Hemifacial Microsomia: Priorities and Sequence of Comprehensive Otologic Management

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Hemifacial microsomia is evidenced by the clinical spectrum of *microtia*, *mandibular deformities*, and *middle ear malformations*. This constellation of defects suggests that the most efficacious treatment will be multidisciplinary management, which includes the otolaryngologist, audiologist, plastic surgeon, and temporal bone radiologist. In the past, microtia—the most clinically apparent malformation—usually has been studied and treated with respect to the area of special interest of the investigator, whether from an otologic, reconstructive, or radiologic perspective. Newer management, however, utilizes the combined skills of the clinical and behavioral disciplines in the *treatment of the microtic auricle, associated middle ear pathology, and hearing loss.* Such multidisciplined approach, commencing during the first year of life, is deemed advisable to cope with the severe unilateral or bilateral hearing loss which may lead to potentially serious sensory deprivation and impede adequate language development in these children.

Introduction

The hallmark of hemifacial microsomia is the clinical spectrum of microtia, mandibular deformities, and middle ear malformations. This constellation of defects suggests the need for a multidisciplinary approach to diagnosis and treatment, and the object of this presentation is to recommend guidelines for such comprehensive management of the microtic auricle in relation to associated middle ear pathology and hearing loss. Correction of mandibular deformities, including technique of pinna reconstruction, will not be discussed, since their management is usually undertaken separately from that of otologic anomalies. Contemporary variations in treatment of the microtic auricle will be considered in relation to various pathologic findings.

Microtia—usually the most clinically apparent malformation—often is selected for independent treatment according to the special interests of individual investigators (Caldarelli, 1977; Gill, 1969; Grabb, 1965; Jafek et al., 1975; Marx, 1926; Meurman, 1957; and Petasnick, 1973). However, hearing loss associated with hemifacial microsomia may result in auditory deprivation so severe as to retard language development. Therefore, the management of microtic auricle, associated middle-ear pathology, and hearing loss should commence during the first year of life and involve the multidisciplinary resources of the otolaryngologist, the audiologist, the plastic surgeon, and the temporal bone radiologist.

Prudent clinical management of this syndrome presupposes a knowledge of patterns of occurence and interrelationships of the deformed structures. Investigation of 70 patients with hemifacial microsomia was undertaken by various specialty services to map the patterns encountered and establish the interrelationship among characteristic facial malformations, leading to a rationale for treatment (Caldarelli et al, in press).

In this study, the external ear deformity

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(microtia) was graded as originally described by Maurman (1957). The grading system is as follows: Grade I—distinctly malformed auricles with smaller size than a normal outer ear, showing most of the characteristic features of the auricle; Grade II—the rudimentary auricle consists of a low, oblong elevation, hook formed at the cranial end corresponding to the helix; and Grade III—the auricle is more defective, showing only a part, often a malformed lobule, while the rest of the pinna is totally absent. Temporal bone deformities were radiographically assessed by polytomography, and middle-ear anomalies and external auditory canal deformities were correlated separately with grades of microtia and mandibular deformity. Findings showed that the severity of middle-ear anomaly and external auditory canal malformation significantly paralleled severity of both microtia and mandibular deformity. Microtia in mandibulofacial dysostosis did not correspond to a similar correlation of the microtic auricle, middle ear deformity, and mandibular deformity (Hutchinson and Caldarelli, 1977). This anatomic disparity suggests that microtia may be syndrome specific.

Further support for this concept was suggested in Poswillo's animal models of hemifacial microsomia and mandibulofacial dysostosis (Poswillo, 1975). He postulated that the etiopathogenesis differed between these syndromes, despite a similarity of certain anomalies.

Audiometric Evaluation

Optimal auditory habilitation in children with hemifacial microsomia often depends on accurate audiometric assessment within two years after birth. If hearing loss is unattended during this period, serious sensory deprivation and language lag may ensue, with significant educational consequences (Webster and Downs, 1978). Ideally, each patient requires (1) periodic hearing assessment, (2) placement in an habilitative program, (3) parent counseling, and (4) early amplification in the bilateral microtias.

Hearing assessment of patients should be sequential. In the neonate and young infant, auditory brain stem potentials (BSER) and the impedance test battery are included in traditional behavioral assessment techniques. The neonate should show standard behavioral responses to suprathreshold auditory stimulation. If responses are clearly identifiable, the infant usually has sufficient auditory acuity for language development. If such responses are not initially elicited, a hearing loss should be assumed, and subsequent testing should be performed for degree and type of loss.

