Craniocervical abnormalities

A comprehensive surgical approach

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Guidelines are proposed for surgical management of symptomatic abnormalities of the craniocervical junction. Experience with 17 recent cases is described. Gas or metrizamide (Amipaque) myelograms with pluridirectional tomograms revealed the etiology and mechanisms of compression of the cervicomedullary junction, as well as its reducibility. Stabilization was the goal in treatment of reducible lesions. Decompression of the cervicomedullary junction was paramount in irreducible cases. Ventral compression was treated in nine patients by transoral transpalatine resection of the odontoid-clivus complex, and all nine improved. A posterior decompression was carried out when bone impingement was present from the dorsal aspect. Fusion was performed in cases in which stability was not achieved by either procedure.

KEY WORDS □9 atlantoaxial dislocation □9 basilar invagination □9 spinal cord □9 medulla oblongata □9 spinal fusion □9 metrizamide

Abnormalities of the craniovertebral junction have only recently emerged from the realm of anatomical and pathological curiosity into the practical field of clinical neurosurgery. Since the first case of spontaneous atlantoaxial dislocation reported by Bell in 1830,4 numerous papers have been published concerning the radiological and pathological components of lesions found in this area.6,9-11,13,14,17,19,22,28,35,42,43,64,66,71-75 Of the 24 patients with developmental abnormalities in the region of the foramen magnum reported by Spillane, et al.,64 basilar invagination was seen in 20, Klippel-Feil deformity in two, chronic atlantoaxial dislocation in two, and occipitalization of the atlas in seven; each patient had neurological deficit. A similar distribution of such pathological entities has been reported by others.5,10,40,42,62,72,75 The surgical treatment of these conditions has generally been posterior decompression by enlargement of the foramen magnum and removal of the posterior arch of the C-1 vertebra. However, the mortality and morbidity associated with such treatment in irreducible lesions with cervicomedullary compression remains high.10,13,29,46,66,69

From our recent experience with 17 patients, we propose a plan for surgical management based upon the preoperative demonstration and understanding of the craniocervical dynamics.

Clinical Material

The basis of this report is a study involving 17 neurologically disabled patients with bone abnormalities in the region of the foramen magnum. Table 1 summarizes the symptoms, pathology, diagnostic studies, type of operation, and results of treatment of these patients.

The pathological anatomy of the cervicomedullary junction can be categorized for treatment purposes into reducible and irreducible pathology. (A reducible lesion refers to the capacity for restoration of normal anatomical relationships of the craniovertebral junction.) This is further classified into five major categories, based on the anatomical mechanisms causing neurological symptoms and signs (Fig. 1):

Group A: Reducible anterior compression of the cervicomedullary junction, requiring posterior stabilization (Case 1)
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**CRANIOCERVICAL JUNCTION ABNORMALITIES**

- Reducible (Needs 1st stabilization)
- Irreducible (Needs 1st decompression)
  - Encroachment
    - Ventral
    - Dorsal
  - Fusion
    - Transoral Decompr.
    - Posterior Decompr.
    - (A) Stable
    - (B) Unstable
    - (C) Stable
    - (D) Unstable
    - (E) Stable
    - Fusion

**Fig. 1.** Approach to abnormalities of the cervicobasilar junction.

**Group A:**

- Case 1. This 12-year-old rheumatoid arthritic girl had a 3-year history of progressive spastic quadriparesis, slurred speech, and neurogenic bladder. Midline polytomes with air myelography demonstrated no invagination of the odontoid process, but instability of the atlantoaxial articulation was evident (Fig. 2). In the flexion position, there was ventral impingement of the cervicomedullary junction by the odontoid process, with reduction of the “effective” anterior to posterior diameter of the cervical canal requiring stabilization (Cases 7–10)

**Group B:**

- Nonreducible anterior compression of the cervicomedullary junction by the dens, not requiring stabilization (Cases 2–6)

**Group C:**

- Nonreducible anterior compression of the cervicomedullary junction by the dens, requiring stabilization (Cases 7–10)

**Group D:**

- Nonreducible or partially reducible posterior compression of the cervicomedullary junction, requiring stabilization (Cases 11 and 12)

**Group E:**

- Nonreducible posterior compression of the cervicomedullary junction, not requiring stabilization (Cases 13–17)

Representative cases from each group are summarized below.

