Audiologic and Educational Issues in CHARGE Syndrome

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CHARGE syndrome is a group of congenital anomalies that can have complex effects on health and on many aspects of development. The prevalence of ear anomalies and hearing loss is 90% and, in any individual, all parts of the auditory system may be involved. The first part of this article deals with the presence and consequences of anomalies, medical and health factors, and special issues that need to be considered in the assessment and treatment of hearing loss. The second part of the article deals with the selection of a mode of communication and with the recommendation for inclusive education for a child with CHARGE. The role of the audiologist in the formulation and attainment of educational goals is stressed.

Introduction

In 1979, Hall reported on the relationship between choanal atresia (blockage of the nasal breathing passages) and other congenital anomalies. In 1981, Pagon and her co-workers reported on a group of patients with similar features. They identified the group of anomalies as a genetic association and named it using an acronym for the major diagnostic anomalies: C-coloboma of eye (keyhole defects in the iris and/or retina) H-heart defects A-atresia choanae R-retardation of growth and/or development G-genitourinary anomalies E-ear anomalies and/or deafness

Since 1981, the name for the disorder and the diagnostic criteria for inclusion have been discussed at length. The name \textit{CHARGE association} has been used by geneticists who believe that the disorder is appropriately described as a nonrandom collection of birth defects of unspecified cause; the name \textit{CHARGE syndrome} has been used by those who believe that a specific genetic cause will be identified in the future. No consensus has been reached about the preferred name. However, agreement has been reached to change the diagnostic criteria for \textit{CHARGE} (Blake et al., 1998; Hefner & Davenport, 1999). There are only four major features in the revised diagnostic criteria. They are listed below with the frequency of occurrence of each anomaly in parentheses:

- Coloboma (80-90%)
- Choanal Atresia (50-60%)
- Cranial Nerve Dysfunction
  - I-lack of smell (Frequent)
  - VII-facial palsy (40%-+)
  - VIII-sensorineural hearing loss or vestibular problems (70-85%)
  - IX/X-swallowing dysfunction (70-90%)

\textbf{Characteristic CHARGE Ear}

- \textbf{External ear:} short, wide ear with little or no lobe, snipped off helix, prominent antihelix discontinuous with tragus, triangular concha, decreased cartilage, asymmetric pinnae often protruding laterally (90%) (See Figures 1 and 2.)
- \textbf{Middle and inner ear:} abnormalities of the stapes, absent stapedius tendon, cochlear anomalies including Mondini's dysplasia (90%?)

![Figure 1. Photographs of auricles from children with CHARGE](image1)

![Figure 2. Line drawings of characteristic auricles of children with CHARGE](image2)

There is also a list of minor diagnostic criteria: characteristic \textit{CHARGE} face and hand, genital hypoplasia, congenital heart defects, cleft lip/palate, tracheoesophageal fistula, middle-ear infection, hypotonia, renal anomalies, and growth deficiency. Other common findings include the following: brain abnormalities, apnea, seizures, laryngomalacia (softness of laryngeal...
cartilage), nipple anomalies, thymic or parathyroid hypoplasia, webbed neck, abdominal wall defects, scoliosis, limb/skeletal anomalies, autistic-like behavior, and behavior problems. Recurrence risk for parents with one child with CHARGE is 1-2% (Davenport, 1999a).

The prevalence of CHARGE syndrome is estimated at 1:10,000 (Blake et al, 1998). Most cases are sporadic, but there is evidence CHARGE may be inherited. It has been reported in four generations of one family (Mitchell, Giangiacomo, Hefner, Thelin, & Pickens, 1985), in association with chromosomal abnormality (North, Wu, Cao, Whiteman, & Korf, 1995), and in fraternal twins (Brown & Israel, 1991). Several patterns of inheritance are possible (Toriello, 1995).

The presence of particular abnormalities and the severity of involvement for any particular structure or system vary greatly across individuals. Although individuals with CHARGE often have many of the same physical and sensory disorders, they may function very differently because of varying degrees of abnormality for each involved system. Since very few individuals can be considered as typical, the most beneficial approach to diagnosis and treatment is to consider the specific abilities and disabilities of each individual.

