Identification and management of high-risk patients with Klippel-Feil syndrome

MAHMOUD G. NAGIB, M.D., ROBERT E. MAXWELL, M.D., PH.D., AND SHELLEY N. CHOU, M.D., PH.D.

Division of Neurosurgery, Hennepin County Medical Center, and Department of Neurosurgery, University of Minnesota, Minneapolis, Minnesota

Patients with Klippel-Feil syndrome are often at high risk for neurological injury. The cervicomedullary junction and cervical spinal cord are especially vulnerable. Twenty-one patients examined and treated over a 20-year period are reviewed. The salient features of the syndrome are identified, and an approach to management is proposed.

KEY WORDS • Klippel-Feil syndrome • craniocervical abnormalities • scoliosis • spine

The classic triad of low posterior hairline, short neck, and limitation of neck motion is present in only 50% of patients with Klippel-Feil syndrome. Restriction of neck motion is the most common finding. Following their initial description of the syndrome, Klippel and Feil proposed a classification based on the site and extent of the cervical fusion: Type I applied to patients with extensive cervical and upper thoracic spinal fusion; Type II referred to patients with one or two interspace fusions, often associated with hemivertebrae and occipitoatlantal fusion; and Type III classified individuals with both cervical and lower thoracic or lumbar fusion. Type II is considered the most usual and asymptomatic form of this anomaly, and the C2-3 and C5-6 interspaces are most often fused. Fusion of the C2-3 interspace is thought to be autosomal-recessive. Although this classification is of genetic significance, it is of limited usefulness from the clinical and prognostic standpoint. Patients who have potential cervical instability or are at high risk for spinal cord injury may not be recognized. Cervical spine instability complicating the fusion pattern was identified by McRae. Hensinger, et al., described three potentially unstable cervical fusion patterns: fusion of C-2 and C-3 with occipitalization of the atlas, a long fusion with an abnormal occipitocervical junction, and a single open space between two fused segments. Commonly, the risks for neurological damage are due to abnormalities other than the fusion pattern of the cervical vertebrae. These include abnormalities at the occipitoatlantal junction, spinal canal stenosis, and scoliosis.

Visceral congenital anomalies associated with the syndrome may be the greatest threat to these patients. Cardiovascular anomalies have been recognized in patients with Klippel-Feil syndrome. Various lesions occur, but ventricular septal defects are the most common. Associated genitourinary anomalies have been identified in 64% of patients by Moore and his colleagues. Congenital absence of the uterus and upper vagina, as well as other abnormalities involving the genital tract have also been recognized. Jalladeau first reported the association of deafness with Klippel-Feil syndrome. Since then, several reports of other otolaryngological anomalies have appeared. A variety of ocular disorders have been recognized in association with Klippel-Feil syndrome. Wildervanck, followed by others, first described the cervico-ocular-auditory dysplasia usually seen in females with this disease. Cleft palate, mandibular malformations, and micrognathia have also been described in patients with Klippel-Feil syndrome.

Skeletal anomalies represent a common feature of the syndrome and include skull asymmetry, platybasia, basilar invagination, and brachycephaly. A 60% incidence of significant scoliosis was reported by Hensinger and his colleagues. Sprengel's deformity has been identified in 25% to 35% of these patients.
Radial and ulnar hypoplasia, finger and toe deformities, and the presence of an omovertebral bone are other rarely encountered anomalies. Dissociate movements of the two hands were identified by Bauman and others in patients with Klippel-Feil syndrome. An impaired pyramidal tract decussation has been proposed as a possible underlying mechanism by some investigators.

A retrospective analysis of 21 patients with Klippel-Feil syndrome admitted to the University of Minnesota Hospitals between 1962 and 1982 reveals that all of the patients had cervical and/or craniocervical abnormalities on plain spine radiographs. Certain patients were found to be at particularly high risk, however, for spontaneous progressive neurological deterioration or the sudden onset of neurological deficits following minor trauma. A classification of cervical spine and craniocervical abnormalities associated with Klippel-Feil syndrome is proposed that may assist the clinician in predicting the relative risk for neurological deterioration and for planning a conservative or surgical therapeutic approach based on the probable mechanism of injury.