**Group A**

**Case 1.** This 12-year-old rheumatoid arthritic girl had a 3-year history of progressive spastic quadriparesis, slurred speech, and neurogenic bladder. Midline polytomes with air myelography demonstrated no invagination of the odontoid process, but instability of the atlantoaxial articulation was evident (Fig. 2). In the flexion position, there was ventral impingement of the cervicomedullary junction by the odontoid process, with reduction of the “effective” anterior to posterior diameter of the cervical canal requiring stabilization (Cases 7–10)

**Group B:**

- Nonreducible anterior compression of the cervicomedullary junction by the dens, not requiring stabilization (Cases 2–6)

**Group C:**

- Nonreducible anterior compression of the cervicomedullary junction by the dens, re-

**Fig. 2.** Case 1. **Upper:** Gas myelogram showing the odontoid process indenting the cervicomedullary junction in flexion. **Lower:** The impingement was reduced in extension.
<table>
<thead>
<tr>
<th>No.</th>
<th>Age (yrs), Sex</th>
<th>Clinical Presentation</th>
<th>Radiological Findings</th>
<th>Treatment</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>12, F</td>
<td>juvenile rheumatoid arthritis age 6 yrs; slurred speech; spastic quadriparesis; neurogenic bladder for 3 yrs</td>
<td>polyarticular rheumatoid involvement; severe atlanto-axial subluxation; CM compression, reducible in extension</td>
<td>halo cast in extension; posterior fusion C1-2</td>
<td>recovered</td>
</tr>
<tr>
<td>2</td>
<td>7, M</td>
<td>occipital headaches; neck stiffness 1 yr; proximal weakness of upper limbs; hyperreflexia throughout</td>
<td>abnormal clivus-odontoid articulation, clivus-canal angle $116^\circ$; ventral indentation of CM junction</td>
<td>tong traction; transoral resection of clivus-odontoid complex</td>
<td>recovered</td>
</tr>
<tr>
<td>3</td>
<td>67, M</td>
<td>diving accident at age 21 yrs; C-1 fracture; 2 yrs difficulty walking, spastic paraparesis</td>
<td>old Jefferson fracture; kinking of CM junction by invaginated odontoid</td>
<td>traction; transoral odontoid resection</td>
<td>improved; CSF leak closed</td>
</tr>
<tr>
<td>4</td>
<td>62, M</td>
<td>sudden blackout followed by dizziness &amp; objects &quot;dancing&quot; in visual field; downbeat nystagmus; unsteady gait</td>
<td>basilar invagination; fusion C2-3; assimilation of C-1; ventral CM junction impingement</td>
<td>traction; transoral clivus-C-1 odontoid resection</td>
<td>recovered</td>
</tr>
<tr>
<td>5</td>
<td>40, F</td>
<td>&quot;migraine&quot; headaches 4 yrs; neck pain, dizziness; short neck &amp; low neck hairline; atrophied supra-spinati; 1st arm dystaxia; hyper-reflexia</td>
<td>abnormal clivus-odontoid-C-1 anterior arch articulation; C2-4 fusion, C-1 assimilation; abnormal clivus-canal angle with severe ventral CM compression</td>
<td>traction; transoral clivus-C-1 odontoid resection</td>
<td>recovered</td>
</tr>
<tr>
<td>6</td>
<td>13, M</td>
<td>easy fatigability; loss of fine movements in hands, limitation of neck motion, Klippel-Feil syndrome; spastic throughout</td>
<td>fusion C2-3 &amp; C7-T1; assimilation of C-1, CM junction compression by invaginated odontoid</td>
<td>1: traction; transoral odontoid resection 2: decompression of foramen magnum &amp; C-1 posterior arch; occiput-C-2 posterior fusion</td>
<td>recovered</td>
</tr>
<tr>
<td>7</td>
<td>13, M</td>
<td>neck pain, gait difficulty, basilar invagination since age 13 yrs; suboccipital craniectomy &amp; C-1 resection; fall 2 yrs before; nystagmus; paraparesis; absent proprioception in legs</td>
<td>ventral indentation CM junction by odontoid process</td>
<td>traction; transoral resection; occiput-C-2 fusion</td>
<td>improved</td>
</tr>
<tr>
<td>8</td>
<td>40, F</td>
<td>hoarse voice, dysphagia; 74-lb weight loss 3 yrs; quadriparesis 1 mo; urinary incontinence; ptosis; 1st vocal cord paralysis; poor gag reflex; rapid worsening; respiratory arrest</td>
<td>basilar invagination; ventral CM junction impingement</td>
<td>traction; transoral odontoid resection; C1-2 occiput fusion</td>
<td>improved immediately; late sepsis, died</td>
</tr>
<tr>
<td>10</td>
<td>66, M</td>
<td>severe rheumatoid arthritis; weakness in arms 3 yrs; progressive quadriparesis 6 mos after a fall; hypalgesia below C-2; bedridden; no function in hands</td>
<td>C-1 compression; odontoid tip 30 mm above foramen magnum; occipital condyles articulate with C-2</td>
<td>traction; transoral clivus-C-1 odontoid resection; occiput-C1-2 posterior fusion</td>
<td>improved, walking</td>
</tr>
<tr>
<td>11</td>
<td>7, F</td>
<td>spondyloepiphyseal dysplasia; repeated falls; paresthias in arms, generalized weakness; nystagmus; spastic quadriparesis; sensation normal</td>
<td>os terminal; unstable C1-2; gliding of occiput, C-1, C-2; dorsal CM compression by C-1 in extension</td>
<td>traction; posterior decompression foramen magnum &amp; C-1; occiput-C-2 posterior fusion</td>
<td>recovered</td>
</tr>
<tr>
<td>12</td>
<td>7, F</td>
<td>spondyloepiphyseal dysplasia; spastic quadriparesis; downbeat nystagmus</td>
<td>os terminal; unstable C1-2 &amp; occiput; CM compression dorsally by C-1 as well as ventral angulation in flexion</td>
<td>halo traction; posterior decompression foramen magnum &amp; C-1; occiput-C-2 posterior fusion</td>
<td>recovered</td>
</tr>
</tbody>
</table>