The degree of hearing loss in CHARGE is often severe to profound. Because both hearing and vision loss are often present, the syndrome is considered by some to be a deaf-blind syndrome. However, problems with the sense of smell and the vestibular sense make it even more appropriately a multi-sensory impairment syndrome. The problems associated with sensory deficits are typically compounded by the presence of physical disabilities. In the first 2-3 years of life, parental focus is on the survival of the child. Once that period has passed, parental focus shifts to mental, physical, and social development. In this period, the importance of hearing becomes obvious, and the resolution of hearing problems becomes a major concern of many parents. The purpose of this article is to describe the audiologic and educational issues that are important to the success of a child with CHARGE syndrome. The article is written from the perspectives of an audiologist (JWT) and of parents of a child with CHARGE who are also psychologists (TSH and NSH).

Early Medical Problems

During the first few years of life, parents and physicians are focused on keeping the child with CHARGE alive. The average child with CHARGE will be treated by 17 different medical specialists and will have more than 20 surgical procedures before 10 years of age (Hefner, 1999). Parental concern with medical issues may result in a lack of attention to communication and learning issues, including issues related to hearing.

Parent Reactions

It is rare that parents have any warning that their child has difficulties, let alone CHARGE. It is very difficult for ultrasounds to detect evidence-of CHARGE, and the physician would have to be looking specifically for indications. Some children with CHARGE are born prematurely due to polyhydramnios or excess amniotic fluid, but this may be associated with numerous other conditions. It would be rare for parents to have even heard of CHARGE. Further, it may also take several days or even considerably longer for the parents to receive a diagnosis. Thus parents may be in shock after the birth, and not in a good position to consider the implications of their child's various medical anomalies. They are also likely to be told that the abnormal external ears are not necessarily associated with hearing loss. Neonatologists and other physicians are likely to be more focused on survival problems, than whether the external ear malformations are related to deafness, and parents will follow the lead of the physicians.

Once the diagnosis of CHARGE is made, many parents make contact with the CHARGE Syndrome Foundation (see end of article for contact information). This organization supports parents and professionals in need of information. It publishes a 130-page management manual for parents and a newsletter, supports a forum for parents on the Internet, and sponsors conferences for families and professionals. As a result, the typical CHARGE parent quickly becomes informed about the unique needs of the child with CHARGE and about the professionals who have the expertise and patience to deal with the complex treatment issues.

Initial survival concerns

Heart. Cardiovascular malformations in CHARGE may include nearly all types, and can range from very mild to life threatening. The most common is tetralogy of Fallot, which is found in 32% of children with CHARGE (Blake et al., 1998). This involves a hole between the ventricles, obstructed outflow of blood to the lungs from the right ventricle, a displaced aorta receiving blood from both ventricles, and enlargement of the right ventricle (Hartshorne & Hartshorne, 1997). While the heart defects may be serious, they are not a major cause of fatalities with CHARGE (Tellier et al., 1998; Wyse, Al-Mahdawi, Burn, & Blake, 1993). Airway and swallowing problems are generally more serious. Airway. Many children with CHARGE have breathing difficulties at birth, and are intubated. Breathing difficulties may be caused by choanal atresia, recession of the jaw, tracheoesophageal fistula, stenosis or laryngeal paralysis, and clefts. A tracheotomy may be performed in 10-30% of cases (Roger et al., 1999). Choanal atresia repair will be necessary, and is now commonly done shortly after birth. However, several follow-up surgeries to dilate or widen the opening may be required (Morgan & Bailey, 1990). Parents will therefore be preoccupied with concerns for helping their child to breathe. Swallowing. Related to the airway problems is difficulty with sucking and swallowing. Secretions tend to pool in the throat and then could be aspirated. This is complicated by gastroesophageal reflux that brings stomach contents up into the throat where they may also be aspirated (Roger et al., 1999). The frequent episodes of pneumonia that result are a major concern and can be life threatening. This is generally treated by way of the insertion of gastrostomy feeding tubes (Blake et al., 1998).
Issues related to development

