Nager Syndrome: An Update of Speech and Hearing Characteristics

Marion D. Meyerson, Ph.D.
Jean Boling Nisbet, M.Ed.

Nager acrofacial dysostosis is a rare syndrome of unknown etiology combining mandibular and thumb/radial hypoplasia. Seven patients evaluated in this study had histories of early respiratory and feeding problems, micrognathia and absent velum, atretic ear canals and conductive hearing loss, upper and lower limb malformations, normal intelligence, and speech/language delays and disorders. These findings, with few exceptions, were consistent with the findings in previously published and unpublished case histories of patients with Nager syndrome. Recommended rehabilitative strategies include prespeech feeding activities (especially if gastrostomy tubes are present), oral language stimulation, individualized speech/language therapy, and early audiologic evaluation and amplification.

In 1977, the early language, hearing, and speech development of two patients with Nager acrofacial dysostosis was reported (Meyerson et al., 1977). The diagnosis of this rare oromandibular hypogenesis syndrome was made on the basis of a combination of craniofacial features similar to those found in Treacher Collins mandibulofacial dysostosis plus thumb/radial hypoplasia. Since that time, several additional patients with Nager syndrome have been reported in the literature.

The etiology of Nager syndrome remains in question. A wide range of variability suggests genetic heterogeneity. Autosomal dominant inheritance with reduced penetrance has been suggested (Halal et al., 1983), but most cases have been sporadic, with no reports of affected parents. Autosomal recessive transmission has also been posited. Gorlin (1978), however, noted that if all cases of Nager syndrome were inherited on a recessive basis, there would be more affected siblings. A few reports of first trimester drug ingestion (Meyerson et al., 1977; Halal et al., 1983; Kawira et al., 1984) have not yielded a firm teratogenic explanation.

Patients with Nager syndrome have been reported in several countries and are of varying racial and ethnic backgrounds (Burton and Nadel, 1977; Matsumura et al., 1981; Wiedemann and Dibbern, 1982; Giugliana and Pereira, 1984; Kawira et al., 1984). The syndrome is rare and should be differentiated from other entities that it may resemble. Among these are Treacher Collins mandibulofacial dysostosis, which has no limb abnormalities and a high incidence of lower lid colobomas (Goodman and Gorlin, 1983); Pierre Robin sequence (Temtamy and McKusick, 1978); postaxial acrofacial dysostosis (Miller et al., 1979); Kelly syndrome (Kelly et al., 1977); and other variants (Carey et al., 1978; Halal et al., 1983; Richieri-Costa et al., 1983).

A number of infants with Nager syndrome have died shortly after birth because of respiratory complications (Gellis and Feingold, 1978; Halal et al., 1983; Pfeiffer and Stoess, 1983; Kawira et al., 1984). Many of the surviving children have required tracheostomies, tongue/lip suturing, gavage feeding, and gastrostomies because of the...
micrognathia and resultant airway compromise (Meyerson et al., 1977; Gorlin, 1978; Temtamy and McKusick, 1978).

The physical features of Nager syndrome generally noted at birth or in infancy are downslanted palpebral fissures, malar hypoplasia, high nasal bridge, atretic external auditory canals, severe micrognathia, and absent velum (Smith, 1982; Golabi et al., 1984). Preaxial limb malformations include hypoplastic or missing thumbs, hypoplastic radii, and shortened humerous bones. Halal et al. (1983) noted a correlation between the shortness of the arms and the lack of mandibular development. Several authors have reported poor growth and short stature in these patients (Gellis and Feingold, 1978; Goodman and Gorlin, 1983; Halal et al., 1983).

Most authors have found normal intelligence in patients with Nager syndrome (Gellis and Feingold, 1978; Smith, 1982; Meyerson, 1985). Goodman and Gorlin (1983) ascribed occasional reports of mild mental retardation to delays resulting from hearing loss. Halal and his colleagues (1983), however, described one patient as being severely mentally retarded.

Ear malformations in Nager syndrome are varied. Stenotic ear canals as well as preauricular tags, pinna and ossicular malformations, and lowset ears have been reported (Goodman and Gorlin, 1983; Halal et al., 1983). Hearing levels are similar to those measured for Treacher Collins syndrome, a conductive loss of 50 to 70 dB bilaterally (Phelps et al., 1981). Even if there are normal auricles and open external auditory canals, the ossicular chain is often malformed and a conductive hearing loss is present. Sensorineural hearing loss has not been associated with Nager syndrome, but it is an occasional finding in Treacher Collins syndrome (Phelps et al., 1981).

