Skull Base Manifestations of Camurati-Engelmann Disease

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Objective: To describe presenting symptoms, evaluation findings, and surgical management of cranial base hyperostosis in patients with Camurati-Engelmann disease (CED).

Design: Retrospective study and literature review.

Setting: The Mayo Clinic, Rochester, Minnesota.

Patients: A total of 306 patients diagnosed as having CED, including 12 primarily evaluated at our institution between 1968 and 2008, and 294 identified in the international literature.

Main Outcome Measures: Presenting symptoms, methods of diagnosis, treatment strategies, and patient outcomes.

Results: One hundred seventy-three of 306 patients (56.5%) had radiographically proven skull base hyperostosis, whereas less than one-fourth were symptomatic. The most common manifestations of cranial base involvement were hearing loss (19.0%), headache (10.4%), exophthalmos (8.2%), and frontal bossing (7.2%); less common were vision changes, vertigo, facial weakness, symptomatic brainstem compression, facial numbness, and hyposmia. Although corticosteroids and bisphosphates may treat torso and extremity involvement, they demonstrate no benefit for symptomatic skull base disease. In select symptomatic patients, aggressive decompression surgery may provide the only means of treatment. Decompression surgery is more challenging with thick sclerotic bone, loss or obscuration of bony landmarks, and decreased supratentorial space. Patients must be counseled on the increased risks associated with surgery and the potential for redeposition of bone and recurrence of symptoms.

Conclusions: Physicians should include CED in the differential diagnosis for patients with radiographic evidence of skull base thickening and synchronous cranial neuropathies or symptoms of elevated intracranial pressure. In mild forms of the disease, the clinical course of patients should be followed with serial examination, audiometric testing, and radiography. In select patients with progressive cranial base symptoms, aggressive wide decompression of involved neurovascular structures may provide benefit.


CAMURATI-ENGELMANN DISEASE (CED), also known as progressive diaphyseal dysplasia, is a rare autosomal dominant disorder characterized by progressive symmetric diaphyseal sclerosis of the long bones and potential cranial hyperostosis. Camurati-Engelmann disease belongs to the larger class of craniofacial bone disorders that include osteopetrosis, craniodiaphyseal dysplasia, and van Buchem disease. Since its first report in 1920 by Cockayne and later descriptions by Camurati and subsequently Engelmann, we identified a total of 306 cases published in the international literature.

The gene linked to the development of CED codes for the β1 subunit of transforming growth factor (TGFB1; OMIM 190180) was isolated to chromosome 19q in 2000. Mutations of this gene result in increased transforming growth factor activity and subsequent stimulation of osteoblastic bone formation and suppression of osteoclastic resorption.

Presenting symptoms commonly include musculoskeletal complaints of lower limb weakness, muscle pain, and gait unsteadiness, whereas cranial base manifestations occur in less than a quarter of patients. The mandible is sclerotic in approximately 25% of patients and infrequently may be enlarged. Symptomatic cranial base involvement is variable and is caused by bony overgrowth of the skull base, leading to foraminal stenosis and diminished cranial vault volume resulting in neurovascular compromise and potentially increased intracranial pressure (ICP), respectively.
After institutional review board approval, we performed a retrospective review from 1968 through 2008 using the electronic medical record, and all patients diagnosed as having CED were included. Data were collected with respect to age, sex, presenting symptoms, evaluation, imaging, surgical management, and treatment outcomes. In addition, a literature review dating back to the first reported case in 1920 was performed to calculate the prevalence and characteristics of presenting symptoms, treatment outcomes, and associated skull base findings.

RESULTS

In the past 40 years, a total of 12 patients (4 male and 8 female) with the diagnosis of CED underwent evaluation and treatment at the Mayo Clinic. Of these, 8 (67%) were found to have radiographic evidence of skull base thickening and concurrent cranial base signs and symptoms of disease (Table 1). The average age at onset of symptomatic cranial base involvement was 24 (range, 15-35) years.