Determination of intellectual ability. Of particular concern is the “R” in CHARGE (N. Hartshorne, 1999). Early investigators, such as Hall (1979) and Pagon et al. (1981), found all or nearly all participants to be mentally retarded, and this finding continues to be cited in the literature in support of this characteristic. However, Blake and Brown (1993) found many children both in their participant group and outside of it who were of normal intelligence. They note that the effects of multi-sensory impairment in early childhood development are not well understood by most professionals and argue that mental retardation should not be considered an essential part of the syndrome, but rather an outcome of the other disabilities involved. N. Hartshorne (1999) gathered adaptive behavior scores and medical histories for 100 cases of children with CHARGE, and found a broad range of abilities, with a mean score above the number required for diagnosis as mentally retarded. Furthermore, low scores were associated with greater medical involvement and sensory impairment. Professionals should not assume that a child with CHARGE will have cognitive delays and even more importantly, that his or her potential is below that of other children. Parents of a child with CHARGE often have the opportunity to see evidence of intellectual function that may not be obvious or adequately assessed in a formal evaluation. If the parents feel that the child’s intellectual capabilities have been underestimated because the evaluation was not appropriate, they typically – and justifiably – have a strong emotional response.

Role of the audiologist. The audiologist is not central to the survival concerns; however, parents will be anxious to find as much information about their child’s condition as possible (T. Hartshorne, 1993). An obvious question, rarely answered by physicians, is whether the misshapen outer ears are associated with hearing loss. It is common when there are concerns to order an auditory brainstem response (ABR) test. Parents will need assistance directly from an audiologist to understand the implications of the report. Finally, the airway, swallowing, and vestibular problems seem to be related to cranial nerve problems including most likely the auditory nerve. The audiologist should be part of the team evaluating both the implications of these difficulties as well as possible interventions.

Many parents have been counseled by knowledgeable professionals or have networked with other CHARGE parents and are aware that children with CHARGE are often very difficult to evaluate audiologically. They are aware that it may take numerous sessions to gain the information needed to fit amplification. However, parents express great frustration when the same procedures are performed unsuccessfully on repeated visits and the hearing remains unspecified. Parents have reported cases in which hearing levels have not been determined after seven years of audiologic evaluations. If an audiologist cannot specify the hearing in a reasonable amount of time, then the family must be referred to a facility with special capabilities for audiologic evaluation.

Characteristics of Hearing Loss in CHARGE

General description of hearing loss. Newborns with CHARGE are at risk for hearing loss using Joint Committee on Infant Hearing criteria (American Speech-Language-Hearing Association, 1994). Most, but not all, individuals with CHARGE have a significant degree of bilateral hearing loss — some mild, but many moderate to profound (Buchanan, 1999; Edwards, Van Riper, & Kilency, 1995; Shah et al., 1998; Thelin, Mitchell, Hefner, & Davenport, 1986). The losses in the two ears are often asymmetrical. The most common type of loss is mixed hearing loss due to ossicular anomalies and cochlear dysfunction. The overall hearing levels may vary as a result of fluctuant conductive loss due to chronic otitis media that is common and persistent when craniofacial anomalies are present. Gradual progression of the hearing loss is uncommon, but has been reported. Examples of CHARGE hearing losses are shown in Figures 3 and 4.

![Graph showing hearing levels and frequencies](image)

Figure 3. Behavioral pure tone audiogram for a 17 year old female with mental retardation. Sound-field results indicate a profound hearing loss in the better ear. Bone-conduction thresholds show the characteristic sloping sensorineural loss in the better ear. The air-bone gap is due in part to chronic middle-ear disease (flat tympanograms and sclerotic tympanic membranes) and, possibly, to ossicular hearing loss. (from Thelin et al., 1986.)

Auditory anomalies and their effects

External ear. The shapes of the auricles in CHARGE are so distinctive that tentative diagnoses of CHARGE have been made on the basis of this criterion alone. The verbal descriptions of the shapes of auricles (see revised diagnostic criteria) may differ significantly across examiners. For this reason, images of characteristic CHARGE ears have been provided in Figures 1 and 2 to illustrate the range of auricular shapes that have been found (Davenport, 1999b; Davenport, Hefner, & Thelin, 1986c). Congenital aural atresia is a rare finding in CHARGE, and malformations of the external ears rarely cause hearing loss. In some cases, the shape of the auricle can be modified with nonsurgical procedures immediately after birth. In most cases, if auricular reconstruction is undertaken, it is done later in childhood.