**Summary of Cases**

Twenty-one patients with Klippel-Feil syndrome were admitted to the University of Minnesota Hospitals from 1962 through 1982. The cases were reviewed with regard to clinical presentation, radiographic findings, response to management, and outcome. Eight of these patients were admitted for evaluation and management of complications arising from the genitourinary, cardiovascular, and otologic anomalies associated with Klippel-Feil syndrome. Each of the patients in this consecutive series was examined and followed by at least one of the authors. There were 10 males and 11 females, ranging in age at the time first examined from newborn to 50 years. Two were infants, both of whom suffered from multiple congenital anomalies and died from congestive heart failure. All patients were followed for at least 1 year, except one patient who died 6 months after birth.

Younger children presented with delayed milestones, mirror movements, and impaired hearing acuity. Older children and adults complained of suboccipital headaches and neck pain, restriction of neck motion, torticollis, sudden or progressive gait disturbances, and sensory loss in the upper extremities. It was common to find abnormalities in several systems in one patient. Table 1 summarizes the frequency of associated abnormalities in this series. All the patients had skull and spine radiographs. Those with neurological deficits had spinal tomography and either Pantopaque or metrizamide myelography. Computerized tomography (CT) has recently been used in conjunction with metrizamide myelography in the radiographic evaluation of this disease. A summary of the neuroradiographic abnormalities demonstrated in these patients is provided in Table 2.

Eleven of the 12 patients who did not develop neurological symptoms or deficit had a single block fusion with no other cervical or craniocervical abnormalities. Nine of the fusions were across a single interspace (Table 2). One patient had partial assimilation of C-1 to the occiput and a C2–3 fusion. This patient had neck pain following a motor-vehicle accident, but never had neurological symptoms or signs. He responded to a brief course of cervical traction.

Nine patients developed neurological deficits either spontaneously or after minor trauma. This group had

### TABLE 1

<table>
<thead>
<tr>
<th>Anomalies</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Skeletal abnormalities</td>
<td></td>
</tr>
<tr>
<td>Sprengel's deformity</td>
<td>8</td>
</tr>
<tr>
<td>Scoliosis</td>
<td>8</td>
</tr>
<tr>
<td>Craniovertebral and craniocervical abnormalities</td>
<td>6</td>
</tr>
<tr>
<td>Cranial abnormalities</td>
<td>6</td>
</tr>
<tr>
<td>Spinal stenosis</td>
<td>2</td>
</tr>
<tr>
<td>Others</td>
<td>6</td>
</tr>
<tr>
<td>Facial abnormalities</td>
<td>6</td>
</tr>
<tr>
<td>Ocular abnormalities</td>
<td>6</td>
</tr>
<tr>
<td>Otolaryngological abnormalities</td>
<td>5</td>
</tr>
<tr>
<td>Genitourinary abnormalities</td>
<td>4</td>
</tr>
<tr>
<td>Cardiovascular abnormalities</td>
<td>2</td>
</tr>
<tr>
<td>Mirror movements</td>
<td>2</td>
</tr>
</tbody>
</table>

