# The Association of Facial Palsy and/or Sensorineural Hearing Loss in Patients with Hemifacial Microsomia

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**Hemifacial microsomia (HFM) is a common craniofacial disorder that is known to be etiologically heterogenous. Phenotypic differentiation of the various subgroups remains unresolved. A review of 50 patients with HFM has yielded data that may help explain different pathogenetic processes. Of particular interest is the association of facial nerve palsy, sensorineural hearing loss (SNHL), or both in a higher percentage of patients than expected. Twenty-two percent had evidence of facial palsy of varying degree. Thirty-three cases had microtia or anotia, and all instances of facial palsy were associated with auricular malformation. Sensorineural hearing loss was found in 16 percent. All patients with microtia and sensorineural hearing loss had facial palsy. Ear tags or pits were found in 21 patients, only two of whom had facial palsy. In all but one case the palsy was found on the more hypoplastic side of the face. In the single exception, both sides of the face were hypoplastic.**

KEY WORDS: *hemifacial microsomia, facial paresis, sensorineural hearing loss, microtia*

The term hemifacial microsomia was first used by Gorlin et al in 1963 to refer to patients with unilateral microtia, macrostomia, and hypoplasia of the mandibular ramus and condyle. Oculoauriculovertebral dysplasia (Goldenhar syndrome) was defined as a variant of this complex and was characterized by vertebral abnormalities, epibulbar dermoids, and other anomalies that may also occur in association (Gorlin et al, 1976).

HFM is a phenotypically and etiologically heterogenous disorder that is usually unilateral but frequently bilateral with more severe expression on one side. It is characterized by varying degrees of facial asymmetry and hypoplasia of the bony and/or muscular and soft tissues of the face; auricular anomalies; skin tags; pits, and different gradations of microtia (Fig. 1).

The purpose of this report is to cite the prevalence of clinical features in a sample of 50 patients with HFM, with special reference to facial paresis, hearing loss, and facial anomalies.

## METHODS

The charts of 50 consecutive patients with HFM were reviewed. The charts represented a partial sample taken in reverse chronologic order to assure that all information was complete. All 50 of the patients had undergone a complete pediatric, dysmorphologic and genetic work-up. In many of the older charts these were not available, and it was therefore decided to use the 50 most recent cases. All patients had undergone full audiometric testing, including auditory brainstem responses with bone conduction, when indicated. Cephalometric and panoramic radiographs, temporal bone tomograms, and, more recently, computed tomography (CT) scans were obtained and reviewed. The temporal bone tomograms and CT findings are important in assessing the degree of external auditory canal atresia and the presence or absence of middle ear ossicular chain, facial nerve canal, cochlea, internal auditory canal (IAC), and vestibular apparatus.

None of the patients had chromosomal anomalies or histories of teratogenesis. A positive family history for facial asymmetry, cleft lip and cleft palate, or ear tags or ear pits was obtained in six cases, but none of these family members were included in the sample.

## RESULTS

There were 27 male and 23 female patients. Forty-eight percent were right-sided, 28 percent were left-sided, and 24 percent were bilateral (Table 1). Thirty-three patients had microtia and 21 had preauricular skin tags or pits, resulting in a total of 94 percent of patients having auricular anomalies.

Peripheral facial nerve paralysis or paresis, total or partial, was found in 11 patients (22 percent). Nine had microtia (Fig. 2) and the remaining two cases had ear tags.

