as neurophysiological monitors.23,35,42,43

Subsequent to surgery the patient was maintained with 5 lbs of skeletal traction. Intravenous hyperalimentation was continued for the first 5 to 6 days and no oral intake was permitted. This was followed by a gradual increase in feeding to a regular diet by the end of the 2nd week. If the dura was opened and a fascial graft used for repair, intravenous antibiotics and the spinal drainage were continued for 10 days after surgery. The tracheostomy was discontinued as soon as the patient's status permitted.

Evaluation of Postoperative Stability

Of the 72 individuals who underwent a ventral decompression, 52 patients required a dorsal fixation procedure. Pluridirectional lateral tomography of the CVJ was obtained 7 days after surgery to determine craniocervical stability. This was done in the flexed and extended positions, as well as with and without traction. An offset at the lateral occipitotlandoaxial articulations during flexion and extension studies or excessive displacement of the spinal-cranial axis with and without halo traction was indicative of instability. Magnetic resonance imaging of the cervicomedullary junction was likewise performed during this time. If instability was identified, posterior occipitocervical or atlantoaxial fusion using bone, acrylic, or bone plus acrylic was carried out at the identified unstable site.37

In these individuals with bone fusion, occipitocervical immobilization was maintained by a halo brace for 5 to 7 months. A sterno-occipitomandibular immobilizer (SOMI) was used in patients with rheumatoid arthritis who had undergone internal fixation with acrylic. The transverse portion of the cruciate ligament and the periosteum of the dens were not interrupted in the seven children below the age of 12 years. They were immobilized postoperatively in a halo or SOMI brace for 3 months. Two of these seven children later showed craniocervical instability and underwent dorsal fixation. The other five exhibited craniocervical stability, and three of these demonstrated new bone formation replacing the odontoid process.23,25 If the ligament was resected, a posterior fixation procedure was required. Dorsal fixation was usually necessary after transoral clivus odontoid resection in abnormalities associated with the atlas assimilation and/or segmentation failures of the C-2 and C-3 vertebrae. When osteoarthritis was present in the lateral atlantoaxial joints, posterior fixation was often not required in our experience. Rheumatoid cranial settling, by definition, is an unstable situation and all such patients required occipitocervical fixation.8,24 We have not utilized a ventral approach for fixation of the occipitotlandoaxial or atlantoaxial joints during the transoral operation for decompression, as the presence of devitalized bone in a potentially contaminated area could lead to infection.

Operative Results

All patients showed neurological improvement. Six individuals who were ventilator-dependent following either trauma or a previous primary posterior decompression had resolution of their neurological symptoms and signs in the postoperative period. Downbeat nystagmus, sleep apnea, and brain-stem signs were prominent features in 15 individuals with basilar invagination and the Chiari malformation. These signs regressed following the ventral decompressive procedure.

Intradural pathology was present in seven individuals. In four of these patients a sequestered odontoid process was found protruding into the pons secondary to rheumatoid cranial settling. In one patient, a clivus chordoma was present. Posttraumatic odontoid invagination and an abnormal location of the occipital condyle-lateral mass of the atlas resulted in disruption of the dura in another individual. One patient had previously undergone a transoral subtotal odontoid resection at another institution following anterior and lateral occipitocervical dislocation, resulting in residual odontoid bone in an intra-arachnoid location. In no instance was a pharyngeal flap or a septomucoperiosteal flap needed for closure of the dura or posterior pharyngeal wall.

Two patients died within the 1st month of operation. One of these was a 79-year-old man with rheumatoid cranial settling and basilar invagination who was markedly improved following his transoral decompression and posterior fixation. He suffered a myocardial infarction 4 weeks after his second operation and died. The other patient was a 52-year-old woman who was admitted quadriplegic and on a ventilator after a motor-vehicle accident. She had odontoid invagination into the pontomedullary junction. She was ambulatory following her transoral decompression with normal respiratory function. She died 3 weeks after the operation of Escherichia coli septicemia from a urinary tract infection that existed at the time of her admission.