In neonates exhibiting disorders associated with middle-ear anomalies, the assumption should be modified to presume that there is sufficient acuity to initiate language development, but such children should have further testing at six to seven months of age using a conditioned-response procedure. The visual reinforcement-audiometric technique (Morre, et al., 1976) is effective for threshold assessment. Between initial evaluation and subsequent attainment of auditory thresholds, monthly contact with the parents is also essential to monitoring the child's language development. Further auditory assessment is indicated if retardation is apparent.

An initial gross test for behavioral changes can be performed using a bone-conduction transducer. If this test evokes behavioral responses, one must then determine if the response is due to auditory or tactile stimulation. Auditory-evoked brainstem potentials may resolve this doubt. The BSER test measures the integrity of the lower auditory pathway from the eighth nerve to approximately the level of the inferior colliculus. The sensory specificity of this test may give a measure of cochlear function regardless of tactile input. Calibration and electrical artifacts currently obscure the interpretation of test results from the bone-conduction transducer. In addition, the observed normal response confuses threshold data in the maturing infant until approximately 18 months of age. For the above reasons, the BSER test should not be used to determine threshold information in these children. However, when used in conjunction with behavioral testing, it may show the presence or absence of functional cochlear activity.

When a tympanic membrane can be visualized in infants exhibiting hemifacial microsomia, the impedance test battery technique is particularly useful in comparing the nonaffected ear with the affected ear in those with

microtia, an external ear canal, and a tympanic membrane. This test battery, through use of the ipsilaterally evoked stapedial reflex, provides early information on the integrity of the middle-ear transduction system and on auditory acuity.

Each technique has inherent advantages and limitations. Both should be employed so that accurate assessment of auditory acuity can be made at the earliest possible age. In addition to comprehensive audiometric assessment, serial testing is helpful in monitoring the constancy or change in the child's audiometric profile with age.

As noted before, the prime objective of early assessment is to prevent sensory deprivation and consequent language delay. If the auditory deficit proves to be minimal, the infant and parents would be enrolled in a habilitative language program. When hearing loss is moderate to severe, amplification should be resorted to in addition.

Most children with hemifacial microsomia exhibit purely conductive hearing loss, which reduces loudness but does not distort intelligibility. The limited frequency and poor tonal characteristics of a bone-conduction hearing aid do not adequately compensate for reduced clarity. Therefore, the speech-language pathologist should supplement amplification with speech and language training. Despite the shortcomings of bone-conduction hearing aids, they provide enough auditory amplification to preclude major delays in language development.

In microtia without an external auditory canal, a bone-conduction hearing aid with a headband transducer is utilized. However, because of the infant's malleable cranial structure, an improperly fitted headband may conceivably injure the scalp and neurocranium. In the older child, a bone-conduction headband often irritates the scalp and may result in localized allopecia. This may be prevented by padding the headband or by altering its position. When major craniofacial surgery is contemplated, careful preoperative planning of the incisions in order to avoid the area of the headband may avert delayed wound healing and permit less interruption of the child's learning through amplification. Selection of an appropriate hearing aid for a child with microtia should follow acceptable contemporary audiologic practices and present few difficulties.

Otologic Management of the Unilateral Microtia

In the unilateral microtia, both otologic assessment and parental counseling regarding care of the nonaffected ear are important. Susceptibility to the usual spectrum of childhood otopathology may result in middle-ear damage and hearing loss with overall auditory handicap. The eustachian tube of the affected ear being patent, this ear also is subject to the usual childhood middle-ear infections. However, a greater risk to the child's health may be acute otitis media resulting from accumulation of pus under pressure in the middle ear. The absence of a tympanic membrane precludes rupture and drainage to the outside. In this case, it is conceivable that infection may involve the labyrinth, the facial nerve, or the brain. In such situations, the sole evidence of infection may lie in careful temporal bone tomography.

Parental Counseling

Parental counseling by the otologist and the audiologist during the first two or three years of life is important. To keep abreast of the child's hearing status, careful and continued otologic surveillance and periodic audiometry throughout the early years are imperative. These children usually exhibit normal but slightly delayed patterns of language development.

Radiographic Assessment

Temporal bone tomograms usually are performed when the child is between the ages of three and five. In certain children, sedation may be required to insure accurate examination. Conventional radiography has limited value in congenital malformations of the temporal bone except to demonstrate the extent of development of the mastoid or, as previously mentioned, to detect acute otitis media in a microtic ear. Temporal bone tomography is performed in at least two projections, frontal and lateral, with one- to two-millimeter sections. In selected situations, special oblique projections may be required.