### TABLE 2

<table>
<thead>
<tr>
<th>Abnormities</th>
<th>Neurological Injury</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Without</td>
</tr>
<tr>
<td>No. of cases</td>
<td>12</td>
</tr>
<tr>
<td>One cervical vertebral block fusion</td>
<td>12</td>
</tr>
<tr>
<td>Across one disc space</td>
<td>9</td>
</tr>
<tr>
<td>Across two disc spaces</td>
<td>1</td>
</tr>
<tr>
<td>Across three disc spaces</td>
<td>1</td>
</tr>
<tr>
<td>Across five disc spaces</td>
<td>1</td>
</tr>
<tr>
<td>Two cervical vertebral block fusions</td>
<td></td>
</tr>
<tr>
<td>With one intervening disc space</td>
<td>2</td>
</tr>
<tr>
<td>With two intervening disc spaces</td>
<td>3</td>
</tr>
<tr>
<td>Subluxation of cervical vertebrae</td>
<td>3</td>
</tr>
<tr>
<td>C-3 on C-4</td>
<td>1</td>
</tr>
<tr>
<td>C-4 on C-5</td>
<td>1</td>
</tr>
<tr>
<td>C-5 on C-6</td>
<td>1</td>
</tr>
<tr>
<td>Myelographic defects</td>
<td>5</td>
</tr>
<tr>
<td>Foramen magnum block</td>
<td>2</td>
</tr>
<tr>
<td>Foramen magnum stenosis</td>
<td>1</td>
</tr>
<tr>
<td>Cervical spinal stenosis</td>
<td>2</td>
</tr>
<tr>
<td>Basilar impression</td>
<td>4</td>
</tr>
<tr>
<td>Occipitomastoiall assimilation</td>
<td>1</td>
</tr>
<tr>
<td>Hypoplastic dens</td>
<td>1</td>
</tr>
<tr>
<td>Cervical spine bifida occulta</td>
<td>1</td>
</tr>
<tr>
<td>Cervicothoracic scoliosis</td>
<td>2</td>
</tr>
<tr>
<td>Thoracic scoliosis</td>
<td>3</td>
</tr>
<tr>
<td>Syringomyelia</td>
<td>1</td>
</tr>
<tr>
<td>Lumbar meningocele</td>
<td>1</td>
</tr>
<tr>
<td>Skull asymmetry</td>
<td>2</td>
</tr>
<tr>
<td>Ventriculomegaly</td>
<td>2</td>
</tr>
<tr>
<td>Porencephalic cyst</td>
<td>1</td>
</tr>
</tbody>
</table>
Management of Klippel-Feil syndrome

more varied and complicated radiographic findings (Table 2). Patients were separated into three distinct groups according to the most significant cervical or craniocervical anomaly and the probable mechanism of injury:

Group I: Patients with an unstable fusion pattern (four patients)
Group II: Patients with craniocervical abnormalities (three patients)
Group III: Patients with associated spinal canal stenosis (two patients).

The clinical presentation, associated abnormalities, radiological findings, management, and outcome for each group of symptomatic patients are summarized in Table 3.

Illustrative Cases

Group I: Case 2

This 17-year-old girl had carried the diagnosis of Klippel-Feil syndrome since early childhood. Associated anomalies included thoracic scoliosis, unilateral pelvic kidney, and congenital aplasia of the fallopian tubes, uterus, and vagina. She became acutely quadriplegic following a minor fall and suffered a respiratory arrest. After she was successfully resuscitated, cervical spine films demonstrated a fracture-dislocation at C3-4. She underwent a limited fusion at another institution and was then transferred to the University of Minnesota. Additional cervical x-ray films were obtained (Fig. 1). Two fused segments of cervical spine with an open interspace at C3-4 were identified. A more extensive fusion was performed to provide stability and facilitate her treatment. The patient has remained quadriplegic and respirator-dependent.

Group II: Case 6

This 21-year-old man came to medical attention after a long history of neck and suboccipital pain. The patient had a short neck with restriction of mobility, and a low hairline. Bilateral hyperreflexia in his upper and lower extremities was evident, as well as bilateral ankle clonus and Babinski signs. An atlanto-occipital assimilation combined with hypermobility at the atlantoaxial joint and fusion of C3-4 was evident on radiographic examination (Fig. 2 left). A myelogram demonstrated no further block at the foramen magnum. Over the following 13 years the patient demonstrated no progression of his neurological deficits. The suboccipital and neck pain, ankle clonus, and Babinski signs did resolve.

The patient presented a second time with a complaint of bilateral hypesthesia involving his upper extremities and bilateral hyperreflexia. A repeat radiological examination demonstrated upward migration of the odontoid process (Fig. 2 right). A transoral resection of the odontoid process was carried out. The patient has remained asymptomatic for over 1 year.