Receptive and expressive language delays marked the early developmental years of the patients reported by Meyerson and her colleagues (1977). Hearing loss, early failure to thrive, frequent hospitalizations, and a variety of health problems were contributing factors. Other authors also described language delays in their patients (Burton and Nadler, 1977; Lowry, 1977; Gellis and Feingold, 1978).

Respiratory and feeding problems in the infant with Nager syndrome require heroic measures that may well impede the normal development of speech skills. Tracheostomies preclude normal vocal play; tongue/lip sutures prevent normal sucking behavior; and gavage feeding and gastrostomy tubes do not allow for normal oral-motor development through prespeech feeding activities. Auditory stimulation and feedback for monitoring and learning are reduced because of hearing loss. Even mild conductive losses appear to slow the acquisition of oral and written language skills and reduce the clarity of speech (Webster, 1983).

In addition to the problems just mentioned, severe micrognathia; restricted jaw mobility; and frequently absent, cleft, or malformed velum are significant hazards to the production of good speech. The hypoplastic mandible leads to abnormal tongue posture. Malocclusion and crowding of the oral cavity may prevent normal articulation for speech. A missing velum must and a malformed velum may result in velopharyngeal insufficiency. The effect of these conditions may, however, be lessened because of tongue carriage, nasal pathway obstruction, and other structural abnormalities. Peterson-Falzone (1982), in describing the resonance problems in mandibulofacial dysostosis, which Nager syndrome closely resembles, noted that supralaryngeal vocal tract abnormality such as a hypoplastic pharynx and aberrant tongue carriage "muffles" the hypernasality resulting from velopharyngeal incompetence. The verbal output of children with Nager syndrome may not sound as hypernasal as would be expected from the palatal morphology but there could be misarticulations, abnormal compensatory adjustments, and an abnormal resonance balance. Each of these factors can reduce intelligibility.

The paucity of information in the literature regarding language, speech, and hearing problems associated with Nager syndrome prompted the following clinical study of seven patients. The children had all been diagnosed as having Nager acrofacial dysostosis. No patient presented a family history of the disorder. Medical, audiologic, and educational reports were obtained from the families, and the children were then given language and speech evaluations in their homes or in a clinical setting by the authors. The following are brief reports summarizing the case histories and clinical evaluations of the patients.

**CLINICAL REPORTS**

**Patient 1**

Patient 1 (Fig. 1) is a 20-month-old male infant who was born with mandibular and malar hypoplasia, absent velum, downslanted palpebral fissures, atretic auditory canals, high nasal bridge, absence of thumbs, and slight shortening of forearms with reduced range of motion at the elbows. A heart murmur and torticolis with possible right sternocleidomastoid hypertrophy were noted in infancy. An unsuccessful tongue tethering
FIGURE 1 Patient 1 at age 20 months.

in infancy was followed by a tracheostomy that was closed by 1 year of age. A gastrostomy performed at 3 weeks was also closed at 1 year. The patient currently wears a cervical collar for the torticollis. Auditory brainstem and behavioral audiometry suggested a mixed hearing loss of moderate degree in the low- and mid-frequency range and a severe loss in the high frequencies. In spite of atretic auditory canals, earmolds and behind-the-ear hearing aids resulted in an improvement of the speech awareness threshold from an unaided level of 75 dBHL to aided responses at 40 dBHL.

The Receptive-ExpressiveEmergent Language Scale (Bzoch and League, 1971), an interview instrument, was administered to the patient’s mother when he was 20 months old. His receptive language score was judged to be at the 14-month level. Because training in signing was begun when the patient was an infant, the authors evaluated expressive language in signs and in oral speech. Expressive language in signing was at the 20-month level. Several emerging language behaviors were demonstrated at higher levels than those at which he scored. He was reported to bring objects upon verbal request, to recognize names of pictures, to comprehend simple questions, and to carry out some two-stage directions. Some verb forms at the 20-month level were understood. His mother reported that he used at least 20 signs and combined them into simple sentences. He used consistent sound approximations for eight words. Little oral output was heard by the examiners except for attention-getting squeals. There was mild intermittent hypernasality.