INITIAL PRESENTATION OF DISEASE

The mean age of first symptom onset was 8.7 (range, 1.5-30) years, with 11 of 12 patients initially reporting lower extremity pain, 9 describing gait unsteadiness or ataxia, and 2 complaining of lower extremity weakness. In addition to pain and gait instability, patient 3 experienced a pathologic fracture of his left tibia at 4 years of age, which led to the diagnosis of CED, and patient 8 required extensive spinal fusion surgery at 19 years of age for severe scoliosis. Two unrelated patients (3 and 5) experienced rapid growth in their later teenage years, one of whom required epiphysiodesis to remedy her uncontrolled lower extremity growth and pain. Both patients underwent additional molecular diagnostic testing and were found to have a heterogeneous missense mutation (c.652C→T [R218C]) of the TGFB1 gene. Nine of 12 patients had a known family history of disease, whereas 3 cases appeared to be sporadic.

OPHTHALMOPATHY, HEADACHE, AND BRAINSTEM INVOLVEMENT

Four patients (mean age, 29 years) developed progressive bilateral symmetric exophthalmia (Figure 1A and B), 3 of whom were asymptomatic without optic neuropathy. The fourth (patient 2) experienced deteriorating visual acuity. All 4 patients had radiographic anterior cranial floor thickening and shallow orbits, and 3 of 4 demonstrated considerable frontal bossing.

At 14 years of age, patient 2 underwent evaluation for increased headaches, blurred vision, and papilledema. During lumbar puncture she was found to have an elevated opening pressure of 35 cm H2O. Skull base radiography demonstrated diffuse bony thickening. At 30 years of age (1976), after worsening headaches and deteriorating vision, she underwent a right subtemporal decompression with postoperative improvement of her neurologic symptoms. She is now 63 years of age and has not had a recurrence of visual symptoms or headache.

Patient 6 experienced headache and vision changes in the absence of exophthalmia or radiographic anterior cranial fossa involvement. Early in the third decade of life, he developed worsening dysphagia to solids and liquids, vision changes, and intermittent debilitating headaches exacerbated by Valsalva maneuvers. Plain radiographs of the skull base demonstrated a stenotic heart-shaped foramen magnum, and magnetic resonance imaging found an Arnold-Chiari malformation. At 41 years of age (2001), he underwent a suboccipital craniectomy with C1 laminectomy, which completely resolved his headaches, visual symptoms, and dysphagia without recurrence.

Four other patients experienced headaches but, in contrast to patients 2 and 6, they did not report vision changes.
or other overt symptoms of increased ICP or brainstem compression.

AUDIOVESTIBULAR AND FACIAL NERVE INVOLVEMENT

A total of 6 patients were diagnosed as having hearing loss (4 bilateral and 2 unilateral), 3 of whom reported concurrent dizziness, while none demonstrated facial nerve weakness or spasm. All 6 patients with hearing loss had pure sensorineural deficits most commonly affecting the higher frequencies, and no patients were found to have mixed or conductive losses. Patients 2, 5, and 8 underwent surgical intervention for progressive internal auditory canal (IAC) stenosis, whereas patients 3, 6, and 7 were conservatively observed with no worsening of symptoms.

By 40 years of age, patient 2 experienced deteriorating hearing and balance without facial nerve weakness. Further testing revealed a hypofunctioning right labyrinth and bilateral moderate sloping to profound sensorineural hearing loss (SNHL). Computed tomography of the head and temporal bones demonstrated significant skull base thickening with bilateral IAC stenosis. In 1987, she subsequently underwent a right retrosigmoid craniotomy with IAC decompression. Postoperatively she experienced profound right-sided SNHL and complete facial paralysis requiring right lateral tarsorrhaphy. Her balance and vision remained stable, and hearing in the left ear (which did not undergo surgery) continued to deteriorate.