Group III: Case 9

This 22-year-old man developed a quadriparesis after his automobile was struck from the rear in a minor motor-vehicle accident. On examination, the patient had a short neck and a low hairline. Hypoesthesia was noted below the C-5 dermatome as well as a bilateral Babinski sign. There was a weak rectal tone and urinary bladder areflexia. On radiological examination, fusion of C2-3 and partial fusion of C3-4 were noted, in addition to cervical spondylosis at C4-5 (Fig. 3). A generous decompressive laminectomy was performed at C4-6; however, the patient failed to improve.

Discussion

All 21 cases of Klippel-Feil syndrome in this series had cervical spine abnormalities visualized on plain spine radiographs. Nine patients (43%) required decompression or stabilization procedures because of neurological deficits which, in all but one case, occurred during the second or third decade of life. Three radiographic patterns were associated with radiculopathies and/or myelopathies. The first pattern consisted of two block vertebral fusions with an intervening open disc space. The second pattern consisted of craniocervical abnormalities such as atlanto-occipital assimilation and basilar invagination. These bone anomalies are commonly combined with hypermobility of the craniocervical junction, foramen magnum encroachment by tight dural bands, or upward migration of the odontoid process. Arnold-Chiari malformation, syringobulbia,
**TABLE 3**

Clinical summary of patients with Klippel-Feil syndrome presenting with neurological deficits*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Presenting Symptomatology</th>
<th>Clinical Features</th>
<th>Radiological Features</th>
<th>Procedure</th>
<th>Follow-Up Period &amp; Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>12, F</td>
<td></td>
<td>head locked to rt while swimming</td>
<td>short neck, mild microcephaly; bilateral hypoplasia over C-6 dermatome</td>
<td>C1–3, C4–5, C6–T1 fusion, subluxation of C-5 over C-6</td>
<td>C3–T2 fusion with autogenous bone graft &amp; halo cast immobilization</td>
<td>over 3 yrs: remained asymptomatic</td>
</tr>
<tr>
<td>2</td>
<td>17, F</td>
<td></td>
<td>quadriparesis &amp; respirator-dependent</td>
<td>paraplegia, thoracic scoliosis, congenital aplasia of fallopian tubes, uterus, &amp; vagina, unilateral pelvic kidney</td>
<td>C1–3 fusion, C4–5 partial fusion, C5–7 fusion, fracture-dislocation at C3–4</td>
<td>Cervical fusion C1–7 with autogenous bone graft</td>
<td>over 1 yr: quadriparesis &amp; respirator-dependent</td>
</tr>
<tr>
<td>3</td>
<td>30, F</td>
<td></td>
<td>neck pain with rt-sided torticollis</td>
<td>short neck, restriction of neck motion; low hairline, bilateral hyperreflexia of lower extremities and ankle clonus, Sprengel's deformity, multiple extraocular muscle palsies</td>
<td>C1–5 &amp; C6–T2 fusion, subluxation of C5–6, cervicothoracic scoliosis</td>
<td>C1–T2 posterior cervical fusion &amp; halo cast immobilization</td>
<td>over 3 yrs: no residual neck pain, no further progression of neurological deficits</td>
</tr>
</tbody>
</table>