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Middle ear. Conductive hearing loss due to malformation of the ossicles is common, and the stapedius muscle may be missing or dysfunctional (Shah et al., 1998; Thelin et al., 1986; Wright, Brown, Meyerhoff, & Rutledge, 1986). The conductive loss due to ossicular anomalies may be large for the low frequencies while the conductive loss in the high frequencies may range from nonexistent to large. With ossicular anomalies, if there is no middle ear effusion, tympanograms obtained using a 226-Hz probe signal may be normal.

Because many children with CHARGE have craniofacial anomalies, eustachian tube dysfunction is ubiquitous and may persist into adulthood. In a survey of parents of children with CHARGE, 98% reported a history of otitis media and 74% reported that tympanostomy tubes had been placed (Thelin & Stephens, 1996). If the conductive loss is due to both the ossicular malformation and middle ear effusion, it may be very difficult to determine the amount of loss attributable to each disorder. Perforated tympanic membranes and draining ears—the sequelae of otitis media—are common. Often the presence of drainage contraindicates the use of one or both of the child’s hearing aids or necessitates use of only one hearing aid at a time.

Moderately severe (or greater) mixed hearing losses are common and are difficult to correct with hearing aids. When the conductive component is significant, high-levels of amplification are required using earmolds that may not seal well and hearing aids that may slip off the ear easily. In these cases, it may be difficult to control hearing-aid feedback without reducing hearing aid gain to inappropriately low levels. In cases with low-frequency conductive loss and high-frequency sensorineural loss, attempts have been made to use a bone-conduction aid on one side and an air-conduction aid on the other. There are no reports on this approach in the literature, but anecdotally, the arrangement is sufficiently cumbersome that compliance is difficult to achieve.

Cochlea. Cochlear hearing loss is common and is typically greatest in the high frequencies (Edwards et al., 1995; Thelin et al., 1986). There may be malformation of the cochlea—such as a Mondini’s dysplasia (Wright et al., 1986)—that can be detected with radiographic imaging procedures or there may be cochlear loss with no observable anatomical abnormality. There also may be vestibular problems associated with cochlear anomalies (Admiraal & Huygen, 1997; Murofushi et al., 1997).

Auditory nervous system. Central nervous system anomalies including cerebral dysgenesis and agenesis of the corpus callosum have been reported (Korf, 1999). In most cases, there are enough questions about the status of the peripheral auditory system that ABR testing is done to resolve those issues rather than to evaluate the function of the auditory nervous system. A case has been reported in which ABR click thresholds were within normal limits and delayed neural transmission times were found using the ABR latency-intensity function (Thelin et al., 1986). Central auditory function has not been reported using other auditory evoked potential measures or behavioral tests of central auditory function. In many cases in which this information would be valuable, the individuals lack the ability to cooperate and/or sufficient hearing to perform the procedures successfully.

Assessment of Hearing
Otoacoustic emissions and ABR. Since hearing loss is almost always present at birth in CHARGE, there is the substantial likelihood that the newborn with CHARGE will fail an otoacoustic emission screening or an ABR screening using a low-level signal. A more effective approach may be to proceed directly to diagnostic evaluations of auditory function and hearing. Since a large part of a newborn’s first few days of life are spent sleeping, this may be an ideal time to perform lengthy
ABR tests without the use of sedation which is often required with infants and toddlers.

Children with CHARGE who have breathing and swallowing difficulties generate physiologic noise that often obscures the ABR responses. If sedation is necessary to perform the ABR test, then risk factors associated heart, breathing, and swallowing problems need to be considered by the physicians responsible for the child. Further, it has been reported that some children with CHARGE exhibit an increased resistance to sedation (Thelin et al., 1986).

Tympanometry and acoustic reflex measures. In children with CHARGE, tympanometry provides a great deal of useful diagnostic information. The use of multifrequency tympanometry and acoustic reflex measures should be considered for detecting otitis media in infants under six months of age and for identifying ossicular anomalies (Hunter & Margolis, 1992; Margolis et al., 1997). Tympanometry should be performed as early in life as possible so that middle-ear function can be assessed without the presence of middle-ear effusion. If normal tympanometry is obtained in the presence of conductive hearing loss, there is evidence of ossicular involvement. Acoustic reflex tests usually are less useful because the reflex is often obliterated by conductive and cochlear loss. However, in cases in which hearing is within normal limits, the absence of an acoustic reflex may indicate absence or dysfunction of the stapedius muscle or tendon. Though this may have little effect on hearing, it may be important in establishing the medical diagnosis (Thelin et al., 1986).