By the middle of the fourth decade of life, patient 5 noted subjective hearing loss, and an audiogram found symmetric bilateral high-frequency SNHL. Computed tomography of the temporal bone confirmed the presence of severe narrowing of her IACs bilaterally (Figure 2A and B). She denied any vertiginous symptoms but felt generally unsteady; results of vestibular testing were within normal limits. Given her worsening hearing, considerable bilateral IAC stenosis, and family history of aggressive phenotypic disease, at 42 years of age she elected to proceed with staged bilateral middle fossa IAC decompressions in 2008 and 2009 (Figure 2C and D). During the year since surgery, she demonstrated no deterioration of hearing.

During the fifth decade of life, patient 8 experienced increasing bilateral high-pitched tinnitus, vertigo, and bilateral hearing loss. Electronystagmography demonstrated bilateral vestibular hypofunction, and audiometric evaluation found profound bilateral SNHL. At 58 years of age (2001), he underwent a planned 2-stage procedure that included middle fossa decompression of his right IAC and subsequent cochlear implantation 2 months later.
He was observed for 6 years after the initial implantation and continued to derive benefit from his device. Soon thereafter, he died of unrelated pulmonary disease.

Patient 3 developed progressive hearing loss and tinnitus spanning a 15-year period starting in the middle of the third decade of life. An audiogram demonstrated normal hearing in the right ear and mild sloping to moderate downsloping SNHL in the left ear. He denied symptoms of dizziness. At 38 years of age, his hearing had remained stable and he had not undergone any surgical intervention.

During his adolescent years, patient 6 developed mild bilateral hearing loss and high-pitched tinnitus without vestibulopathy or facial nerve weakness. Plain radiographs of the skull base demonstrated sclerotic petrous ridges. Despite radiographic evidence of lateral skull base involvement, his audi vestibular symptoms remained stable. At 49 years of age, he showed no further deterioration.

By 38 years of age, patient 7 reported significant right-sided hearing loss, dizziness, and high-frequency tinnitus. Audiometric evaluation demonstrated mild right-sided high-frequency SNHL. Results of subsequent vestibular testing were within normal limits. Given her mild unilateral symptoms, no surgical interventions were pursued. At 48 years of age, she had stable audi vestibular symptoms and had not required surgical decompression.

MEDICAL MANAGEMENT

Eleven of 12 patients were medically treated for chronic lower extremity pain. Of these, only 2 found relief with nonsteroidal anti-inflammatory drugs, 6 improved with the addition of prednisone therapy, and 1 required a combination of nonsteroidal anti-inflammatory drugs, corticosteroids, and narcotic pain medications. Two patients experienced chronic refractory pain and underwent a trial of bisphosphonate treatment with no additional perceived benefit. Despite any reported improvements in lower extremity pain, corticosteroid and bisphosphonate therapy did not improve headache or other neurological symptoms in any patients with skull base involvement.

During the past 30 years, we have examined a total of 12 patients who were ultimately diagnosed as having CED, 8 of whom were found to have asymptomatic skull base involvement (Table 1). We performed a review of the literature dating back to the first account with Cockayne2 in 1920 and, including the present 12 patients, there have been a total of 306 unique cases of CED. The average age at the onset of symptoms was 13.4 years, with 45.7% of patients being male and 54.3% female. Documented radiographic skull base involvement was described in 173 patients (56.5%). The most common skull base symptoms included hearing loss, headaches, frontal bossing, and ophthalmopathy (Table 2). Many of the published cases did not specifically focus on skull base or neurological disease and therefore may have left out certain details; as a result, we acknowledge that our review of the literature may underestimate associated signs and symptoms of skull base involvement.