**Group II: Patients with craniovertebral abnormalities**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Presenting Symptomatology</th>
<th>Clinical Features</th>
<th>Radiological Features</th>
<th>Procedure</th>
<th>Follow-Up Period &amp; Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>4</td>
<td>14, M</td>
<td></td>
<td>right torticollis</td>
<td>brachycephaly, limitation of neck motion; hypertelorism, thoracic scoliosis, mild bilateral conducing hearing loss, developed left hemiparesis; prompting further radiological studies after 1 yr</td>
<td>bilateral invagination, C1–3 &amp; C5–6 fusion, mild ventriculomegaly with a small 4th ventricle seen on CT of the craniocervical junction, obstruction to the dye flow at the foramen magnum seen on myelography</td>
<td>suboccipital decompression with C1–2 laminectomy</td>
<td>14 mos: no neurological deterioration developed, strength &amp; neck motion improved</td>
</tr>
<tr>
<td>5</td>
<td>10, M</td>
<td></td>
<td>gait difficulties, frequent stumbling</td>
<td>short neck, low hairline, restriction of neck movements, Sprengel's deformity, right hemiparesis &amp; hyperreflexia, bilateral Babinski sign</td>
<td>bilateral invagination, C2–3 fusion, C7–T2 spina bifida occulta, thoracic kyphoscoliosis; cervical &amp; thoracic spinal cord widening on myelography suggestive of syringomyelia</td>
<td>suboccipital cranieotomy &amp; C1–3 decompressive laminectomy; anterior spinal decompression performed</td>
<td>over 10 yrs: remissions &amp; exacerbations; died intraoperatively due to uncontrollable DIC</td>
</tr>
<tr>
<td>6</td>
<td>21, M</td>
<td></td>
<td>1st admission: suboccipital &amp; neck pain, 2nd admission: bilateral upper extremity paresis, mostly distally, unsteady gait</td>
<td>short neck, restriction of neck movements, low hairline, bilateral hyperreflexia with ankle clonus, Sprengel's deformity</td>
<td>1st admission: atlanto-occipital assimilation, atlantoaxial joint hypomobility; C3–4 fusion obstruction at the foramen magnum on myelography, 2nd admission: myelographic defect at the cervicoomedullary junction due to upward migration of odontoid process</td>
<td>1st op: craniectomy with C1 laminectomy &amp; fusion of occipital bone remains to C2–2 &amp; C3–3, 2nd op: transoral resection of odontoid process</td>
<td>over 1 yr: gradual resolution of hypoplasia, strength has improved</td>
</tr>
<tr>
<td>7</td>
<td>26, M</td>
<td></td>
<td>acute onset of suboccipital headaches, left facial nerve, shoulder, &amp; arm numbness, unsteady gait</td>
<td>skull &amp; facial asymmetry, palpebral fissure asymmetry, high arched palate, cleft palate, left facial hypoplasia, decreased left corneal reflex, left hemiparesis, right hemihypoplasia, bilateral Babinski sign, bilateral hammer toes, thoracic scoliosis, Sprengel's deformity</td>
<td>bilateral invagination, congenital absence of anterior arch &amp; right lateral mass of C1–1; C2 articularis with skull base; small deformed dens, multiple fused cervical vertebrae, cervicothoracic scoliosis</td>
<td>suboccipital cranieotomy with C1–2 decompressive laminectomy; patch graft of dura to provide more space at craniocervical junction</td>
<td>over 1 year: gradual resolution of hypoplasia, strength has improved</td>
</tr>
</tbody>
</table>

**Group III: Patients with associated spinal cord stenosis**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Presenting Symptomatology</th>
<th>Clinical Features</th>
<th>Radiological Features</th>
<th>Procedure</th>
<th>Follow-Up Period &amp; Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>8</td>
<td>13, M</td>
<td></td>
<td>mental retardation, poor school performance</td>
<td>neck pain, hypoplasia along left C6–7 dermatome; 6 strength of left elbow/flexors, decreased hearing acuity on left</td>
<td>assimilation of C-1 to occiput, C2–3 fusion, C5–7 spina bifida, C5–6 stenosis on myelography</td>
<td>C5–6 laminectomy followed by anterior cervical fusion at C5–6</td>
<td>over 8 yrs: cervical pain &amp; motor weakness improved; died from myocardial infarction</td>
</tr>
<tr>
<td>9</td>
<td>22, M</td>
<td></td>
<td>quadriparesis following motor-vehicle accident</td>
<td>short neck, low hairline, hypoplasia below C-5 dermatome, bilateral hyperreflexia &amp; Babinski sign; decreased rectal tone, bladder areflexia</td>
<td>C2–3 fusion, partial fusion at C3–4; cervical spondylolisthesis at C5–6 visualized on myelography; bilateral aplasia of thumbs &amp; radii, multiple carpal bones</td>
<td>C4–6 decompressive laminectomy</td>
<td>8 yrs: no neurological change</td>
</tr>
</tbody>
</table>