Behavioral audiometry. Behavioral thresholds are the most important results to define the degree and pattern of the hearing loss and for the selection of appropriate amplification. In most cases, the behavioral pure-tone thresholds provide a more detailed description of the hearing than do physiologic measures. If there is concern about poor processing in the auditory nervous system, it is most reassuring to find that the child being tested can provide a voluntary response. However, there are many reasons why children with CHARGE can be very difficult to evaluate behaviorally (Thelin, 1999):

- poor ability to understand the requirements of the tests because of developmental delay due to illness, sensory deprivation, and/or cognitive delay
- inability to speak or otherwise communicate
- poor physical mobility—especially head and neck movement
- voids in the visual fields due to retinal coloboma that make head movements difficult to interpret in localization tasks
- tactile defensiveness and autistic-like behaviors
- noisy breathing
- chronic otitis media

Despite these obstacles, every attempt needs to be made to obtain behavioral thresholds for pure tones and speech at the earliest age possible. This may require innovative adaptation of procedures for behavioral observation audiometry, visual reinforcement audiometry, and conditioned play audiometry.

When the loss is mixed, less conventional forms of bone conduction testing may be useful in specifying the degree of cochlear loss: bone-conduction speech reception thresholds with behavioral audiometry and ABR responses to bone-conducted clicks and frequency-specific signals.

It may be useful to have the parents practice with the child at home by placing earphones on and off and practicing responses to sound. There needs to be commitment to testing as much as necessary in a short period of time to obtain the result that will lead to appropriate treatment. For audiologists who are uncomfortable fitting hearing aids without behavioral data every effort needs to be made to obtain valid and complete results as quickly as possible.

Amplification

The fitting of hearing aids and/or FM systems on children with CHARGE is both a difficult and necessary task. Parents are aware that, if there is significant hearing loss, the development of language and speech depends critically on the use of amplification. Parents may need special encouragement and support in using the aids with their child. They may have difficulties getting the aids to fit properly and avoiding feedback, because, due to mobility problems, their child is lying prone much of the time. Under these conditions, parents often keep close watch on their child so that he or she does not remove the aids and throw them. Doubts about whether their child is benefiting from the amplification contribute to parents not making the wearing of aids a high priority. These difficulties need to be acknowledged, and support for coping with them needs to be offered. However, even with support, a large number of children with significant hearing loss are not wearing amplification on a regular basis or have rejected amplification (Edwards et al., 1995; Thelin et al., 1986; Thelin & Stephens, 1996). All members of the multidisciplinary team who are responsible for the child need to support the use of amplification (Morgan, et al., 1993; Shah, et al., 1998). Support that can be given in the educational setting is discussed in the following section.

External ear anomalies often cause major problems in fitting and successful wearing of hearing aids. In most cases, the ear canals and conchae are small enough that in-the-ear aids cannot be considered. In many cases, the auricles are misshapen in a way that prevents fabrication of an earmold for a behind-the-ear aid that fits securely. Furthermore, behind-the-ear aids have a tendency to slip off auricles that are formed of soft cartilage and rotated posteriorly rotated. Often, the auricles are not large enough to use “Huggie Aids” successfully. Parents have successfully used double-sided ostomy tape and a fine-coil plastic loop used to hold eyeglasses (The Hilco Invisible Hugger Sports Band, Hilco Company, Plainville, MA 02762) (D’Luna, 1997). A number of children with CHARGE have received cochlear implants. Until relatively recently, there has been reluctance to perform this procedure on children with multiple congenital anomalies. At this time, there is no information in the literature on the results of these procedures.
Choice of communication system

Choosing the appropriate communication system is a topic that is sufficiently complex that it is beyond the scope of this article. For children with CHARGE, it may be late in childhood before the final decisions are made. In a survey of 72 parents with children primarily under 10 years of age, the following communication methods were reported: 47% speech, 38% sign language, 50% own method, 7% communication board, and 18% other methods (Thelin & Stephens, 1996).