D I A G N O S I S A N D R A D I O G R A P H I C F I N D I N G S

The symptoms and physical manifestations of CED are extremely variable even among affected kindred.20 Only 74% of patients with TGFB1 mutations express clinical symptoms, whereas radiographic abnormalities are found in approximately 94% of patients.12 Multiple reports in the literature describe asymptomatic patients who were diagnosed as having CED after incidental radiographic findings21–23 or molecular testing.23 Owing to the phenotypic variability and incomplete penetrance, establishing sensitive clinical criteria has proven difficult.7,12 Among reports, a large variety of diagnostic and prognostic tools have been described in the evaluation of CED. Radiography and genomic mutational analysis have proven highly sensitive, whereas other investigatory strategies that include biopsy,24–26 peripheral blood smear, erythrocyte sedimentation rate, and measurement of serum calcium, phosphorus, and alkaline phosphatase levels have been met with less success.23

Radiographic screening with plain x-rays and computed tomography is a sensitive diagnostic tool and provides valuable information for making the initial diagnoses and establishing the extent of involvement. Bone scintigraphy has been shown to complement standard radiographic evaluation when determining disease activity and progression.27 The hallmark of CED is progressive symmetric diaphyseal hyperostosis of the lower extremities and is found in 94% to 98% of patients.12,23 Skull base involvement has been estimated to occur in more than half of all patients12 and, in our review, cranial base sclerosis occurred in 173 of 306 published cases (56.5%).

The defining characteristics and differential diagnosis of CED are well described in previous publications.1,2,9,12,18,29 There are multiple sclerosing bone disorders in patients with normal stature that commonly affect the skull base. Van Buchem disease (generalized corti-

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**Table 2. Prevalence of Skull Base Manifestations Among 306 Published Cases**

<table>
<thead>
<tr>
<th>Skull Base Finding</th>
<th>Cases, No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Radiologically confirmed sclerotic skull base</td>
<td>173 (56.5)</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>58 (19.0)</td>
</tr>
<tr>
<td>SNHL</td>
<td>22 (7.2)</td>
</tr>
<tr>
<td>CHL</td>
<td>4 (1.3)</td>
</tr>
<tr>
<td>MHL</td>
<td>3 (2.6)</td>
</tr>
<tr>
<td>Unspecified</td>
<td>24 (7.8)</td>
</tr>
<tr>
<td>Headache</td>
<td>32 (10.5)</td>
</tr>
<tr>
<td>Exophthalmos</td>
<td>25 (8.2)</td>
</tr>
<tr>
<td>Frontal bossing</td>
<td>22 (7.2)</td>
</tr>
<tr>
<td>Vision changes</td>
<td>17 (5.6)</td>
</tr>
<tr>
<td>Papilledema</td>
<td>14 (4.6)</td>
</tr>
<tr>
<td>Vertigo</td>
<td>13 (4.2)</td>
</tr>
<tr>
<td>Facial paresis/paralysis/spasm</td>
<td>13 (4.2)</td>
</tr>
<tr>
<td>Brainstem herniation and compression</td>
<td>5 (1.6)</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>3 (1.0)</td>
</tr>
<tr>
<td>Facial numbness</td>
<td>2 (0.7)</td>
</tr>
<tr>
<td>Hyposmia</td>
<td>1 (0.3)</td>
</tr>
</tbody>
</table>

Abbreviations: CHL, conductive hearing loss; MHL, mixed hearing loss; SNHL, sensorineural hearing loss.

*Includes patients from the following references: 2 to 4, 7, 8, and 12 to 174.*
cal hyperostosis) is a rare autosomal recessive disease characterized by sclerosis of the skull, mandible, and clavicles. A common, striking feature is a large overbearing mandible, and, rarely, the circumference of the skull is enlarged. Craniodiaphyseal dysplasia can be distinguished with characteristic massive facial bone enlargement, hypertelorism, paranasal bossing, and nasal flattening. Ribbing disease is often viewed as a milder variant of CED; in contrast, Ribbing disease affects the skeletal system in an asymmetric manner.30,31

Genetic screening may be helpful, particularly in cases of no known family history of CED (≤30% of cases are sporadic) or when symptoms are subtle and results of diagnostic tests are inconclusive. In their report, Janssens and colleagues21 reviewed 100 cases from 24 separate families and found mutations in the gene coding for TGFBI in virtually all symptomatic patients.