*CM = cervicomedullary; CSF = cerebrospinal fluid.
### Table 1 (continued)*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Clinical Presentation</th>
<th>Radiological Findings</th>
<th>Treatment</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>13</td>
<td>16, F</td>
<td>Klippel-Feil syndrome; easy-fatigability; sleep apnea; could breathe when asked; on ventilator for 5 mos elsewhere; stiff neck, nystagmus, absent position sensation in limbs; pulmonary function 40% of normal</td>
<td>Klippel-Feil syndrome; basilar invagination; assimilation of C-1; vertebral angiogram &amp; myelogram; cerebellar tonsillar herniation</td>
<td>posterior decompression of foramen magnum &amp; C1-3; Arnold-Chiari malformation; adhesions lysed; 4th ventricle opened</td>
<td>sleep apnea resolved, improved</td>
</tr>
<tr>
<td>14</td>
<td>13, F</td>
<td>psychomotor retardation from infancy, slurred speech; quadripareis after fall 6 mos before; stiff neck, atrophy of hand muscles, spastic in legs, unable to stand</td>
<td>fusion of C-1 &amp; C-2 bodies; dens fused to foramen magnum; no instability</td>
<td>posterior fossa &amp; C1-2 decompression; lysis of adhesions &amp; band at CM junction</td>
<td>improved, ambulatory</td>
</tr>
<tr>
<td>15</td>
<td>48, M</td>
<td>vitamin D-resistant rickets, headaches, vertigo, weakness in arms; spastic hyperreflexic throughout; passing out spells with head in extension</td>
<td>hyperostosis of craniovertebral bones, shallow posterior fossa; C1-3 junction compression by C-1 arch, trigeminal artery</td>
<td>foramen magnum &amp; C-1, C-2 posterior decompression; release of vertebral arteries at C-1 from bone canal</td>
<td>recovered</td>
</tr>
<tr>
<td>16</td>
<td>60, M</td>
<td>unable to walk, bladder incontinence, drop attacks 2 yrs; webbed neck; spastic paraparesis</td>
<td>basilar invagination, neural arch of C-1 indenting CM junction, no instability; CT scan normal</td>
<td>suboccipital &amp; C1-2 decompression</td>
<td>improved, no drop attacks</td>
</tr>
<tr>
<td>17</td>
<td>40, M</td>
<td>numbness in hands 1 yr, weak grips; gait difficulty, spastic quadripareis, legs &gt; arms; hypalgiesia in arms; low hairline, cervicodorsal kyphosis</td>
<td>assimilation of C-1; hypoplastic odontoid; fusion at C2-3; dorsal CM compression by C-1; no instability</td>
<td>suboccipital &amp; C-1 decompression; Arnold-Chiari malformation, no syrinx</td>
<td>improved</td>
</tr>
</tbody>
</table>

*CM = cervicomedullary; CSF = cerebrospinal fluid.

