Congenital and Acquired Deafness in Clefting and Craniofacial Syndromes

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A study of 284 *craniofacial* defect patients determined factors associated with or predisposing to *acquired* or *congenital hearing loss*. Complete *otolaryngic* and *audiologic* evaluation was done at the initial assessment of a large group of patients. From there, a smaller number who had had adequate workup and followup was selected. The patients were classified into seven *clefting* categories, *microtia*/atresia, *facial* defects, *cranial* defects and miscellaneous anomalies of the head and neck. X-ray findings, those present at middle ear surgery and those studied at autopsy, are summarized. The *incidence* of hearing loss is 88 per cent. The likelihood of congenital hearing loss increases with the number of defects. Certain unifying concepts that may help identify a specific child as being in urgent need of otologic and audiologic testing are presented.

Introduction

The infant who has craniofacial defects presents physician and parents with many concerns regarding his ultimate function. Craniofacial defects have a high potential for involving adjacent sensory organs. The clinician needs to know which patients may benefit from early evaluation and which evaluations have high yield. He needs reasonable guidelines for justifying each test and consultation. Available references do not address this concern. An experience with 284 selected patients is presented with the goal of helping the non-otologist know when to call in the otologist.

The normal child acquires language through a gradual coordination of sensory input and central processing. This, of course, is a simplification of a complex process, which is far from being well understood. For the first few months of life, the process seems to be somewhat independent of environmental input in that the child vocalizes and babbles spontaneously. However, during this time, the hearing child is surrounded by sounds to which he may be alerted reflexively in the late prenatal and neonatal periods but to which he eventually begins to attend, then to try to imitate and to integrate with other sensory input. Finally, he is able to initiate his own productions and to use them to communicate. Basic concepts appear to be well established by about three years of age, although language continues to be enriched and polished throughout life (Northern and Downs, 1974).

Disruptions of this process occur in the child who has frequent, perhaps prolonged, bouts of partial hearing loss due to otitis media of early onset. Studies have demonstrated significant gaps between performance and verbal items on intelligence tests (Northern, Downs, 1974) Defects in language acquisition faced by the child who has congenital bilateral hearing loss are usually greater, although proportional to the degree of loss. The clearest example is afforded by the profoundly deaf child who hears almost nothing without amplification. Reflex startle responses occur only at very high sound intensities presented close to the young deaf baby. However, those who are otherwise normal are so visually alert that they may fool the parents into thinking that hearing is normal until the baby fails to take the next steps of imitation and initiation of language. The handicapping effects of various degrees of childhood hearing loss are shown in Table 1. A profoundly deafened child to whom the world of sound and language has not been opened during the first two to three years of life will have a difficult if not insurmountable barrier to language

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acquisition through the auditory route. Other areas of language acquisition and function may also be affected also (Sterritt, Camp, 1966). The problems of the child bereft of vision and hearing, whose central nervous system and or intellect are impaired as part of a craniofacial defect syndrome, or who has defects of the peripheral speech mechanism are very complex.

Ideally, a hearing handicapped child should be identified and helped in the first months of life, but this ideal is not always realized. The profoundly deaf infant waits in a soundless environment an average of 21 months after birth before his loss is detected (Bergstrom, Hemenway, 1971a).

Materials and Methods

Virtually all patients with craniofacial defects were referred for otologic consultation as part of their routine initial evaluation. All underwent general otolaryngologic examination, including, in most instances, viewing their ears under a Zeiss operating microscope; audiometry, including, where possible, puretone and speech testing; and acoustic bridge testing using either the Madsen ZO 70 electroacoustic unit or the Grason-Stadler, Model 1720, Otoadmittance meter. Where appropriate, plain mastoid films and petrous pyramid polytomography were obtained. From this larger group of patients 284 were selected whose workup and followup were judged adequate. Their ages ranged from a few days of age to 50 years at the first encounter.

Findings

The patients were divided into 11 subgroups of which seven were cleft palate subgroups, three of these syndromal. One, the "cleft palate plus," group was comprised of patients with clefts and other defects which could not be classified as representing any particular syndrome. Four additional groups were classified according to locus of the anomaly. Eighty-eight per cent of the patients had hearing losses, 152 as the result of otitis media or microtia/atresia, lesions readily discernible by physical examination (Table 2). Of the remaining 98 patients, nearly two-thirds had sensorineural losses or subtle defects of the conducting system requiring otologic and audiologic examination for confirmation. Furthermore, eight patients had serous otitis media superimposed on congenital ear lesions; seven of the microtia/atresia patients had hearing losses in the normal appearing ear or superimposed sensorineural loss in microtic/atretic ears.