SKULL BASE MANIFESTATIONS AND MANAGEMENT

Although more than 50% of patients with CED will have radiographically confirmed skull base involvement, less than 25% experience cranial nerve dysfunction. Owing to the symmetric but often irregular cranial base involvement,29 the laterality and progression of individual cranial neuropathies are unpredictable. Some patients may note abrupt unilateral hearing loss, facial nerve paralysis, or vision loss; others may experience gradual or fluctuating unilateral or bilateral symptoms.16,32 Hyperostosis is 2 to 3 times more common in the anterior and middle fossa compared with the posterior fossa.29 Cranial neuropathies are thought to result from foraminal narrowing or increased ICP.18,19,33 Generalized vasculopathy occurs in many patients, resulting in thickening of vessel walls,13,34-38,175 which may further compromise tenuous vascular flow through already stenotic bony canals.

Given the rarity of CED, there are currently no randomized controlled trials documenting pharmacologic effectiveness of corticosteroid or bisphosphonate therapy. Multiple isolated reports document improvement in lower extremity gait coordination, muscle pain, exercise tolerance, and appetite after corticosteroid administration.12,15,17,23,32,35,57 Radiographic and histological reversal of disease has been described in select cases.40,42,46-48 whereas others report no such improvements.49 Bisphosphonate therapy has been met with less success; few reports document improvement,50 whereas most report no change51,52 or detrimental effects.46 Medical therapy, including corticosteroids and bisphosphonates, does not appear to benefit cranial base disease progression or symptoms.18,42,43,55,57

The primary management of progressive symptomatic skull base involvement remains surgical decompression in carefully selected patients. Surgery is technically demanding, with sclerotic bony changes.18,177 decreased pneumatization of paranasal sinuses,25,38,29 reduced space for supratentorial access,5 and potential obstruction or loss of critical landmarks.16,53,55 Patients should be counseled on the increased risks associated with surgery,17,18,56,57 and additional measures such as intraoperative image guidance should be considered particularly for cranial nerve decompression. Cranial base manifestations of disease are mainly caused by foraminal stenosis and increased ICP; as such, interventions addressing both conditions are met with the most success.17,19 Bony regrowth after decompression remains a concern. In our literature review, 3 of 28 decompression procedures had radiographically proven bony regrowth.16,23,54,58

Temporal bone involvement may result in cochleovestibulopathy and facial nerve weakness. In our review of 306 cases, hearing loss was the most commonly cited cranial base symptom and occurred in 58 of 306 patients (19.0%).8 In a 1996 review, Higashi and Matsuki5 found a similar rate of 17.7% (23 of 130). Hearing loss can be conductive, caused by external auditory canal stenosis,18,35,60 ossicular9 or stapes fixation,17 chronic effusion secondary to eustachian tube narrowing and dysfunction,63,67,76 overgrowth of the round or oval window,5 and cholesteatoma formation.83 Sensorineural hearing loss may result from otic capsule involvement78,79 or IAC neurocompression, and mixed hearing loss can occur from a combination of these factors. The type of hearing loss was documented in 34 of the 58 cases (64.7% SNHL, 11.8% conductive hearing loss, and 23.5% mixed hearing loss). Gait unsteadiness is a frequently encountered symptom, whereas vertigo was found in only 13 of 306 patients (4.2%).16,30,53-55,63,67,70,77,78 Similar to many patients with SNHL, vertigo is most likely related to neurovascular compression from critical IAC stenosis.53,55 Facial nerve weakness or spasm is the most common motor cranial neuropathy and has been documented in 13 cases (4.2%).† Less commonly, trigeminal neuropathy occurs, manifesting as facial numbness with or without masticytic weakness.17,17