Of the 11 patients with VII cranial nerve palsy, six had sensorineural hearing loss (Fig. 3), whereas only two out of

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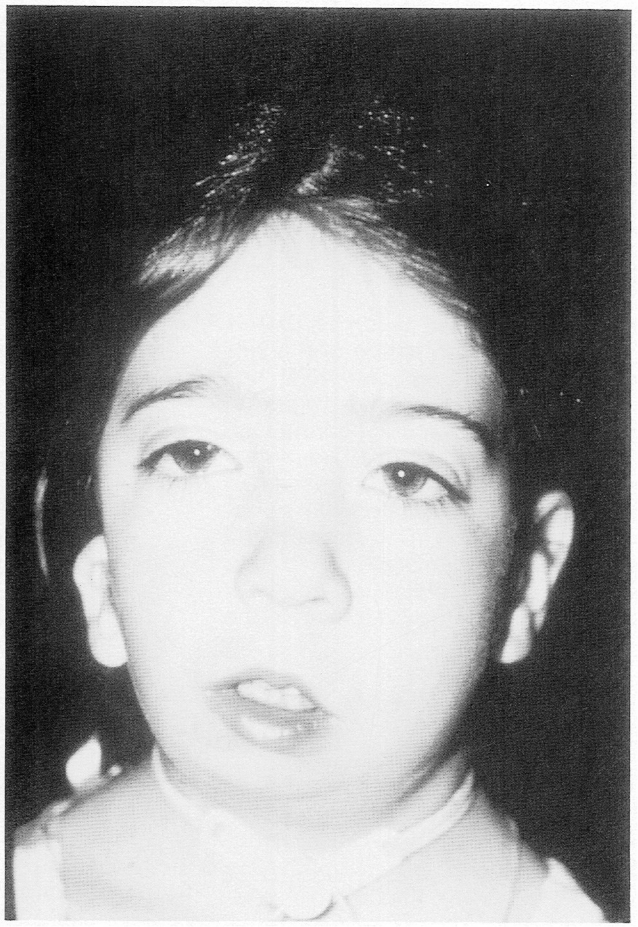

**FIGURE 1 Patient with right-sided hemifacial microsomia without facial palsy.**

39 patients with VII nerve findings had sensorineural hearing loss (Table 2). Of the eight patients (16 percent) with sensorineural hearing loss, four had right-sided HFM and the other four had left-sided HFM. Of the five cases with left-sided facial nerve findings, four had bilateral sensorineural hearing loss. All four patients with bilateral sensorineural hearing loss had left-sided facial nerve findings (see Table 2).

Of the eight patients with sensorineural hearing loss, two had CT scans of the temporal bones and three had tomograms. One CT scan showed a normal cochlea and vestibule on the affected side with sensorineural hearing loss. The other CT showed an absent cochlea and abnormal vestibule on the severely affected side in a patient with bilateral sensorineural hearing loss.

Of interest are the findings of an abnormal cochlea and vestibule on the CT scan of a patient with no sensorineural hearing loss and of a middle ear and mastoid soft tissue density (cholesteatoma) in a patient with grade III microtia and external auditory canal atresia (Fig. 4). The three polytomographs revealed atresia of the internal auditory canal with unilateral sensorineural hearing loss and absence of the facial canal in one patient with VII nerve paresis.

**TABLE 1 Distribution of Sample According to Laterality**

| | *No of Patients* | *Percentage* |
|---|---|---|
| Right | 24 | 48 |
| Left | 14 | 28 |
| Bilateral | 12 | 24 |
| Total | 50 | 100 |

## Discussion

The prevalence of facial nerve involvement varies in the literature. It was reported to be an occasional finding by Converse et al (1973), in whose article it is also mentioned that, in more severe deformities, the facial nerve and the fallopian canal may be absent and the nerve often has an abnormal course through the temporal bone.