(between the dens and posterior arch of C-1) to 7 mm. With extension, there was an increase of the diameter between the dens and the posterior arch of C-1 to 20 mm, with no evidence of impingement upon the neural elements. Accordingly, the patient underwent a posterior fusion of C1-2 in extension, followed by fixation in halo traction for 4 months and complete resolution of her neurological symptoms (Fig. 3).
Group B

Case 2. This 7-year-old boy had suffered from occipital headaches and neck stiffness for 1 year. Physical examination disclosed proximal weakness in the upper limbs, hyperreflexia throughout, and bilateral Babinski responses. Midline air-contrast lateral polytomography revealed an abnormal clivus-odontoid articulation with no evidence of invagination of the dens above Chamberlain's line (the diagonal between the hard palate and the posterior rim of the foramen magnum) (Fig. 4). The clivus-cervical canal...
angle measured 116°, compared with the normal angle, which is greater than 130°. The air-contrast myelogram demonstrated impingement of the cervicomedullary junction ventrally and a large subarachnoid space posteriorly. Subsequent to a transpalatine oral odontoidectomy and resection of the caudal clivus, there was resolution of neurological deficit. The patient demonstrated no instability after operation, obviating the need for fusion.

**Case 5.** This 40-year-old woman had a 6-month history of sudden syncope, followed by symptoms of dizziness and objects “dancing” in her visual field. Physical examination demonstrated downbeat nystagmus, diminished position and vibration sensation in the lower limbs, and unsteady gait. There was invagination of the dens above Chamberlain’s line, with ventral compression of the medulla and cervical cord (Fig. 5 left). In addition to odontoid invagination, an assimilation of C-1 to the occiput and failure of C2-3 segmentation were seen. Following transoral odontoid and caudal clivus resection, a normal clivus-cervical canal angle was reestablished (Fig. 5 right), and the patient’s symptoms cleared 6 weeks after surgery. There was no evidence of instability at the craniocervical junction.

**Group C**

**Case 7.** This 13-year-old boy presented with progressive fatigue. Physical examination demonstrated diminution of fine finger movement, increased tone in both upper and lower extremities with hyperreflexia, and limitation of flexion, extension, and lateral motion of the neck. The dens protruded 16 mm above Chamberlain’s line. Assimilation of the atlas into the occiput posteriorly, and failure of segmentation of the C-2 and C-3 vertebrae, were seen anteriorly. The mechanism of ventral compression and angulation and cervicomedullary structures by the dens was illustrated by a midline tomogram with air-contrast myelography (Fig. 6). No significant dorsal impingement was present. The dens and distal one-half of C-2 were removed by the transoral approach, with anterior decompression of the neural structures. The patient was maintained in skeletal traction with 5 lb of weight for 10 days after surgery. Following removal of traction, roentgenograms demonstrated rostral-caudal and ventral instability of the C-2-occipital junction. On the 10th postoperative day, the patient underwent posterior bone fusion and wire stabilization of C2-3 to the occiput. He was maintained in a halo brace for 6 months, and subsequently all neurological deficits resolved.

**Group D**

**Case 9.** This 66-year-old woman represented the only mortality in our series. She presented with rapidly progressive quadriplegia and respiratory arrest. She underwent anterior removal of the odontoid process and rostral one-third of the C-2 body, followed 2 weeks later by posterior fusion. She demonstrated marked improvement and became ambulatory, but 40 days after operation she died as a result of sepsis of urinary tract origin.

**Case 11.** This 7-year-old girl with spondyloepiphyseal dysplasia had a 3-year history of progressive generalized weakness, manifested by frequent falling and transient paresthesias in the upper extremities. Neurological examination demonstrated...
spastic quadripareisis, more in the upper limbs, and nystagmus to lateral gaze. The sensory examination was normal. Midline tomograms with metrizamide (Amipaque) myelography showed dorsal compromise of the cervicomedullary junction by the C-1 lamina posteriorly in both flexion and extension (Fig. 7). In addition, a reducible ventral encroachment was seen in flexion and relieved by extension. An ossiculum terminale was present. The patient underwent C-1 posterior arch decompression accompanying occipital-C1-2 lateral fusion because of gliding of the occiput on C1-2 and instability. Six months after surgery and subsequent bone fusion, there was no evidence of neurological deficit.