A proper tomographic study can provide

the otologist with information about the degree of tympanic-bone abnormality, which may range from a minor deformity to complete agenesis, and about the position of the sigmoid sinus, the middle fossue dura, the juglar bulb, the mandibular condyle, and the course of the facial nerve.

Congenital cholesteatoma has been reported medial to the atretic plate. In some patients, the facial nerve may exit directly laterally in the mastoid bone immediately below the microtic auricle. Knowledge of this anomaly is important to the otologist in planning reconstruction of the external auditory canal and the middle ear, since it places the facial nerve in a position of vulnerability during auricular reconstruction. Following a review of temporal tomography, the parents are counseled on the potential for reconstructive surgery. Our current policy in patients with unilateral microtia and one normal-hearing, nonaffected ear is to defer middle-ear surgery until adulthood. Based on our own experiences and the reports of others, we note that approximately 50% of patients will improve enough to have binaural hearing following surgery. Deferring surgical decision until adulthood allows for consideration of the patient's educational or employment needs. In certain cases where the external auditory canal and tympanic membrane are reconstructed but the ossicular chain reconstruction fails to provide hearing improvement, we have utilized air-conduction hearing aids with specially designed molds.

Auricular Reconstruction

Since the initial parental concern is for the deformed auricle rather than the degree of hearing loss, parental counseling by the plastic surgeon as to the timing of the auricular reconstruction is important. Initially the parents are informed that to achieve ideal results, the timing of auricular reconstruction and middle-ear reconstruction must occur in proper sequence. Sequential management may vary according to the severity of the auricular deformity and the experience and reconstructive technique of the plastic surgeon.

Auricular reconstruction may often precede middle-ear reconstructive surgery. However, at the time of auricular reconstruction, the otologic surgeon may place a metallic marker over the mastoid in the area selected for a prospective external auditory canal. This will allow the plastic surgeon to reconstruct the auricle in a position most appropriate for future external auditory canal reconstruction. In unilateral microtia, otologic surgery usually may be delayed until adulthood, as stated earlier. However, the severity of middle-ear deformity in patients with grade II or III microtic auricles usually precludes adequate hearing restoration merely with middle-ear surgery. In patients in whom reconstructive middle-ear surgery does not achieve hearing improvement but in whom the external auditory canal remains patent and the tympanic membrane intact, air-conduction hearing aids with custom-designed molds may be required.

Grade I Microtia with an External Auditory Canal

Within the graded microtia classification, patients with bifid tragus, preauricular tags, and cupped, tilted, or low-set auricles may have less severe anatomic middle-ear and external auditory canal deformities than the grade II and III microtias. Typically, the external canal is narrowed, the tympanic membrane is underdeveloped, and the ossicles have subtle malformations resulting in deficient ossicular mobility or continuity. The degree of hearing loss in these patients often equals that for the grades II and III microtic ears. Elective reconstructive middle-ear surgery usually is performed after five years of age, and the potential for significant hearing improvement is good.

Patients with Bilateral Microtia

This group presents the most difficult problems in management. Children with bilateral microtia and absent external auditory canals have severe bilateral conductive hearing losses necessitating early audiometric evaluation and habilitation. The audiologist should work closely with parents to ascertain if the hearing aids are working properly and also to re-evaluate the child's hearing status and the effectiveness of the hearing aids in providing the maximal auditory input for language development. In certain situations, it may be beneficial for a speech-language pathologist to assess the child's language development and

work closely with the audiologist and parents to offer additional guidance in audiometric habilitation. Again, as in the unilateral case, the otologist should counsel the parents and the pediatrician regarding possible acute middle-ear infections and potential dangers to the child's health. Conventional temporal bone tomography using sedation may be necessary to assess the possibility of acute otitis media. Temporal bone tomography is performed under sedation during the third or fourth year of life. The otologist, on reviewing the tomograms with the radiologist, determines the potential for hearing improvement with middle-ear surgery. If surgery is indicated, it may be performed as early as four years of age. Auricular reconstruction in patients with bilateral microtia is sequenced as in the unilateral microtia. If reconstructive middle-ear surgery does not provide any hearing improvement and if the external auditory canal remains patent and the tympanic membrane intact, an air-conduction hearing aid with a specially designed mold may be utilized. Contralateral middle-ear surgery can be performed electively if there is a hearing improvement in the repaired ear.

Summary

The multidisciplinary nature of effective clinical management of the microtic auricle with associated temporal bone anomalies and hearing loss has been presented. Such a comprehensive approach appears to provide the best potential for sensory habilitation and cosmetic reconstruction in patients with hemifacial microsomia.

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