Converse et al (1973) had described the anomalous course of the facial nerve in nine of 47 patients with microtia who underwent middle ear exploration for the purpose of establishing restoration of conductive hearing. The facial nerve was found to course over the temporomandibular joint rather than to exit through the stylomastoid foramen. Grabb (1965) reported a prevalence of facial paresis of 10 percent. It is not known whether the muscle weakness observed was caused by a primary deficiency of the mesoderm of the branchial arches, the neural ectoderm of the cranial nerves, or both of these primary germ layers. It was again determined that the chorda tympani branch was functionally intact. Gorlin et al (1976) reported a 10 percent prevalence of involvement of the lower mandibular branch, possibly related to bony involvement in the region of the facial canal. The higher prevalence of facial paresis in our sample may reflect an ascertainment bias, although this is probably not the case because the referral sources were numerous (i.e., pediatricians, orthodontists, speech pathologists, and others). Our findings are supported by the report of Bergstrom and Baker (1980), who found that in syndromes associated with facial paralysis there was a 23 percent prevalence of facial palsy in 12 patients with HFM, "Goldenhar syndrome", or both. The overall prevalence of neonatal facial palsy has been reported to vary from 0.25 percent to 6 percent.

Sekhar et al (1978) reported on temporal bone findings in HFM. The facial nerve fibers were nonexistent except for the nervous intermedius component (chorda tympani nerve). More recently, Sando and Ikeda (1986) described the facial nerve as hypoplastic in its entire course through the temporal bone. The tympanic segment was widely displaced in the fallopian canal, and the mastoid segment lacked a bony covering and had an anomalous course exiting cephalad to the stylomastoid foramen. The chorda tympani nerve was not visualized in the temporal bone.

Rapin and Ruben (1976) reported on unilateral weakness of the muscle innervated by the cranial VII nerve in six out of 16 children with malformed ears. Five of six had facial hypoplasia that was frequently associated with hypoplasia of the ramus of the mandible and malformation of the condyle.

The prevalence of sensorineural hearing loss associated with HFM has not been fully delineated in the literature (Budden and Robinson, 1973; Sando and Ikedo, 1986). Our report of a 16 percent prevalence of sensorineural hearing loss may reflect our careful scrutiny of our cases rather than an ascertainment bias.