Fig. 7. Case 11. Upper: Metrizamide myelogram with cervicomedullary midline tomogram showing ventral and dorsal compression of the junction in flexion. Lower: In extension, the compression was mainly dorsal.
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Group E

Case 15. This 48-year-old man with Vitamin D-resistant rickets had a 5-year history of headache, vertigo, and progressive weakness in the upper limbs. Several episodes of transient syncope occurred simultaneously with neck extension. On physical examination, there was increased tone in all extremities with hyperactive reflexes. Syncope was reproduced with neck extension. Roentgenograms demonstrated hyperostosis of the craniovertebral bone structure and a shallow posterior fossa (Fig. 8). With positive contrast myelography (metrizamide), a narrow cervical canal was visualized, with the neural arch of C-1 indenting the cervicomedullary junction posteriorly. Four-vessel angiography demonstrated stenosis of both vertebral arteries at the foramen magnum between the occiput and the posterior arch of C-1. A persistent trigeminal artery was present. The posterior rim of the foramen magnum was removed along with the laminae of C-1, C-2, and C-3, with simultaneous decompression of the vertebral arteries. He had no postoperative instability. A year later his neurological examination was normal and he was free of syncopal attacks.

Discussion

Perhaps the most interesting feature of craniocevical abnormalities is their diversity. Each abnormality may vary in degree of deformity, its clinical effects, and in the pattern of association with neighboring skeletal structures. Basilar invagination was present in nine patients. The congenital type is often associated with atlantooccipital fusion. Acquired basilar invagination is found in diseases such as Paget's disease, osteogenesis imperfecta, rickets, osteomalacia, and hyperparathyroidism. The patient in Case 8 had posterior decompression for basilar invagination, but subsequently suffered neurological deterioration. She only improved after transoral resection of the invaginated odontoid process.

Klippel-Feil syndrome was seen in five cases, two of whom had Chiari malformation in conjunction with occipital assimilation of the atlas. Several authors report that up to 25% of patients with occipitalization of the atlas have Chiari malformation. The incidence of Chiari malformation in our series is similar to that reported by Spillane, et al., who recognized three such cases in 24 patients with developmental abnormalities in the region of the foramen magnum.

Dysgenesis of the odontoid process, commonly referred to as an unfused odontoid process, or os odontoideum, encompasses a variety of clinical conditions. There is enough supporting evidence to call the os terminale a congenital abnormality. With os odontoideum, ossiculum terminale, hypoplastic odontoid process, or non-union of odontoid fracture with upward displacement of the proximal segment, we believe that the cruciate ligament is incompetent. This results in an instability of the atlantoaxial articulation, and may produce cervicomedullary compression. If this is detected, a fusion procedure is essential after relieving the compressive pathology.

There are numerous conditions in which the occipital condyles may be hypoplastic, resulting in a forward gliding of the skull in its relationship to the spine. These have been described in Morquio's disease, spondyloepiphyseal dysplasia, and Conradi's syndrome. Spondyloepiphyseal dysplasia was present in a set of twins with ossiculum terminale and unstable occiput-atlas-axis relationships.

The abnormal clivus-odontoid articulation in two children (Cases 2 and 6) may represent a "third occipital condyle" with its confluent origin from the proatlas. Correction of this anomaly allows a change in angulation and reversal of symptoms. Two patients had rheumatoid arthritis, each with a decrease in the effective diameter of the sagittal canal at the level of the foramen magnum. The synovial bursae that surround the odontoid process and its associated ligaments are involved in the osseoligamentous destruction by this disease, with subsequent loss of stability. In a consecutive series of 104 autopsies on patients with rheumatoid arthritis, atlanto-
To axial dislocation was the main cause of death in eight and contributory in two patients.44 Sudden death had occurred in seven of 10 patients. Surgical indication to correct instability of the upper cervical spine in this disease is less than 5%.69 Ferlic, et al.,24 reported a series of 550 patients with rheumatoid arthritis who were admitted for surgical procedures; 12 had atlantoaxial and atlanto-occipital dislocation, and there were no children in the series.