* CT = computerized tomography; DIC = disseminated intravascular coagulation.
† Two years later, gait difficulty recurred, CT confirmed previous myelographic picture of syringomyelia; syrinx cavity shunted. Progressive improvement for 1 yr, then recurrent gait difficulties; kyphoscoliosis progressed.
and syringomyelia may also be associated with these anomalies. The third pattern consisted of fusion of one or more cervical vertebrae associated with stenosis of the cervical spinal canal. This combination of findings has not received as much attention in the literature as have the first two radiological patterns. The stenotic segment may occur above, below, or at the level of fusion. The question of when does hypermobility become significant instability enough to warrant further reduction of neck motion by a fusion procedure has not been well defined in the absence of progressive symptomatology or sudden injury to the spinal cord or nerve roots. The possible role of prophylactic surgical stabilization of the cervical spine in the asymptomatic patient or in patients with neck pain is a debateable issue, however. We thus propose the following approach for patients at high risk for neurological injury.

**Group I Patients**

If a potentially unstable cervical spine is recognized, the patient is advised to avoid contact sports and other activities placing the spine and spinal cord at risk. Spinal fusion may be indicated if the patient becomes symptomatic or if a progressive deformity is radiologically evident, as this group of patients appears to be particularly prone to disastrous spinal cord damage following trivial trauma. As in Group I, patients in Group II are encouraged to avoid stressful physical activities. If neurological deficit and/or radiological changes occur, a surgical approach is considered. Deficits resulting from cranio-cervical abnormalities are often due to instability, basilar invagination, or occipitoatlantal assimilation. In these instances we have followed an approach similar to that recommended by Menezes, et al. A precise knowledge of the anatomical relationships between the bone framework, cerebrospinal fluid space, and neural elements is essential. Pluridirectional tomography with metrizamide or gas myelography through the cranio-cervical junction provides valuable information. Careful flexion/extension during the latter studies demonstrates the presence and degree of instability. The recent use of CT following metrizamide myelography can clearly demonstrate the pathological anatomy, particularly the area of bone encroachment. The association of posterior fossa abnormalities and syringomyelia can also be visualized. An attempt at careful reduction by skeletal traction is recommended if basilar invagination is present. In the event of successful reduction, posterior stabilization is indicated, provided that additional impingement in the area of the foramen magnum is not present. In the event of an irreducible lesion, which is usually the case in our experience, ventral or dorsal...
fig. 3. case 9. left: lateral view of the cervical spine demonstrating fusion of c2-3 and partial fusion of c3-4. center and right: cervical myelograms, anterior and lateral views, demonstrating a significantly spondylotic segment at c4-5.

decompression is indicated, depending upon the site of encroachment. posterior stabilization may be necessary if craniocervical instability is present or develops postoperatively.

group iii patients

patients in group iii are encouraged to avoid contact sports or other stressful physical activities. if these individuals develop symptoms related to the stenotic spinal canal, decompression of the spinal cord is indicated.

operative procedures

a summary of the suggested surgical approaches for symptomatic patients is provided in fig. 4. whenever a surgical procedure is considered for patients with klippel-feil syndrome, a meticulous physical examination is essential since serious associated anomalies which are not evident at first might be present. cardiopulmonary evaluation to exclude associated congenital cardiac anomalies or respiratory insufficiency should be undertaken. routine performance of an intravenous urogram has been recommended. consultation with otolaryngologists and oral surgeons is important, as problems during intubation are a common occurrence. the presence of oral deformities or cleft palate might render a transoral approach for de-
Management of Klippel-Feil syndrome

compression particularly difficult. A labio-mandibular-transoral approach as described by Delgado and his colleagues might prove necessary.