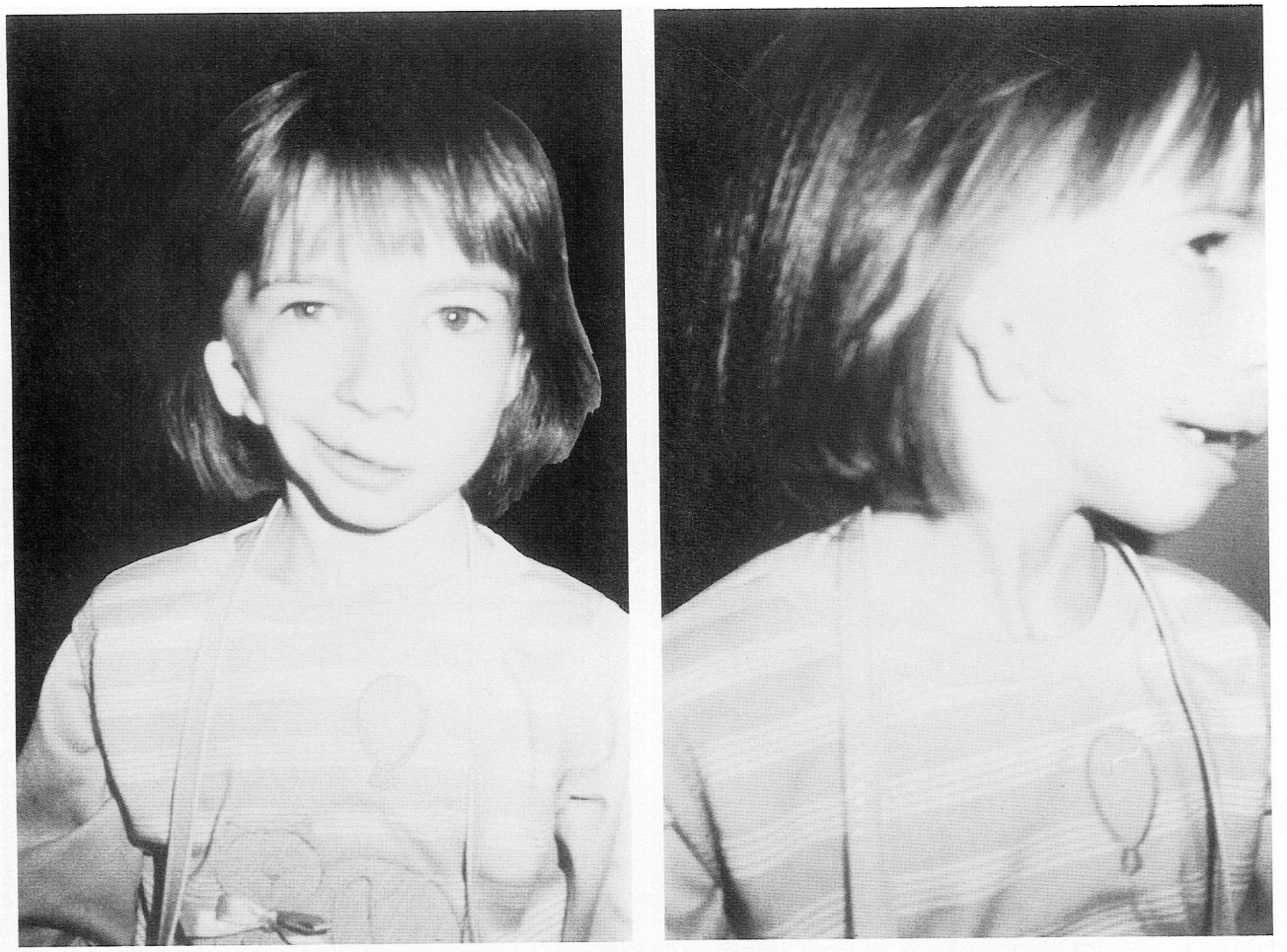

**FIGURE 2 Patient with right-sided hemifacial microsomia, grade II microtia, bilateral sensorineural hearing loss, and left-sided facial paralysis.**

The earlier developmental theories hypothesize that the otic capsule is resistant to the insult causing the spectrum of anomalies in HFM (Grabb, 1965; Poswillo, 1975). However, the more recent studies of the embryologic formation of the orofacial tissues and of the otic capsule have shown important interactions between the different mesenchymal tissues and epithelium in the development of the external ear, middle ear, and other facial structures, as well as the sensory receptors of the developing inner ear and the neural elements of the VIII cranial nerve ganglion (Wiznitzer et al, 1987). These occur during cytodifferentiation and spatial patterning and migration of the neural crest cells from the neural fold (Sulik, 1984; Noden, 1986).

Depending on the nature of the embryologic insult and the developmental sequence of events, varying degrees of anomalies may become manifest. These anomalies include sensorineural hearing loss in patients with HFM with structural abnormalities of the cochlea, vestibule, and IAC (in