Basilar invagination, Klippel-Feil syndrome, spondyloepiphysyal dysplasia, dysgenesis of the odontoid process, third occipital condyle, and rheumatoid arthritis all have potential or actual instability of the cervical spine. These are often seen in children with structural defects in either bone or ligamentous elements.14,25,26,34,36,49,73,75 These congenital or acquired defects may be associated with excessive motion and be vulnerable to minor trauma.

Although no cases are represented in this series, atlantoaxial subluxation associated with infection of the neck tissues occurs almost exclusively in children, and has yet received little attention.20,33,39,41,46,68 The lymphatic drainage of the occipitotlantalontoid joints is primarily into the retropharyngeal glands and then into the deep cervical chain.20,68 In Bharucha and Dastur's study64 of 40 patients with craniovertebral anomalies, the precipitating factor for the neurological deficit was trauma in 15 and infection in seven.

In the present series, the odontoid was abnormally placed, being invaginated into the posterior fossa as a result of non-union of an odontoid fracture in two patients, and from compression fracture of C-1 in two other patients. In Case 4, a posterior fixation of C1–3 had been previously performed for odontoid fracture. Over the ensuing 10 years, the sequestered odontoid apical segment migrated upward and resulted in verticacervicomedullary compression.

At the cervicomedullary junction, compromise of the neural structures results in a multiplicity of symptoms and signs which may present as myelopathy, cervical root and cranial nerve dysfunction, and/or vascular insufficiency.5,8,13,26,31,38,40,43,47,49,56,57,66 In the majority of symptomatic patients, both a sensory and motor disturbance were evident. Evidence of myelopathy was present in 16 of 17 patients, each presenting with different degrees of weakness in the upper and lower limbs. Paraparesis was a presenting feature in two patients. Motor symptoms may range from easy fatigability to obvious quadriplegia. Compromise of the pyramidal tracts by repetitive trauma and chronic compression of neural elements are presumed to be the etiology of motor deficits. Myelopathy mimicking the “central cord” syndrome was especially prominent in patients with basilar invagination. Following chronic implantation of hygroscopic plastic tumors in the foramen magnum, Taylor and Byrnes70 found progressive weakness in one or both upper extremities in monkeys. Examination of the spinal cord 6 weeks later demonstrated progressive downward dilatation of pericentral veins, anterior horn-cell hypoxic changes, small perivascular hemorrhages in the posterior horn cells of the lower cervical enlargement, and relative sparing of the white matter. They thus attribute the false localizing signs of high cervical compression to stagnant hypoxia secondary to venous distention. Studies have suggested that venous drainage of the cervical gray matter is rostral between T-1 and C-1, and that a separate drainage exists for the gray and white matter, resulting in the “central cord” syndrome.70 Similar studies based on autopsy examinations have demonstrated the selective vulnerability of the spinal cord gray matter to compressive anoxic changes.13,72

Brain-stem signs such as downbeat nystagmus were evident in two of the patients with cervicomedullary compromise, and nystagmus on lateral and upward gaze in an additional four patients. Downbeat nystagmus has been well documented accompanying cervicomedullary pathology.12 A relatively rapid medullary dysfunction was present in the patient in Case 9, her symptoms presenting as hoarseness, dysphagia, ptosis, and depressed gag reflex followed by respiratory arrest. Sleep apnea occurred in Case 13, demonstrating posterior impingement on the cervicomedullary junction secondary to Arnold-Chiari malformation and Klippel-Feil syndrome. Pulmonary function was assessed to be 60% impaired, and CO2 response was abnormal. Both of these patients demonstrated resolution of their respiratory symptoms after anterior and posterior decompression, respectively. Similar patients with respiratory insufficiency have been described by others, with resolution after decompression.32,37,38 Both clinical and experimental neurophysiological studies delineating the pathophysiology of respiratory arrest and sleep apnea have been published.5,37,38,48 If the afferent component of the central respiratory pathway is disrupted at the cervicomedullary level, normal facilitatory and modulatory feedback pathways are unavailable to make fine adjustments in the respiratory servo-mechanisms, although gross responses are handled satisfactorily when awake.57 Hence, routine blood gases and vital capacity may be deceptively normal.