There is no room for complacence in looking at the records of patients whose hearing losses were due at least in part to otitis media. There was an average delay of 3½ years in diagnosing serous otitis media, reported present at birth in nearly 100 per cent of cleft palate patients (Paradise and Bluestone, 1969). In only five patients was serous otitis media diagnosed at birth; three of those patients had palate defects. A 36-year-old patient with submucous cleft palate denied previous knowledge of ear disease, but a cholesteatoma was discovered in one ear. In 11 additional patients ranging in age from five to 27 years, evidence of chronic otitis media was found with no suspicion on the part of patient or parents that otitis media had ever occurred. Nine of the 11 had had regular pediatric care, tonsillectomies, hospitalizations, or multiple examinations by specialists involved in care of their defects.

There were definite delays in diagnosing bilateral congenital hearing loss in many other patients in this series, delays ranging from 12 to 288 months for conductive lesions

TABLE 1. Handicapping effects of hearing loss

degree of loss	*what can be heard without amplification*
Slight (15-25 dB-e.g. serous otitis, perforation, sensorineural loss (SNHL)	All vowels. May miss unvoiced consonants
Mild (25-40 dB-same pathology as above)	Some of speech sounds, louder voiced sounds
Moderate (40-65 dB-e.g. chronic otitis media, middle ear anomaly, SNHL)	Little of speech sounds at normal conversational level
Severe (65-95 dB-e.g. SNHL or mixed HL)	No speech sounds of normal conversation
Profound (95 dB-e.g. SNHL or mixed HL)	No speech, no other sounds

Courtesy of Marion Downs, University of Colorado Medical School.

TABLE 2. Hearing loss in craniofacial defects

no. of pts.	*types of defect*	*hearing loss*			
		conductive		*SN*	*mixed*
		congen.	*acq.*		
24	Cleft lip & palate		16	3	
51	Cleft palate		46		
21	"Cleft palate plus"*	4	18	5(1)	1
14	Pierre Robin	2	8	1	
4	Treacher Collins	4			
7	Trisomy 13	7			
60	Submucous cleft	7	16	9(1)	6(2)
36	Microtia/atresia	36(3)		1(4)	6(5)
33	Facial defects	8	7	14	3
14	Cranial defects	5	2	5	1
20	Misc. head & neck defects	5	1	6	3
284		78	114	44	20

(1) 3 had OM also (2) 5 had OM (3) 5 had congen. conductive loss in contralat. ear (4) in contralat. ear (5) 5 had superimposed SNHL in atretic ears, 1 in contralat. ear.

* The otologic findings are complex. One patient who had a congenital conductive loss had SNHL in the opposite ear, and 3 had OM superimposed on congenital middle ear defects.

and from 12 to 156 months for sensorineural or mixed deficits.

Analysis within subgroups and illustrative case histories

I. Clefts and Cleft Syndromes

A. In cleft palate, whether occurring alone or in combination with cleft lip, nearly all hearing loss was due to otitis media detected in the early months of life (Table 2). In several notable exceptions, diagnosis was not made until the early school years. Young infants are difficult to examine, but gentle, expert immobilization, ear cleaning, pneumatic otoscopy, and use of the microscope for examination minimize such difficulties.

Inadequate examination or lack of awareness may have accounted for some delays in diagnosis of otitis media in these patients.

Case Report. A five-year-old boy failed school hearing tests. Followup after cleft lip and palate repair had not included screening for ear disease. Initial otologic examinations were all but impossible because of the child's terror of doctors, but eventually good rapport was obtained. Bilateral serous otitis and an audiometric speech reception threshold of 34 dB were found. Repeated myringotomy and grommet tube placement were performed over the ensuing six years to maintain ventilation of the middle ear. School progress has been good.

The literature suggests that teenage and adult cleft palate patients outgrow their tendency toward middle ear disease (Moller, 1975). However, six older children and adolescents had chronic otitis media requiring tympanoplasty or mastoidectomy. Continuing vigilance is perhaps indicated. The sensorineural hearing losses found in this group were apparently unrelated to the cleft, but a specific cause was never found. Apparently no postmortem examination of the ears of patients suffering from cleft palate as their only anomaly have been reported.