One postoperative pharyngeal wound infection occurred in an 82-year-old man with rheumatoid cranial settling and odontoid invagination. He underwent a transoral resection of the compressive granulation tissue and the odontoid with subsequent posterior fixa-
tion. He had two episodes of epistaxis secondary to a retropharyngeal abscess that required drainage.

Representative Cases

Case 1

This 24-year-old woman was admitted with a 2-year history of excruciating occipital headaches and difficulty in using her left arm. She had previously been evaluated for Klippel-Feil syndrome, hearing loss, and facial numbness. Examination revealed limitation of neck movement, bilateral internuclear ophthalmoplegia, and hypalgesia in the trigeminal distribution on the left with a left hemihypalgesia. She had truncal ataxia and was hyperreflexic in her arms and legs. Pluridirectional tomography demonstrated complete atlas assimilation into the clivus and basilar invagination of the occiput and odontoid (Fig. 1 left). There was atlantoaxial instability on flexion-extension roentgenograms. Magnetic resonance imaging revealed vertical penetration of the odontoid into the posterior fossa causing ventral cervicomedullary junction compression (Fig. 1 center). Excessive granulation tissue present circumferentially around the odontoid process was best illustrated by the MR axial view (Fig. 1 right). The patient underwent a transoral resection of the abnormal clivus odontoid complex. Subsequent posterior occipitocervical fusion was necessary because of instability, and she had complete resolution of her neurological symptoms.

Case 2

This 12-year-old girl presented with headaches, difficulty with swallowing, thoracic scoliosis, and a spastic quadriparesis. A unilateral atlas-occipital assimilation was present, with the contralateral atlas fused to the axis (Fig. 2 upper). Vertebral angiography was markedly abnormal (Fig. 2 center). Computerized tomography with Amipaque outlined a Chiari I malformation. The odontoid process was transversely located, occupying the ventrolateral aspect of the foramen magnum. The cerebellar tonsils occupied the dorsal one-third of the foramen magnum, and a compressed medulla was seen in the center. Magnetic resonance imaging defined a lateral indentation of the pontomedullary junction by the abnormal occiput-atlas fused mass; a cervical cord syrinx was also revealed (Fig. 2 lower). The patient’s paresis and swallowing improved after ventral resection of the entire odontoid process. She subsequently underwent foramen magnum enlargement via the posterior route. A muscle plug was used to occlude the communication between the fourth ventricle and the central canal of the cervical spinal cord, and a fourth ventricle to subarachnoid shunt was made. A dural graft completed the posterior decompression, followed by an occipitocervical fusion. The patient has since shown marked neurological improvement.

Case 3

This 79-year-old man with rheumatoid arthritis was admitted with a tracheostomy in place after having aspirated on numerous occasions due to bilateral ninth, 10th, and 11th cranial nerve palsies. Other abnormal neurological findings included an internuclear ophthalmoplegia and a severe quadriparesis in addition to his peripheral rheumatoid arthritic changes. An MR image revealed a ventrally located mass indenting the cervicomedullary junction, with cranial settling (Fig. 3 upper). Pluridirectional tomography with metrizamide enhancement of the CSF showed the abnormality to contain both a sequestered odontoid process and pos-
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possible granulation tissue (Fig. 3 lower left). This sequestered odontoid process was best shown on the axial CT myelogram, where it was seen indenting and adhering to the pontomedullary junction with penetration of the dura (Fig. 3 lower center and right). At operation, the dural penetration was identified and the sequestered piece of odontoid was found adherent to the ventral surface of the medulla. This was isolated from the remainder of the mass and removed. Leakage of CSF was prevented by the technique outlined above under Operative Procedure, and a dorsal fixation was made. The cranial nerve deficits improved within the first 2 weeks after operation, and the patient was then ambulatory with assistance. He died of a myocardial infarction 4 weeks following his second operation.