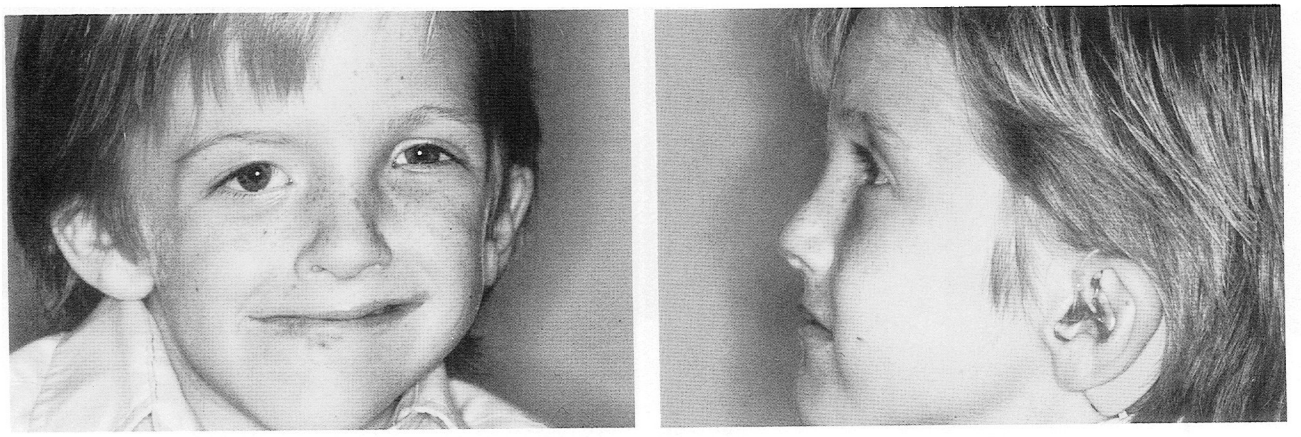

**FIGURE 3 Patient with hemifacial microsomia, right facial paralysis, and sensorineural hearing loss.**

**TABLE 2 Sample Population Distribution According to Facial Nerve Disorders and/or Sensorineural Hearing Loss**

| | *Microtia* | | *VII Nerve Palsy* | | | *Hearing* |
|---|---|---|---|---|---|---|
| *Case #* | *Laterality* | *Grade** | *Paralysis* | *Paresis* | *Branch* | *SNHL* |
| 1 | Right | I | | Yes | Right buccal | Right |
| 2 | Left | III | Yes | | Left total | |
| 3 | Left | III | | Yes | Left temporal | Bilateral |
| 4 | Right | I | | Yes | Left buccal and mandibular | |
| 5 | Right | II | Yes | | Left total | Bilateral |
| 6 | Right | III | | | | |
| | Left | II | | | | |
| 7 | Right | III | | | | Right |
| 8 | Right | III | | | | Right |
| 9 | Right | III | Yes | | Left total | Bilateral |
| | Left | III | | | | |
| 10 | Right | III | | Yes | Right mandibular | Right |
| 11 | Right | I | | Yes | Right total | |
| 12 | Right | III | Yes | | Right temporal | |
| 13 | Right | III | Yes | | Right total | |
| 14 | Right | II | | Yes | Left buccal and mandibular | Bilateral |
| | Left | III | | | | |
| Total | | 14 | 5 | 6 | | 8 |

* Based on Meurman Y. (1957). Congenital microtia and meatal atresia. Arch Otolaryngol 66:443–463.

some) and normal outer, middle, and inner ear structures with possible isolated sensory and neural involvement (in others).

The interesting segregation of facial palsy, sensorineural hearing loss, ear malformations, and laterality may be explained in a number of ways. The spectrum of anomalies seen by us may reflect the well established etiologic heterogeneity of the disorder. One might then wonder whether the various entities observed represent distinctly different "syndromes" or phenotypic overlap within a single sequence with etiologic heterogeneity. None of our patients had identifiable chromosomal aneuploidies or histories of teratogenesis, and the diagnosis of a specific known syndromic disorder other than HFM was not possible. There were no cases of Towne syndrome or of other known genesis malformation syndromes with ear tags, pits, or facial asymmetry. Furthermore, the presence of six cases with some familial findings does not necessarily imply that these cases are distinctly different from the nonfamilial cases. For one thing, in the familial cases, all of the family members had either minor malformations that might be considered to be within the spectrum of HFM anomalies, such as an ear