B. Complicated Cleft Palate Entities

As might be expected, cases of cleft palate complicated by other anomalies tended to show a higher incidence of congenital hearing loss. Major associated anomalies included cleft lip, abnormal pinnae and external canals, microcephaly, craniostenoses, micrognathia, posterior choanal atresia, hemifacial microsomia, limb anomalies, central nervous system, cardiac, renal, chromosomal and ocular defects.

Case Report. A four-month-old male infant had persistent serous otitis media for which he underwent bilateral myringotomies and placement of tubes. He had cleft palate, facial tags, branchial defect, deformed right pinna, multiple bony anomalies of cervical and upper thoracic vertebrae, and intravenous pyelogram showed congenitally malrotated kidneys. At two years, audiometric test-

ing showed no response in the right ear. Petrous pyramid polytomograms revealed right middle ear anomalies, a bony plate occluding the right oval window and virtual absence of the right internal auditory canal. A hearing aid was fitted to the good ear because further surgery on his only hearing ear was deemed risky and because persistent serous otitis media was likely to impair speech and language development.

C. Cleft Palate Syndromes

Infants with Treacher Collins' mandibulofacial dysostosis may be grotesque in appearance, but many have normal intellectual capacity and generally normal internal architecture. However, renal and cardiac anomalies have been described (Hilson, 1957; Maran, 1964). Their hearing losses respond well to amplification. Surgery may not be feasible because of severe defects of middle ear structure.

Pierre Robin patients have a smaller incidence of congenital hearing loss but more variety in the types and degree of loss. In neonatal life, their very survival may depend on tracheostomy. In a few instances, children have somehow survived both respiratory and feeding difficulties.

CASE REPORT. At three months of age a cachetic female infant was admitted. Her mother, harrassed by poverty, a large family, and poor health, reported that the child took only a small amount of formula with great effort. The child had a large palatal cleft, micrognathia, glossoptosis and serous otitis media, and she seemed, even in skilled nursing hands, barely able to swallow. Development was grossly retarded, and she was placed in a state home for the retarded where she became the pet of the staff. She gained weight, then the ability to swallow, then began to catch up developmentally. Her efforts to talk intelligibly were impeded by her unrepaired palate. Because it appeared that she had more potential than was originally realized, the palate was repaired and unremitting serous otitis media treated by myringotomy and tubes. She was placed in a foster home and, when last seen at the age of six, was progressing in speech and language.

At the present time consideration for long-term care of trisomy-13 patients is not practical because of their lethal anomalies. Nevertheless, these mistakes in embryogenesis are of research interest, at least as far as the development of the ear is concerned. A recent autopsy study of 14 ears, the largest series accumulated to date, showed remarkable consistency in the pathologic findings, which included middle ear anomalies, severe primitive dysgenesis of inner ear vestibular organs but only minor shortening of the cochlea, and normal Corti's organ (Sando, Lieberman, 1975).

It is of interest that these ear anomalies bear some general resemblances to those found in Treacher Collins and in Pierre Robin. Histopathologic examination of the ears in a Treacher Collins patient showed bilateral external auditory atresia, absence of malleus and incus, anomalous stapes, and severely aberrant course of the facial nerve. The vestibular structures were abnormal in one inner ear. However, the cochlea and neural structures were normal. The author has examined postmortem temporal bone specimens from three Pierre Robin neonates who died of hypoxic complications or of associated defects. The inner and middle ears of one were normal; those of another showed questionable neural atrophy in the middle cochlear coil of one ear. The third showed an anomalous stapes and crowding of malleus and incus in the epitympanum. The cochlea was destroyed in removal and could not be evaluated (Bergstrom, Hemenway, 1968). Similar findings have recently been reported by Igarashi et al., who also noted minor anomalies of the vestibular portion of the inner ear and minor partition defects of the cochlea (Igarashi, Filippone, 1976). Sensorineural loss has been reported, but the pathology is unknown.

In summary, these three unrelated syndromes, similar only in having cleft palate, have findings in the ear which bear certain similarities: 1) gross middle ear anomalies; 2) anomalies of the vestibular compartment of the inner ear; 3) normal cochleas or ones containing minor anomalies of questionable significance.

D. Minor Clefts

Submucous cleft of the palate is generally asymptomatic in terms of nasal regurgitation of liquids or poor speech. However, it should be emphasized that 40 per cent of these patients have chronic middle ear disease (Bergstrom, Hemenway, 1971 b). One etiology may be poor mechanical stabilization of the tensor

and levator veli palatini muscles whose combined action is necessary to open the Eustachian tube. Congenital hearing loss occurred in 13 patients, five of whom had submucous cleft as their only other anomaly.