Case 4

This 34-year-old woman had suffered from progressive diplopia, headaches, and arm weakness for 5 to 6 years. She underwent posterior fossa decompression for a Chiari I malformation with some improvement of her neurological function. Three months following the operation her neurological abnormalities were exacerbated with increasing headaches and gait difficulties. Magnetic resonance imaging showed the cerebellar vermis and tonsils impacted into the foramen magnum; the ventral pontomedullary junction was draped over an acutely angulated clivus-spinal canal junction (normal angle 125° to 137°) (Fig. 4). There was displacement of the posterior fossa contents into the cervical canal. The patient underwent resection of the clivus-odontoid complex by a ventral approach and a subsequent dorsal fixation. Her neurological condition improved following the operation.

Discussion

A transoral approach through the posterior pharyngeal wall has been an operation used for drainage of retropharyngeal abscesses for many years. Although this route provides direct access to the CVJ, it has not gained its well-deserved place in the neurosurgical armamentarium. The reasons for this can be attributed to initial reports of infection, limited exposure, CSF leakage, and unacceptable patient morbidity and mortality rates. Scoville and Sherman32 stated in 1951 that “the angulation of the medulla over the abnormally high odontoid process is the chief offender and causation of neurologic signs and disability in platybasia. Future surgical advance lies in the development of a successful removal of the odontoid, possibly through the mouth.” Over the past three decades, the value of the anterior surgical approach to ventral spinal canal pathology has been defined in several disorders.5,10,26,29,33,36 This operation is being increasingly used, incorporating the transoral approach to the ventral CVJ.1,8,11–14,16,18,19,23,27,28,31,35

In 1977, we formulated a physiological approach to correct pathology secondary to abnormality of the CVJ.23,25 With these guidelines, the high morbidity and mortality rates previously associated with posterior decompression of ventrally placed lesions were not seen. The advantages of the transoral-transpalatine approach to the CVJ compared to other operative approaches in irreducible ventral pathology include: 1) the impinging
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FIG. 3. Case 3. Upper Left: Midsagittal T1-weighted magnetic resonance (MR) image of the craniovertebral junction (CVJ) revealing odontoid invagination and a ventral mass (white arrow) displacing the medulla (m) posteriorly. Upper Right: Axial T1-weighted MR image 15 mm above the foramen magnum. The medulla (m) is displaced dorsally by a mass (white arrow) behind the clivus. Lower Left: Metrizamide myelotomogram in the midsagittal plane at the CVJ. The anterior atlantal arch is below the midportion of the axis body. The odontoid process (open arrow) is fractured at its base and angled dorsally and rostrally into the medulla (m). The fractured odontoid and fourth ventricle are outlined with dotted lines for clarity. Lower Center and Right: Axial computerized myelotomograms obtained 10 mm (center) and 15 mm above the foramen magnum (right). The odontoid sequestrum (arrow) displaces the medulla (m) dorsally and the vertebral arteries (arrowheads). In the more rostral section (right), the tip of the odontoid is intra-arachnoid (open arrow) and intimately related to the vertebral arteries and ventral aspect of the medulla (m).

Bone pathology and granulation tissue that accompany chronic instability are accessible only via the ventral route; 2) the patient is placed in the extended position, as opposed to a flexed position, thus decreasing the angulation of the brain stem during surgery; 3) surgery is performed through the avascular median pharyngeal raphe and through the clivus.