Conclusions

The clinical presentation and course of 21 patients with Klippel-Feil syndrome examined and treated over a 20-year period is presented. Their ages ranged from newborn to 50 years old. No sex predominance was noted. Eight of the patients came to medical attention for associated abnormalities unrelated to the spinal or craniocervical junction deformities. Recognition of the syndrome in this group of patients should prompt extensive evaluation of the craniocervical junction and spine for early identification of patients at high risk for neurological damage.

Three distinct groups of patients are recognized: Group I: Patients with an unstable fusion pattern; Group II: Patients with craniocervical abnormalities; and Group III: Patients with cervical spinal canal stenosis. The approach for the management of each group is proposed in light of the clinical picture and radiological investigations. Whenever a surgical procedure is considered, careful attention should be given to the presence of associated anomalies. Significant surgical risks are thus avoided.

References

37. Mc Rae DL: Bony abnormalities in the region of the foramen magnum: correlation of the anatomic and neu-
logical findings. Acta Radiol 40:335–354, 1953
39. Mol W, Detmar S: The combination of Sprengel's de-
formity with Klippel-Feil disease and the presence of an os 
40. Moore WB, Matthews TJ, Rabinowitz R: Genitourinary 
abnormalities associated with Klippel-Feil syndrome. J Bone 
41. Morrison SG, Perry LW, Scott LP III: Congenital brevi-
collis (Klippel-Feil syndrome) and cardiovascular anom-
42. Mündnich K: The dysplasias of the middle and the inner 
ear in different types of malformation. Proc R Soc Med 
67:1197–1198, 1974
43. Notermans SLH, Go KG, Boonstra S: EMG studies of 
associated movements in a patient with Klippel-Feil syn-
44. Palant DI, Carter BL: Klippel-Feil syndrome and deaf-
ness. A study with polytomography. Am J Dis Child 
123:218–221, 1972
45. Park II, Jones HW Jr: A new syndrome in two unrelated 
females: Klippel-Feil deformity, conductive deafness and 
syndrome of vertebral anomalies. Report of six cases and 
syndrome of Wildervank. ZWR 83:157–163, 1974
49. Shenoi PM: Wildervank's syndrome. Hereditary malfor-
mations of the ear in three generations. J Laryngol Otol 
86:1121–1135, 1972

50. Sherk HH, Dawoud S: Congenital os odontoideum with 
Klippel-Feil anomaly and fatal atlanto-axial instability. 
51. Singh SP, Rock EH, Shulman A: Klippel-Feil syndrome 
with unexplained apparent conductive hearing loss. A 
52. Sommerfeld RM, Schweiger JW: Cleft palate associated 
27:737–739, 1969
53. Stadnicki G, Rassumowski D: The association of cleft 
palate with the Klippel-Feil syndrome. Oral Surg 33: 
335–340, 1972
54. Stark EW, Borton TE: Hearing loss and the Klippel-Feil 
55. Strax TE, Baran E: Traumatic quadriplegia associated 
with Klippel-Feil syndrome: discussion and case reports. 
Arch Phys Med Rehabil 56:363–365, 1975
56. Wildervanck LS: [Klippel-Feil syndrome, associated with 
abducens paralysis, bulbar retraction and deaf-mutism.] 
Ned Tijdschr Geneeskd 96:2752–2756, 1952 (Dut)
57. Willemsen WNP: Combination of the Mayer-Rokitas-
ny-Kistler and Klippel-Feil syndrome — a case report 
and literature review. Eur J Obstet Gynecol Reprod Biol 
58. Wycis HT: Lipoma of the spinal cord associated with 

Manuscript received September 1, 1983. 
Accepted in final form March 27, 1984. 
Address reprint requests to: Mahmoud G. Nagib, M.D., 
Division of Neurosurgery, Department of Surgery, Hennepin 
County Medical Center, 701 Park Avenue South, Minneapo-
lis, Minnesota 55415.