CASE REPORT. A three-year-old boy was referred by a speech pathologist because of tongue thrust, poor articulation, and mouth breathing. Oral examination was difficult because the patient strenuously resisted objects placed there. However, he readily cooperated for ear cleaning, and serous otitis media was found. Audiometry showed bilateral 45 dB conductive hearing loss which resolved after myringotomy and tube placement. Submucous cleft palate was detected while the patient was under general anesthesia prior to concomitant adenoidectomy which was cancelled pending further assessment.

In our group of 60 patients, 11 had had prior adenoidectomy or adenotonsillectomy, five without improvement of middle ear disease after surgery. One patient's pre-existing speech difficulty became worse postoperatively.

II. MICROTIA AND AATRESIA

The most striking finding in 36 microtia/atresia patients was the incidence of superimposed hearing loss in the atretic ear or the finding of unsuspected congenital sensorineural, conductive, or mixed hearing loss in the contralateral, normal-appearing ear. Twelve patients had bilateral microtia/atresia. Two had concomitant renal defects, one ipsilateral to a unilateral ear defect, the other affecting both kidneys in a patient who had bilateral microtia/atresia. Neither patient had facial asymmetry or hypoplasia.

At surgery or autopsy, reported ear findings vary considerably, ranging from the favorable, although rather infrequent, situation in which only a bony or fibrous plate is found, but tympanic membrane and middle ear are present and normal. Occasionally cholesteatoma forms deep to an atresia plate where tympanic membrane is present. More often the findings include mildly to grossly deformed or fused ossicles, absent ossicles, hypoplastic middle ear space, misplaced facial nerve. In more severe cases surgery has to be abandoned. X-ray studies should be done prior to reconstructive pinna or middle ear surgery to delineate the anatomic status of the middle and inner ear and the probable course of the facial nerve. Occasional facial nerve injury occurs during surgery.

Tuning fork tests and audiometry pose special challenges in these patients and need to be done carefully and correlated with other findings to assure that surgery is not performed on a dead ear or on the only hearing ear.

III. FACIAL DEFECTS

The incidence of acquired ear disease was low in this group of patients, but the proportion having congenital hearing loss was rather high. Several had potentially correctible middle ear defects. Three cases of auditory atresia were included because facial and other defects were detected long before deep-seated atresia of the external canal was recognized. Two of these children were referred for presumed mental retardation which disappeared after they had used hearing aids for some time.

Autopsy on one patient believed to have Goldenhar syndrome showed a hypoplastic oval window, deformed stapes, and slight shortening of the cochlea.

IV. CRANIAL DEFECTS

There were only 14 cases of primary cranial defects in this series, five of Crouzon's and four of Apert's. However, eight other patients had cranial defects, at least one of whom required surgery. Two Apert's patients have undergone middle ear surgery. In one, a widely patent cochlear aqueduct with gush of perilymph and cerebrospinal fluid aborted attempted surgical correction of a congenitally fixed stapes footplate (Bergstrom, Neblett, 1972). In a second patient, stapedectomy and prosthetic replacement have been successful. Congenital stapes fixation has also been reported at autopsy on an infant affected with Apert's (Lindsay, Black, 1975).

V. MISCELLANEOUS HEAD AND NECK DEFECTS

This potpourri of patients usually presented multiple defects. Except for three patients who had lethal chromosomal aberrations, there were few defects below the clavicle in this group. Although all but two were children, the incidence of otitis media was low. Fifteen of 21 had congenital hearing losses or

autopsy lesions compatible with congenital hearing loss.

Four patients in this group have died. The temporal bones of two trisomy-18 patients, one previously published, (Sando, Bergstrom, 1970) showed abnormal external ears and ossicles, especially the stapes, and aberrant course of the facial nerve. A neonate who died of the complications of partial deletion of chromosome 13 had minor middle ear anomalies and dysplasia of cochlea and saccule which likely would have produced sensorineural deafness (Bergstrom, Hemenway, 1972). A young man who died in a traffic accident had well-documented profound bilateral congenital deafness. He had micrognathia, posterior choanal atresia, shallow pharynx, and hooked nose. At autopsy, one temporal bone appeared to be completely normal; the other showed organ of Corti atrophy.