Treatment of hearing loss requires a keen understanding of the disease process and potential sites of involvement.78 Conductive hearing loss may be managed by conventional hearing aid amplification,50,57 bone-anchored hearing devices,50 stapedectomy,78 or ventilation tube insertion,79 depending on the underlying cause. Those with early or mild SNHL and radiographic IAC stenosis may initially benefit from conventional hearing aids77 but should consider middle fossa9 or retrosigmoid IAC decompression with the goal of longer-term hearing preservation. Those with advanced SNHL may benefit from cochlear implantation with79 or without57 IAC decompression, depending on the degree of IAC stenosis. Current knowledge dictates that critical IAC stenosis is a contraindication to cochlear implantation78, however, we have found that staged cochlear implantation after adequate IAC decompression is beneficial for hearing restoration in patients with CED.79

Internal auditory canal decompression in CED is technically demanding given the sclerotic nature of the bone and obscuration of normal surgical landmarks. Including patients in the present series, we identified a total of 14 IAC decompressions (in 11 patients) performed for hearing loss, vestibular symptoms, or facial nerve weakness18,17,23,32,35,53,55,57; 3 of these reported iatrogenic injuries to the facial or cochleovestibular nerve coma.
plex,17,51 8 demonstrated lasting hearing improvement or stabilization,16,12,33,55 and 3 were met with initial hearing stabilization followed by eventual hearing decline and recurrence of symptoms.10,23 Two reports discuss isolated 8- and 15-dB high-frequency SNHL after successful IAC decompression for vestibular symptoms; they implicate acoustic trauma or heat injury associated with prolonged drilling with a diamond burr.33,55

The progression of facial nerve involvement is unpredictable.16,32 It has been hypothesized that those with an intermittent progressive course may be afflicted by repeated acute viral inflammation resulting in neural edema and arterial tamponade, as in Bell’s palsy.16,179 Those with a more indolent course may have progressive venous congestion associated with foraminal overgrowth, resulting in edema and subsequent arterial occlusion.16 Although there are multiple reports of successful facial nerve decompressions in other skull base hyperostotic syndromes,13,179,180 only 4 patients with CED underwent decompression for facial nerve weakness or spasm.16,17,32,57 Two of these documented long-term success, attributing favorable outcomes to extensive IAC and fallopian canal decompression.16,32 Given the potential for multi-level involvement, full facial canal (or, at a minimum, IAC and labyrinthine segment) decompression is advocated.32,33,179

Ophthalmopathy occurred in less than 10% of patients and included exophthalmos,28,30,32 papilledema,† and optic nerve atrophy.30,33,54,60,89 Ocular changes are attributed to bony overgrowth of the orbit, optic canal stenosis, and increased ICP.89 Anterior fossa involvement commonly manifests with frontal bossing and exophthalmos,28 and hyposmia has been reported in 1 patient.23 Most patients with globe protrusion are asymptomatic, but a small percentage may experience diplopia and, in extreme circumstances, globe subluxation.85 Early reports had attributed vision loss and blindness to optic nerve compression, and surgical management primarily involved optic nerve sheath fenestration and narrow field optic nerve or orbital decompression; however, such strategies were met with only limited success.54,58,60,63 Growing evidence suggests that ophthalmologic deterioration is caused by the effects of optic nerve compression and increased ICP; only after aggressive broad cranial decompression (usually through a subtemporal craniotomy) and restoration of normal ICP are orbital symptoms consistently improved.19,54,70,81,82

Increased ICP is thought to result from diminished intracranial volume from diffuse bony overgrowth and jugular foramen stenosis, resulting in compromised internal jugular venous outflow.17,76 The true incidence of elevated ICP in CED is unknown because only symptomatic patients may undergo lumbar puncture.19,62 Presentation is variable and may include headache28 (10%), vision changes‡ (4%), papilledema (3%), and other cranial neuropathies. Associated gait changes may be difficult to appreciate given that most patients demonstrate lower extremity weakness and muscle pain as presenting symptoms independent of skull base involvement.12

Including 1 patient from this report, a total of 5 patients with CED had symptomatic brainstem herniation.18,19,81,82 The exact mechanism is unclear but is theorized to result from caudal displacement of the brainstem due to progressive intracranial volume loss combined with elevated ICP.18,19,81,82 All 5 patients experienced severe headaches and vision changes, had progressive imbalance,18,19,81 and 2 experienced dysphagia.18 There has been only 1 report of dysphagia and diminished gag reflex outside those with symptomatic brainstem compression.17 Jugular foramen syndromes manifesting with compression of cranial nerves IX, X, and XI have not been associated with CED; we suspect this is because of the infrequent involvement of the posterior fossa29 and the relatively large diameter of the jugular foramen.