Prognosis for language development was judged to be excellent and for oral speech good if training emphasized an aural/oral approach. The patient has been visited once weekly by a teacher from a center for the infant deaf. She reported an expressive sign vocabulary of 25 to 30 words with combinations such as “where doggie,” and “please milk.” The parents are considering future placement in an oral program.

Patient 2

Patient 2 (Fig. 2) is a 2½-year-old girl noted at birth to have severe micrognathia, malar hypoplasia, absent velum, downslanted palpebral fissures, atretic ear canals, missing thumbs, fused ulnae and radii, and shortened forearms. She also had dislocated hips and had worn a cast for several months. Tracheostomy and gastrostomy were performed in infancy and remain open to date. Auditory brainstem response audiometry indicated normal cochlear function. When patient 2 was 15 months of age, speech detection thresholds were obtained with behavioral audiometry at 65 dBHL bilaterally. Unaided soundfield warbled tone thresholds were 60 to 70 dBHL across the speech frequency range. A bone conduction hearing aid fitted at 6 months of age improved speech detection to 10 dBHL and warbled tone thresholds to 10 to 25 dBHL across the speech frequencies.

At chronologic age 28 months, a Receptive-Expressive Emergent Language Scale (Bzoch and League, 1971) administered to the patient’s mother indicated receptive language age at 27 months and

FIGURE 2 Patient 2 at age 2½ years.
expressive language age in signing at 20 months. Although the decoding of oral language appeared within normal limits, there were some gaps in naming smaller body parts and in understanding personal pronouns and complex sentences. She had at least 30 signs that she recognized and used and had been putting them together in two-sign combinations to represent concepts such as “Jason crying.” She appeared to understand both English and Spanish and was able to point correctly to 26 items on a nonstandardized preschool receptive bilingual language inventory. After pointing to the items, she correctly and spontaneously made signs for 15 of them. Considerable stoma noise was heard throughout the testing. She locomoted on her buttocks and could pull herself into an upright position.

Prognosis for language development is excellent and for speech development, fair. Eventual plugging of the stoma and release of the severely restricted mandible should improve the prognosis. She is currently visited regularly by a teacher of the infant deaf.

Patient 3

Patient 3 (Fig. 3) is a 4-year-old girl who at birth had malar and mandibular hypoplasia, downslanted palpebral fissures, absent velum, bilateral radial hypoplasia with small thumbs and decreased range of motion, small first toes bilaterally, and club feet that were casted for several months. Tracheostomy and gastrostomy were performed in infancy and eliminated before her second birthday. Some oral feeding was accomplished concurrent with the gastrostomy tube. The mandible was not restricted and some early mandibular growth was observed. Otologic history revealed recurrent otitis that was treated with bilateral myringotomies and tympanostomy tubes. At age 3 years, a moderate conductive loss was identified through play audiometry and a hearing aid with shell mold was recommended. A year later the otitis had cleared; however, a presumed ossicular malformation appeared to be responsible for the continued conductive loss bilaterally with poorer hearing in the lower frequencies. Binaural behind-the-ear amplification improved thresholds to 20 to 30 dB HL across the speech frequencies. A prosthetic obturator with a speech bulb was fitted at age 3 ½ years and was retained with wire clasps and orthodontic bands on deciduous molars. The bulb was enlarged at age 4 years but no improvement in resonance was observed.

On the Preschool Language Scale (Zimmerman et al., 1979) at chronologic age 4–1, patient 3 scored at the 4–11 level with a quotient of 120 in auditory comprehension and at a 4–5 level with a quotient of 108 in verbal ability. Her age equivalent on the Expressive One-Word-Picture Vocabulary Test (Gardner, 1979) was 4–8, which placed her in the sixty-sixth percentile. Although she had generally excellent language and cognitive skills, the patient made errors in receptive and expressive vocabulary at the 3 ½–to-4-year level. However, the patient could identify right from left, smaller body parts, and action-agent constructions, and she could count at levels appropriate for her age. Verbal ability testing yielded deficits in repeating function words and morphemes and in answering complex questions about remote events. Nevertheless, she did answer questions about the senses that are normed as typical for ages 4½ to 5 years. Word-finding and expressive vocabulary skills were good. Sentence length of 4 to 8 words was within normal limits for her age (Hedrick et al., 1975).

Patient 3 demonstrated variable hypernasality and nasal emission. She used many compensatory articulations such as mid-dorsum stops and glottal stops. Some glottal stop coarticulations and frequent final consonant omissions were observed. There was no difference in articulation with or without the obturator. Because of less oral crowding than in most cases of Nager syndrome, there was no muffled quality. Laryngeal function appeared normal with adequate loudness and pitch range.