Summary of Case Findings

Some associations between the presence of certain defects and concomitant hearing loss can be made. Congenital hearing losses occurred in 87 per cent of patients having ocular defects, in 67 per cent of those having microcephaly, in 60 per cent of those having cranial nerve palsies, and in 50 per cent of those having central nervous system defects. Many of the losses were sensorineural. This makes sense when one recalls that the otocyst, from which the inner ear is derived, the optic vesicle, and the central nervous system are all derived from primitive ectoderm. Congenital cranial nerve palsies were divided between those associated with central nervous system defects and those of the facial nerve where extensive defects of branchial origin coexisted. Micrognathia had a 70 per cent associated incidence of hearing loss, all but two of which were conductive in nature. Where pinna defects other than microtia/atresia occurred, 75 per cent of those affected had congenital hearing loss and nearly two-thirds of the 75 per cent were conductive in nature, as demonstrated by audiometry, petrous pyramid polytomography, surgery, or autopsy.

Palatal, facial, vertebral, and skull anomalies and posterior choanal atresia were associated with congenital hearing loss in 90 per cent, 81 per cent, 100 per cent, 65 per cent, and 77 per cent of the cases respectively. Defects in these areas may be conceptualized as disorders of embryologic mesodermal growth or organization. Defects of the heart, another mesodermal derivative, were associated with congenital hearing loss in 72 per cent of the cases. 83 per cent of patients having renal defects also had congenital hearing loss. However, our population of microtia/atresia patients showed only a five per cent incidence of renal anomalies. Perhaps, in the one instance, the high correlation of renal and ear defects may be seen as a fault in mesoderm formation since the ear defects in most instances involved bony structures related to otic capsule development. In microtia, the low correlation may reflect only coincidental branchial and renal defects.

There is one other observation of interest. The likelihood of deafness rises rather steeply as the number of defects rises, as can be seen in Table 3.

Other Considerations

Factors believed to be extraneous to the craniofacial defect must be remembered when an individual child is evaluated. These include intrauterine infections such as rubella, cytomegalovirus, herpes virus type II, and perhaps toxoplasmosis and viral infections

TABLE 3

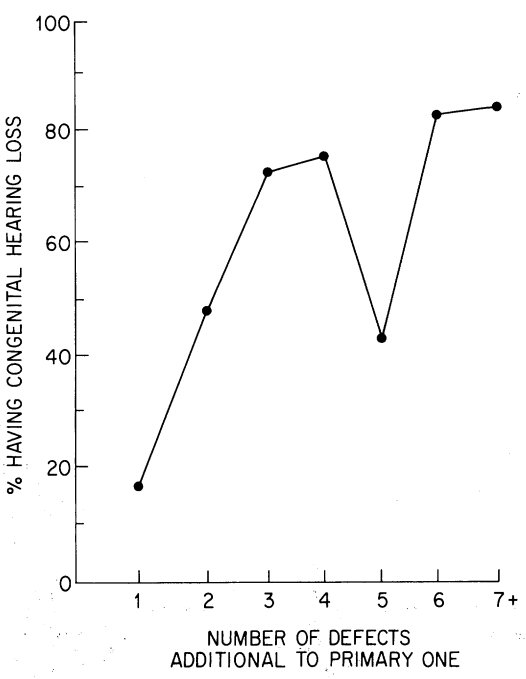

TABLE 4. Craniofacial syndromes having hearing loss*

Sensorineural	
Marshall syndrome	General cortical hyperostosis
Stickler syndrome	Cleidocranial dysostosis
Lop ears, imperforate anus, triphalangeal thumbs	Stippled epiphyses and goiter
	Agenesis of carotid artery
	Craniodiaphyseal dysplasia
Conductive	
Pierre Robin	Otopalatodigital
Treacher Collins	Hemifacial microsomia
Cleft palate	First and second branchial syndrome
"Cleft palate plus"	Frontonasal dysplasia
Apert	Oculodentodigital
Oral facial digital II	Ocular hypertelorism
Pfeiffer	Chondrodystrophia calcificans
Hallermann-Streiff	Congenital facial palsy
Otofaciocervical	Lop ears, micrognathia
Malformed, low-set ears	Kniest's syndrome
Conductive, Sensorineural or Mixed	
Goldenhar	Pre-auricular pits, cervical fistula syndromes
Crouzon	Facial hemihypertrophy
Moebius	Fibrous dysplasia
Microtia	Mucopolysaccharidoses
Wildervanck	Craniometaphyseal dysplasia
Acrodysostosis	Dominant and recessive "ear-kidney syndromes"
Partial deletion 18 chromosome	Diastrophic dwarfism
Osteopetrosis	Frontometaphyseal dysplasia
Thalidomide	Klippel-Feil
Cup ears, lacrimo-auriculo-dento-digital	Rubella syndrome
	Sclerosteosis

* From the Literature.

other than the aforementioned. Family history of congenital defects, including deafness should be sought. Neonateal or perinatal insults such as low birth weight, hypoxia, hyperbilirubinemia, ototoxicity, or neonatal meningitis may damage the organ of hearing (Bergstrom, Hemenway, 1971 a).