The indications for a transoral operation at the anterior CVJ must be exact. A precise definition of the pathology usually requires more than one radiological procedure. The diagnostic investigations should be chosen to complement each other in order to gain maximum information, as in our Case 3. Identification of a ventral abnormality at the CVJ, such as in rheumatoid cranial settling, does not by itself form an indication for a transoral operation.6 Unless the reducibility of the lesion has been attempted by cervical traction, a ventral operative decompression is of questionable value. Of the 65 patients in our series with rheumatoid basilar invagination and cranial settling, only 14 required a ventral decompression procedure. The remaining 51 patients had reduction of the odontoid invagination with cervical traction and required only a posterior stabilization procedure.24,37

The primary approach to intra-arachnoid lesions at the foramen magnum should not be via the transoral route unless other approaches prove ineffective. Several patients with clivus chordoma, meningioma, and schwannoma who were referred to our facility for an anterior transoral resection have undergone a suboccipital posterolateral operation with complete surgical resection of the pathology.
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The extent of surgical exposure of the anterior CVJ in the transoral approach is limited by the emergence of the hypoglossal nerves 1½ to 2 cm lateral to the clivus midline, by the vertebral arteries, and by the eustachian tubes lying just below the base of the skull. However, these structures allow 3 to 4 cm of lateral exposure which is sufficient for the removal of the pathology from the lower half of the clivus to the C2–3 interspace. The median labiomialdibular glossotomy combined with the transoral approach adds little to the exposure of these lesions rostral to the C-3 vertebra. The importance of a tracheostomy cannot be overemphasized. It allows for increase in the operative exposure and is a safeguard against airway obstruction from postoperative lingual swelling. It may obviate the need for reinsertion in the postoperative period with patients who have had respiratory insufficiency secondary to medullary compression or the need for reintubation if subsequent posterior decompression is required.

The soft-palate incision provides access to the lower clivus and the high CVJ. This technique provides superior operative exposure compared to suture of the soft palate and its retraction into the nasal cavity as advocated by some authors. Our surgical experience has satisfactorily demonstrated to us that the odontoid should be exposed to its rostral tip prior to its removal in a cranial-caudal direction. If the odontoid is first detached at its base and resected in a caudal-cranial direction, unfolding of the dura and ventral soft tissue impairs the resection and may result in dural tearing.

This latter technique has been proposed by some who advocate the extrapharyngeal route to the craniocervical region.

A Chiari malformation was identified in 15 of the 38 patients with an atlas-occipital assimilation and basilar invagination. Nine of these patients had undergone a primary operation consisting of a posterior fossa and upper cervical canal decompression. Rapid deterioration ensued in four patients, and in five cases an initial improvement was followed by progressive deterioration in neurological function. In each of these nine instances a postoperative MR image showed an increase in ventral cervicomedullary junction compression and angulation secondary to “peg-like” basilar invagination. Four patients had an associated increased caudal herniation of the cerebellum into the dorsally created enlargement in the cervical canal. This resulted in further foramen magnum impaction. In contrast, all 15 patients with assimilation of the atlas and basilar invagination with Chiari malformations improved after an anterior operation. The improvement involved the spasticity as well as reversal of the central spinal cord deficit. The visual abnormalities of internuclear ophthalmoplegia and the cranial nerve palsies also improved. This improvement could be attributed to relief of brain-stem angulation, improved blood supply, or changes in CSF craniospinal pressure dynamics.

The basic transoral approach has been used for clipping of midline aneurysms of the vertebrobasilar circulation. Disadvantages of this operation include accommodating the length of aneurysm clips in the
exposure and difficulty in obtaining dural approximation over the ventral aspect of the brain stem.

There is probably an inherent host immunity of the nasopharyngeal structures to the normal oral flora. Experience with otolaryngological procedures indicates that it is safe to conduct major surgical operations in the upper airway, using appropriate antibiotic coverage over when pathogenic flora are identified. Pharyngeal healing is remarkably rapid. A retropharyngeal abscess, with subsequent osteomyelitis of the atlas, occurred in one individual and required drainage. His immune status was questionable because of a 40-year history of rheumatoid arthritis and administration of steroids for many years.

In summary, this series has shown that the ventral transoral-transpharyngeal approach to the lower clivus and upper cervical spine is safe, rapid, and efficacious. This approach is important in relieving ventral irreducible pathology at the CVJ.

References
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Manuscript received October 1, 1987.
Accepted in final form May 23, 1988.
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