Prognosis for language and speech is favorable because of the relatively mild expression of the syndrome and the patient’s excellent cognitive abilities. She will attend a regular preschool class with once weekly speech therapy with emphasis on articulatory placement, on closing syllables, and on the use of morphemes and function words. She has never used signing.

Patient 4

Patient 4 (Fig. 4) is a 5½-year-old girl who at birth had severe micrognathia and malformed temporomandibular joints, retroverted and low-set ears, hypoplastic outer and middle ears, fused elbow joints, severe humeral and radial hypoplasia with inward rotation of forearms, hypoplastic thumbs and toes, and missing fingers on the right hand. A bilateral club foot deformity was treated with casts. At 2 weeks of age she underwent a tracheostomy, which was closed at age 2½ years. She did not start using voice, however, until the age of 5 years. A gastrostomy was performed at 9 months of age and remains open to date. Tongue
retraction and jaw restrictions have prevented normal sucking and chewing. Strabismus was corrected surgically at age 2 years, and her vision is now normal. Bilateral temporomandibular joint release and reconstruction with an interdental splinting device were done at age 5 years. Visual response audiometry at 2 years of age yielded a speech awareness threshold of 70 dBHL. A bone conduction hearing aid was fitted, and she responded to voice at 30 dB and to warbled tones at 20 to 40 dBHL.

On the Preschool Language Scale (Zimmerman et al., 1979) at chronologic age 5–10, patient 4 responded at a 5–2 level with a quotient of 89 in auditory comprehension and at a 3–6 level with a quotient of 60 in verbal ability. Her age equivalent on the Expressive One-Word-Picture Vocabulary Test (Gardner, 1979) was 4–9, which placed her in the twenty-first percentile for her age. On the auditory comprehension portion, she passed all items at the 3- to 3½-year level and most vocabulary items at 3½ to 4 years. She passed some scattered items at higher levels but did not respond correctly to prepositional phrases, adjective items, and math problems. She passed no items at 6 to 7 years. Her mother reported that she used no voice or speech until age 5, although she had been signing. At present she speaks first and then signs if not understood. The basal age on the verbal test was 2 years with few correct consonants, inconsistent use of plural markers, and few answers to complex questions. Syntax was good. Average sentence length and variety of sentence types were low for her age.

Intelligibility was below 50 percent when context was unknown. This was attributed to consonant and vowel distortion. The patient used /n/ correctly and /d/ inconsistently but produced few other consonants, perhaps in part because of the severity of the micrognathia and oral crowding. Hypernasality and nasal emission were mild but resonance was muffled. The tongue tip appeared hypoplastic but was mobile. The voice was diplophonic and of low intensity.

In summary, the patient’s language delay may have been exacerbated by conductive hearing loss, inadequate prespeech feeding activity, and frequent hospitalizations. Significant gains in the last year suggest a better prognosis for improvement in language skills. Gains in speech intelligibility will probably require increased oral mobility, slower rate of speech, and careful placement with light contact of the articulators. She attends a special education class for the deaf and multihandicapped children and receives speech therapy once a week. Total communication is used in her class.

**Patient 5**

Patient 5 (Fig. 5) is a 5½-year-old girl who was noted at birth to have malar and mandibular hypoplasia, with an ankylosed temporomandibular joint, small mouth, absent velum, small ear canals, posteriorly rotated auricles, small tongue, and a preaxial limb defect with short thumbs and toes. An early attempt to release the ankylosed TM joint did not improve jaw mobility. Tracheostomy and gastrostomy performed in early infancy were eliminated just before age 5 years. Visual response audiometry showed a conductive hearing loss and an air-bone gap of 25 to 60 dBHL in the speech frequencies. A bone conduction hearing aid was not used until the patient was 5½ years old. Nonverbal psychometric testing at age 3½ suggested normal intelligence with many age-appropriate skills.

On the Preschool Language Scale (Zimmerman et al., 1979) at chronological age 5–8, the patient scored auditory comprehension and verbal ability levels of...
On the Expressive One-Word Picture Vocabulary Test (Gardner, 1979), the age-equivalent level was 2–2, which was below the first percentile. She failed some vocabulary items at lower levels, but could identify action words, match block patterns, recognize colors, and understand the numerical concept of three. She did not use plurals or answer complex questions. Responses in expressive language were scattered. Average sentence length was 4 to 5 words. Her voice was slightly high-pitched and of low intensity.