Case Report

A healthy male infant underwent cleft lip repair as a neonate and palate repair at 18 months. An older sister was congenitally deaf. He was noted to have serous otitis which was treated medically. The parents suspected hearing loss when he was six months old. At two years of age he had his first audiogram which showed no response in the right ear and responses in the left ear at 65 dB at 250 Hz, 90 dB at 500 Hz and questionable responses at 110 dB at 1000 Hz.

The presence of a craniofacial defect is not the only factor to be considered when deciding whether or not otologic consultation and audiogram are needed. This case exemplifies some of the defects in our present case-finding methods. First, the family history was apparently not appreciated. Second, the parents' worries were not listened to. Third, the lack of responsiveness to sound was attributed to a mild middle ear problem which, incidentally, was not reassessed to see if it had cleared. Fourth, consultation and hearing testing were not obtained until very late in the child's critical learning period for language although he had two excellent reasons for their being ordered.

Conclusions

It would appear from this series of patients that the likelihood of acquired or congenital hearing loss occurring in patients who have clefting or craniofacial syndromes is high enough to warrant obtaining otologic and audiologic consultation routinely.

References

BERGSTROM, L., HEMENWAY, W. G., and DOWNS, M. P., A high risk registry to find congenital deafness, *Otol.*

Clin. No. Amer., 4, 369–399, 1971 (a).

BERGSTROM, L., and HEMENWAY, W. G., Otologic problems in submucous cleft palate, *So. Med. J., 64,* 1172–1177, 1971 (b).

BERGSTROM, L., HEMENWAY, W. G., and SANDO, I., Pathological changes in congenital deafness, *Laryngoscope 82,* 1777–1792, 1972.

BERGSTROM, L., NEBLETT, L. M., and HEMENWAY, W. G., Otologic manifestations of acrocephalosyndactyly, *Arch. Otolaryng., 96,* 117–123, 1972.

HILSON, D., Malformation of ears as a sign of malformation of the genitourinary tract. *Brit. Med. J., 2,* 785–789, 1957.

IGARASHI, M., FILIPPONE, M. V., and ALFORD, B. R., Temporal bone findings in Pierre Robin syndrome. *Laryngoscope 86;* 1679–1687, 1976.

LINDSAY, J. R., BLACK, F. O., and DONNELLY, W. H. JR., Acrocephalosyndactyly (Apert's syndrome): temporal bone findings, *Ann. Otol., Rhinol. and Laryngol., 84,* 174–178, 1975.

MARAN, A. G. D., The Treacher Collins syndrome, *J. Laryngol., 78,* 135–151, 1964.

MÖLLER, P., Long-term otologic features of cleft palate patients, *Arch. Otolaryngol., 101,* 605–607, 1975.

NORTHERN, J. L., and DOWNS, M. P., *Hearing In Children.* Baltimore: Williams and Wilkins, pp. 61–74, 226–227, 1974.

PARADISE, J. L., and BLUESTONE, C. D., Diagnosis and management of ear disease in cleft palate infants, Trans. Amer. Acad. Ophthal., *Otolaryng., 73,* 709–714, 1969.

SANDO, I., HEMENWAY, W. G., and MORGAN, W. R., Histopathology of the temporal bones in mandibulofacial dysostosis, *Trans. Amer. Acad. Ophthal. and Otol., 72,* 913–924, 1968.

SANDO, I., BERGSTROM, L., WOOD, R. P. II, and HEMENWAY, W. G., Temporal bone findings in trisomy-18 syndrome, *Arch. Otolaryng., 91,* 552–559, 1970.

SANDO, I., LIEBERMAN, A., BERGSTROM, L., IZUMI, S., and WOOD, R. P. II, Temporal bone histopathological findings in trisomy-13 syndrome, *Ann. Otol., Rhinol. and Laryngol., 84,* Suppl. 21, 1975.

STERRITT, G. M., CAMP, B. W., and LIPMAN, B. S., Effects of early auditory deprivation upon auditory and visual information processing. *Percept. and Motor Skills 23,* 123–130, 1966.