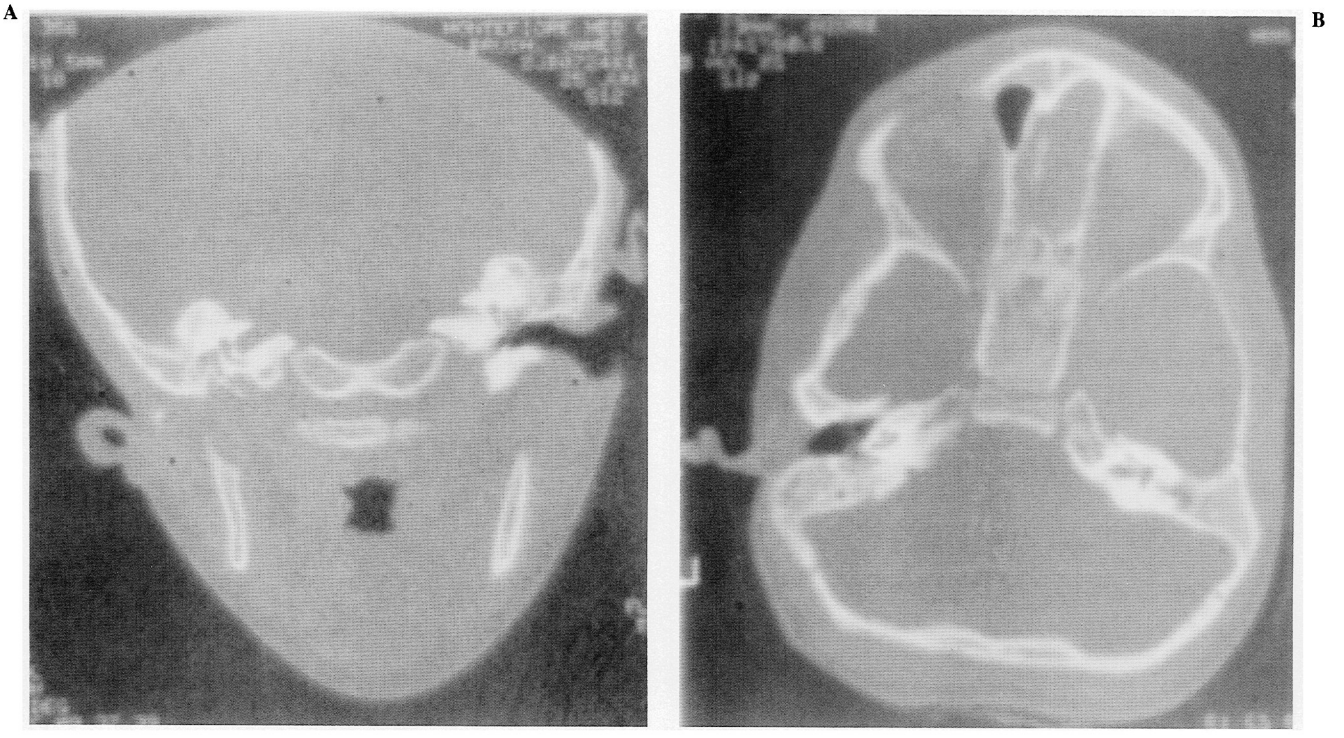


**FIGURE 4 CT scan of patient with middle ear and mastoid soft tissue density representing cholesteatoma.**

tag, or they had cleft lip and cleft palate. None of the family members had obvious manifestations of HFM. It is therefore unclear as to whether the familial cases represent specific malformation syndromes of known genesis. Also, there were no phenotypic differences between the familial and nonfamilial cases, and facial paresis and sensorineural hearing loss were present in both the familial and nonfamilial cases. No family members were found with either facial paresis or sensorineural hearing loss. Until specific pathogenetic mechanisms of this fairly common spectrum of anomalies are more precisely defined, the relationship between facial paresis, sensorineural hearing loss, and the rest of the facial anomalies associated with HFM will remain a matter of speculation.

An alternative explanation for the observed variability is that all of the patients examined have the same developmental sequence, with the observed differences being attributed to variable expression. It is, of course, impossible to confirm any hypothesis of causation based on our retrospective data. But if one accepts the premise that the HFM phenotype is nonspecific and may have many possible causes, clinicians are then obligated to search carefully for facial weakness and sensorineural hearing loss as a possible clinical feature in every case of facial asymmetry.

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