The speech intelligibility of the patient was low. She could pronounce /p, b, m/ and could approximate /t/. She exhibited hypernasality, nasal emission, and a number of compensatory articulations including glottal and midpalatal stops. Many final consonants were omitted. Speech output sounded muffled because of the restricted jaw mobility and small oral cavity.

The prognosis is thought to be good if intensive remediation is given. The patient lived abroad for part of the preschool period and was not in any educational program. With attendance at an oral communication class after her return, important gains were made. The use of amplification plus enrollment in a school program should help language skills in the future. If a total communication class is the only option available in her community, emphasis should be placed on oral speech.

Patient 6

Patient 6 (Fig. 6) is a 10½-year-old boy who was noted in infancy to have malar and mandibular hypoplasia with an ankylosed temporomandibular joint, broad nasal bridge, downslanted palpebral fissures, narrow ear canals, absent velum, and hypoplastic thumbs. A tracheostomy was performed in infancy and closed at 9 months. He was hospitalized until 2½ months of age because of severe respiratory and feeding problems. Release of the ankylosed temporomandibular joint was performed at 5 years of age. Nasal passages were narrow and the patient was and is a mouth breather. A conductive hearing loss was identified in infancy, and bone conduction amplification was used. At age 10, the presence of a moderate conductive loss was established using standard speech reception and pure tone thresholds. Speech discrimination was excellent. With his bone conduction aid in place, his thresholds were in the normal range.

Early language development was marked by mild receptive delays and significant expressive lags. At chronologic age 10–7, he scored an age equivalent of 9–9 on the Utah Test of Language Development (Mecham and Jones, 1978), placing him in the fifth percentile. His age equivalent on the Expressive One-Word Picture Vocabulary Test was 11–5, which put him at the sixty-sixth percentile. Word finding and vocabulary were his best language skills, but there were deficits in reading, auditory comprehension, and spelling. Successful test responses were scattered. On the Lindamood Auditory Conceptualization Test (Lindamood and Lindamood, 1971), he scored at a first-grade level with notable deficits in auditory tracking. (This test can be administered in spite of peripheral hearing loss.) Syntax in spontaneous speech and average sentence length were within normal limits.

Oropharyngeal crowding appeared responsible for the muffling of hypernasality. Some inconsistent cul-de-sac nasality alternated with nasal emission. Articulation was marked by mid-dorsum palatal stops for /t/ and /d/. Many of the phonemes could be produced correctly with auditory stimulation, but the patient’s average rate of speech was too fast to allow for careful self-monitoring. Habitual pitch seemed somewhat high for his age.

Patient 6 has been in special classes for the hearing impaired and for language-handicapped children since early childhood. Signing was taught with speech for a brief period, but is no longer used. Partial mainstreaming into a fourth-grade class is planned. Mandibular advancement and mobilization is planned for the near future and should help to improve sound production and general intelligibility, as well as appearance. Continued emphasis on reading mechanics and comprehension is warranted. Drooling can and should be controlled with nonverbal reminder signals.

Patient 7

Patient 7 (Fig. 7) is an 11-year-old girl who was noted at birth to have malar and mandibular hypoplasia, microstomia, moderate glossoptosis, downslanting palpebral fissures, atretic ear canals, aplastic thumbs, hypoplastic toes, and rotation deformities of the legs. Her legs were placed in casts in early infancy and a combination of tube and oral feeding was undertaken. She resided in a nursing home for the first 8 months of life and was given language stimulation by the hospital staff and visiting clinicians. Surgical release of the ankylosed mandible was attempted at age 3 years with only partial success. Shortly before her eleventh birthday, the release of bilateral temporomandibular joint ankylosis and mandibular advancement were accomplished, and dental splints were wired in place. A bone conduction body aid was fitted at 5 months of age after a moderate conductive loss was
determined through speech reception thresholds. Binaural bone conduction hearing aids brought thresholds to normal levels and yielded excellent speech discrimination scores. A psychometric evaluation when she was 9 years old indicated average cognitive and visual-motor skills as measured by two standardized scales.

The patient’s early years were marked by a mild receptive language deficit and significant expressive delays. On the Utah Test of Language Development (Mecham and Jones, 1978) at age 11, she scored two years below her chronologic age, placing her at the fourth percentile.