Acetazolamide therapy,19,62,82 therapeutic lumbar puncture,19,62 and ventriculoperitoneal shunting55,81 have been used for temporary management, but most patients eventually require decompressive cranietomies. Isolated suboccipital craniectomy procedures have been met with mixed success, and 1 case documents intraoperative death.18 It is generally recommended that patients with increased ICP with or without brainstem herniation undergo broad decompressive cranietomies to improve or restore cranial vault space.81,82

**RECOMMENDATIONS FOR FOLLOW-UP**

The management of cranial base involvement requires multidisciplinary surveillance and treatment. On diagnosis, those with CED should undergo screening radiographic bone surveys, and all patients with skull base involvement should have baseline computed tomography of the head and neck to confirm the presence and extent of disease. Since more than half of all patients with skull base hyperostosis are asymptomatic, and because there may be concordance between the degree of sclerosis and symptoms, only patients who are symptomatic require further imaging.

Surveillance in patients with asymptomatic skull base involvement should include annual ophthalmologic, otolaryngologic, and neurologic examinations. Eye examination should evaluate for proptosis, exposure keratitis, papilledema, and loss of visual acuity; evaluation by the otolaryngologist may provide insight into cranial neuropathies and audiovestibular abnormalities; neurological assessment should emphasize cranial nerve involvement but also evaluate for signs of increased ICP and brainstem herniation. More rigorous monitoring is required for those with progressive symptomatic disease who would consider surgical intervention because earlier decompression may result in more favorable outcomes.

Camurati-Engelmann disease is a rare disease that results in symptomatic cranial base hyperostosis in less than one-fourth of patients. Hearing loss and headache are the most frequent findings, whereas ophthalmopathy, facial nerve weakness, brainstem herniation, and

†References 23, 24, 38, 54, 58, 60, 62, 75, 76, 80-83.

‡References 23, 24, 38, 54, 58, 60, 62, 75, 76, 80-83.
trigeminal neuropathies are less common. In asymptomatic and mild forms of disease, patients should be followed up with serial examination, audiometric testing, and imaging.

Unfortunately, there remains no known pharmacologic or surgical strategy for disease reversal. In contrast to lower extremity disease progression, medical therapy has not been successful in treating associated skull base symptoms. Surgery with aggressive wide bony decompression is technically challenging but remains the only option for patients with advanced disease.

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Author Contributions: All authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Carlson, Beatty, Link, and Driscoll. Acquisition of data: Carlson and Driscoll. Analysis and interpretation of data: Carlson, Beatty, and Neff. Drafting of the manuscript: Carlson, Neff, and Driscoll. Critical revision of the manuscript for intellectual content: Carlson, Beatty, Link, and Driscoll. Study supervision: Carlson, Beatty, Neff, Link, and Driscoll.

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REFERENCES

Correction

Error in Figure. In the Original Article titled “Educational and Employment Achievements in Prelingually Deaf Children Who Receive Cochlear Implants” by Venail et al, published in the April issue of the Archives (2010;136[4]:366-372), an error occurred in Figure 3 on page 370. In the figure’s key for part A, the bar color for children 16 to 18 years old should have been dark blue instead of light gray, and the bar color for children older than 18 years should have been medium blue instead of dark blue. The corrected figure is printed here with its legend.

**Figure 3.** Types of educational support, in addition to the cochlear implant, used by children without (n=74) (A) and with (n=26) (B) additional disabilities in this study. Data are presented according to the age of the children at the time of the survey. FM indicates frequency-modulated.