Symptoms related to vascular disturbances of the cervicobasilar junction have been documented by several authors.13,48,49,51,53,54,57,60,71,72 These vascular symptoms include syncope, vertigo, intermittent attacks of altered consciousness and confusion, episodic hemiparesis, and transient loss in visual fields. Pathological conditions associated with vascular compromise include basilar invagination, atlantoaxial instability with and without ossiculum terminale,41 and an abnormally placed C-1 ring. The excessive mobility of an unstable atlantoaxial joint may cause repeated trauma to spinal cord vessels and intermittent obstruction of the vertebral and anterior spinal arteries upon neck motion. Both angulation and stretching, as well as occlusion of the vertebral arteries, have been
Pressive procedures for irreducible lesions of the posterior decompression resulted in improvement associated with the "central cord" syndrome, many. When the Chiari malformation was in only one-third of patients, the remainder being posterior decompression of the cervicomedullary junction is necessary when neural compromise occurs dorsally. If instability is present following either decompression operation, posterior or posterolateral fusion is carried out.

Before the era of skeletal traction, lesions of the occipitoatlantoaxial joint were marked by failure to achieve reduction in all except acute dislocations. After introduction of skeletal traction, it was apparent that the majority of dislocations, whether chronic or acute, could be reduced even when treated years after injury. Initially, posterior decompression was the treatment of choice with or without stabilization. The high operative risk associated with posterior decompressive procedures for irreducible lesions of the craniocervical junction has been the experience of many. When the Chiari malformation was associated with the "central cord" syndrome, posterior decompression resulted in improvement in only one-third of patients, the remainder being either worse or unchanged. Indeed, Dastur, et al., demonstrated six cases of hemorrhage within the medulla and upper cervical spinal cord after posterior decompression for basilar invagination or stable atlantoaxial dislocation when the compromise was ventrally situated. Two of our patients (Cases 4 and 8) had previously undergone posterior decompression or fusion procedures, with continued progression of neurological deficit.

The ventral transoral-transpalatine approach to the clivoatlantoaxial region has been used for fractures, infections, tumors, rheumatoid arthritis, and congenital abnormalities. A transpharyngeal approach has also been utilized. The operative technique that we have used is similar to that described by Greenberg, et al. In children, the transverse portion of the cruciate ligament complex and the cruciate notch on the axis is left intact to provide decompression with axis stability. Since the periosteum is not violated, this allows for future new bone formation and spontaneous fusion, which occurred in Cases 2 and 7. We have performed tracheostomy in each patient before transoral odontoectomy to provide for exposure during operation and adequate airway in the postoperative period. The patient is maintained in skeletal traction with intravenous feedings for 6 days, followed by graduated feeding orally to full diet by 14 days. Polytomies in flexion-extension are carried out 7 to 10 days after surgery; if instability is present, a posterior fusion is done with bone from either rib or iliac crest. External immobilization is accomplished using a halo cast for 6 months to achieve adequate fusion. None of the patients in this series who underwent transoral resection has experienced complications secondary to surgical infection and/or vascular injury.

The transpharyngeal route of stabilization of the atlanto-occipital joint in cases of trauma and tuberculosi was reported by Fang and Ong. They reported complications of vascular injury and infection of the pharyngeal wound. Others have refined the technique and demonstrated its safety and feasibility. De Andrade and MacNab extended the lateral extrapharyngeal approach to the craniocervical junction. They reported four patients who underwent anterior occipitocervical fusion, and reported subsequent stability in each case. Further refinement of these techniques may result in their future use.

Conclusions

From our recent experience, we have formulated a physiological approach to correct the pathology secondary to abnormality of the craniocervical junction. The factors that are used to influence specific treatment are: 1) etiology of the lesion and mechanisms of compression, and 2) whether the bone abnormality can be reduced to its normal position. These factors are determined by tomography, gas or metrizamide myelography, and vertebral angiography. The
primary aim of treatment of lesions that can be reduced with resolution of spinal cord-medullary compression is stabilization. Decompression of the cervicomедullary junction is paramount in irreducible cases. Ventral compression is treated by transoral these procedures, a fusion is performed. From the dorsal aspect. If instability follows either of decompression when bone impingement is present from the dorsal aspect. If instability follows either of these procedures, a fusion is performed.

References
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