Before mandibular advancement, her severely restricted vocal tract resulted in “muffling” of inconsistent hypernasality and nasal emission. Intelligibility was fair with a slowed rate of speech. The velum was absent; however, cinefluorography revealed that during production of pressure consonants, the posterior portion of the tongue appeared to make contact with the posterior pharyngeal wall at the level of the atlas. Following surgery, a marked improvement was noted in the production of labiodental consonants but hypernasality increased noticeably.

Prognosis for improved speech intelligibility has improved following mandibular advancement surgery. After several years in classes for the hearing impaired, the patient has completed a regular fifth-grade class with marginal grades. The authors suspect that patient 7 has a learning disability based on central auditory processing deficits. With placement in regular classes she should continue with remedial reading, spelling, and other academic programs being provided by resource specialists.

**DISCUSSION**

Physical characteristics found in our seven subjects included hypoplasia of the mandible, malar bones, velum, external auditory canals, and thumb and radial bones (see Table 1). Most of these seven children had lower limb involvement including toe abnormalities, club foot, and hip dislocation. The two children in our study sample with the most serious limb malformations (patients 2 and 4) appeared to have the most severely hypoplastic mandibles. These physical characteristics were consistent with those described in the additional published reports cited earlier and in unpublished case reports of Nager syndrome contributed by colleagues.

Our seven patients and all additional contributed case histories described early feeding problems, conductive hearing loss, and subse-
TABLE 1 Selected Physical and Speech Characteristics of Seven Patients with Nager Acrofacial Dysostosis

<table>
<thead>
<tr>
<th>Data and Characteristics</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>1 2 3 4 5 6 7</td>
</tr>
<tr>
<td>Sex</td>
<td>M F F F M F</td>
</tr>
<tr>
<td>Severe micrognathia</td>
<td>+ + - + + +</td>
</tr>
<tr>
<td>Absent velum</td>
<td>+ + + + + +</td>
</tr>
<tr>
<td>Thumb/radial hypoplasia</td>
<td>+ + + + + +</td>
</tr>
<tr>
<td>Lower limb anomalies</td>
<td>- + + + + -</td>
</tr>
<tr>
<td>Atretic ear canals</td>
<td>+ + + + + +</td>
</tr>
<tr>
<td>Early tracheostomy</td>
<td>+ + + + + -</td>
</tr>
<tr>
<td>Early gavage feeding</td>
<td>+ + + + + +</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>+ + + + + +</td>
</tr>
<tr>
<td>Early receptive language delay</td>
<td>+ + - + + +</td>
</tr>
<tr>
<td>Poor speech intelligibility</td>
<td>+ + + + + +</td>
</tr>
</tbody>
</table>

+ = present
- = not present

quent speech delays and disorders. Most of the patients had been tracheostomized because of early respiratory distress resulting from severe micrognathia. Most were fed either by gastrostomy or nasogastric tubes for several months or years because of temporomandibular ankylosis and tiny mouth opening.

Since severe jaw restriction and open tracheostomies in some of the children precluded attempts at vocal play and early speech activities, sign language was taught as an expressive tool in spite of missing digits. The most mildly affected youngster in the study (patient 3) never used signs. All the children over the age of 5 years could vocalize and verbalize, and the use of signs decreased in favor of oral speech. Hearing loss, locomotor limitations because of upper and lower limb involvements, and frequent respiratory illness and multiple hospitalizations may have slowed the rate of early receptive language development. A few unpublished case reports indicated that at least three young adults with Nager syndrome had completed high school and had adequate speech with some problems in articulation and resonance.

No subject in the present sample had what could be called good speech in that resonance and articulation problems were immediately obvious. However, the five older patients communicated primarily with oral speech. In many, oropharyngeal crowding masked the expected hypernasality and altered the oral resonance, producing a muffled quality. The most mildly micrognathic child (patient 3) exhibited the hypernasality, nasal emission, and compensatory substitutions that are typical of velopharyngeal incompetence. Severe micrognathia in most of the children precluded the correct production of the bilabial and labiodental consonants.

Orthognathic surgery in early childhood and in the preteen years has achieved variable success in improved mandibular movement and better speech. Vargervik et al (1985) proposed a schedule for Nager syndrome patients beginning with early fluoride treatment and a sugar-free diet, release of mandibular ankylosis and subsequent orthodontia at age 3 or 4 years, and mandibular advancement at age 11 to 12 years.

Fortunately, most of the subjects in this study had access to early audiologic evaluation and amplification. Behavioral audiometry was combined in many cases with brainstem response and impedance audiometry. All the subjects had a moderate to severe conductive loss, and patient 1 had a mixed loss. Most of the children were fitted with bone conduction hearing aids, although patients 1 and 3 appeared to benefit from binaural behind-the-ear hearing aids with earmolds. Because bone-conducted sound is readily transmitted to both cochleas, some audiologists question the value of prescribing two instruments for a child. Others feel justified in trying dual receivers. The patients for whom binaural bone conduction aids have been fitted were said to have demonstrated small gains in localizing sound and in hearing in noisy environments. There were no reports of indentation of the mastoid caused by the pressure of the bone oscillator.

Otologic surgery has been undertaken to correct auricular and middle ear malformations in other craniofacial syndromes with varying success (Phelps et al, 1981). Temtamy and McKusick's (1978) Nager syndrome patient underwent external and middle ear surgery with "disappointing" results. Aside from routine myringotomies to treat chronic otitis media, no child in this study had otologic surgery. Cosmetic correction of ear deformities may be proposed when the children are older.

Although no serious emotional illness has been documented in any of the seven patients in this study, it is important to recognize the psychological impact of facial deformity and the trauma of surgery. Parental and patient counseling may be important adjuncts to habilitation. Genetic counseling should also be sought. No patient in the study had an affected parent or sibling. Until the mode of transmission becomes clear, most genetic counselors would give a recurrence risk figure of no more than 8 percent (Halal et al, 1983) to families of the patients.
REMEDIAL RECOMMENDATIONS

Analysis of diagnostic findings, therapy reports, and clinical insights has yielded a number of recommended intervention strategies for those involved in the language, speech, and hearing habilitation of children with Nager acrofacial dysostosis. Although nutritional needs may require gavage feeding, prespeech feeding training with maximum sucking and chewing experience should be encouraged. Oral language stimulation should be maximized from infancy. Signing should be incorporated with speech only if the mandibular restrictions and tracheostomies render early speech play impossible. Total communication, if continued through early childhood, should favor attempts at oral speech when possible and use signing as a backup in successful communication.

Speech therapy should begin early in life and should incorporate parents and siblings in the reinforcement of all attempts at expressive language. Oral and tongue mobility should be maximized within the limits of mandibular restriction. Drooling can be deconditioned with a behavior modification paradigm using a subtle reminder sign. Compensatory articulations must be judged individually. Those that are logical adaptations to an aberrant vocal tract and enhance speech intelligibility should be reinforced. Other compensations that are unnecessary and impede intelligibility should be avoided before they become habits. Slowed rate, articulatory placement, and maximum mandibular excursion are vital to increased intelligibility and warrant early modeling. In addition, prosodic variation, informative facial expression, and body language should be incorporated into pragmatic exchanges so as to maximize communication success.

Some authors have postulated that conductive hearing loss during the first few years of life, when the brain is maturing, may affect a child's eventual central processing of auditory input (Webster, 1983). The oldest patients in this sample, patients 6 and 7, show evidence of learning and language problems in spite of apparently normal intelligence. Some of the younger children exhibit language deficits that may be reflective of central auditory processing problems and future learning disabilities.

The children in this study tended to use simplified sentences. This may represent an unconscious but rational effort to aid others in understanding their poorly intelligible speech. Instead of focusing on increased sentence length, the goals for language should be enriched syntax and clarity of speech.

Lastly, early and expert audiologic evaluation and amplification can, in most cases, bring aided thresholds into a normal or close-to-normal range. The decisions regarding the use of one or two bone conduction hearing aids must be made on an individual basis with the child's desires and performance in mind. It is hoped that high-fidelity bone conduction devices, possibly but not necessarily implanted bone-anchored hearing aids (Tjellstrom and Hakansson, 1981), will replace the awkward and unattractive headbands, oscillators, and hearing aids that are in use at present.

Because most children with Nager syndrome have normal cognitive abilities, it is critical that their skills be highlighted. Rational, individualized, remedial approaches to their many problems must be developed.

Acknowledgement. The authors wish to acknowledge the contributions of Rosalie Goldberg, Patricia Lindamood, Susan Creighton, Cynthia Curry, Karin Vargervik, and our patients and their families.

References

Kelly TE, Cooke RJ, Kesler RW. Acrofacial dysostosis...