From the parents’ standpoint, the choice of a communication system is one of the most important as well as most difficult decisions a family must make. It forces them to come to terms with the degree of their child’s limitations. It can have an impact on family functioning for years to come (for example, if family members must learn a sign system). It also requires knowledge and expertise of complex communication issues that very few parents have acquired. Parents need to receive as much information as possible regarding the choices that exist and the important considerations to make regarding the potential benefits and limitations of each alternative. Sometimes different consultants have their own biases about what system is best, and parents can rapidly become very confused (as can other educational personnel). Delays can take place as parents and schools implement and then change the primary communication approach. A consistent approach is essential, and it is worth taking some time for everyone, particularly the parents, to review alternatives and make a choice that everyone can support. After the primary communication approach has been chosen, a plan for evaluation of the child’s progress should be established to determine that the approach is beneficial. After a trial period (possibly 6-12 months), modification of the approach may be considered if progress has not been satisfactory.

Inclusive education

Inclusive education is a growing trend in the field of special education. More and more children who are deaf-blind, and specifically children with CHARGE, are being fully included in general education classrooms (Hartshorne & Hartshorne, 1998). The concept of inclusion is one that is often confused with mainstreaming (Stengle, 1996). The term “mainstreaming” refers to the practice of allowing a child to participate in the general education curriculum when he or she is capable of doing so without special supports. For example, a mainstreamed child may attend a special education resource room for part of the day for support in the area of reading, but remain in the general education classroom for math instruction. The child is permitted to remain in the general classroom only because he or she has demonstrated the ability to keep up with the class in that area with no special supports. Conversely, the term “inclusion” refers to the practice of providing any needed special education supports and services to a child within the general education classroom, regardless of the child’s current skill level. Special education in this case is seen as a service, and not a place, and can thus be provided to the child in the environment that removes him or her from his non-disabled peers the least. Thus, inclusive education provides a way for schools to meet legal obligations to educate children with disabilities in the least restrictive environ-
ment. In addition, it should be noted that children in inclusive placements should always be receiving the level and quality of special supports and services they would be receiving if they were being educated in a more segregated placement.

If inclusion is well planned, there are many benefits to the student and to his or her non-disabled peers. Recent research has indicated that children in inclusive settings meet a greater number of their IEP goals, are more motivated, have natural models for appropriate behavior, and have sustained interactions with peers without disabilities, to name a few benefits (Stengle, 1996). Additionally, children without disabilities will have the opportunity to socialize in the educational environment with children who have disabilities.

In order to meet the special needs of children with CHARGE in an inclusive setting, curricular adaptations will likely be necessary. There are several approaches to consider, the choice depending upon the individual child. 1) A child may be able to access the general education curriculum in some areas, with few or no adaptations. 2) A child may need special adaptations such as Braille or large print, an FM system, or a sign language interpreter in order to participate. 3) A child who has not yet developed formal communication may need a parallel curriculum in some areas. For example, as the fifth grade class is practicing long division, the child with disabilities may be learning to sort functional objects by color, thus “dividing” them into groups. When the class is reading silently, a child who is deaf-blind may be looking at experience stories, books constructed from objects familiar to the child based on a certain experience. The child with CHARGE is also “reading”, and this represents working within a curriculum that is parallel to that accessed by the children without disabilities. At certain times, a totally alternative curriculum may be chosen for a child in an area for which adaptations or parallels do not avail themselves. For example, while the other children are studying social studies, a child with CHARGE or deaf-blindness may be working on a targeted objective that does not easily fit into any particular general curriculum area (such as bathroom skills). The team, in developing these curricular adaptations, should keep in mind the principle of “least-restrictive materials” (Stratton, 1984, p.3), which recommends only making adaptations when necessary. They may not be necessary in all activities.

A particularly useful method of making these decisions is through the use of a curriculum matrix. A curriculum matrix is a chart on which are listed all of the student’s objectives as written on the Individualized Education Plan. These objectives are plotted on one axis of the chart, while the general education curricular schedule and lesson plans are plotted on the other. The educational team then can clearly see where the student’s goals and objectives fit into the general education curriculum, and can devise ways to meet the objectives within each lesson. The team must make certain that each objective is targeted in such a way that the child is able to have enough practice to meet it.

In inclusive education, teams must meet regularly in order to plan and keep up with the frequent changes in the general education curriculum (Ronel & Byrne, 1995). In one model, the entire team (including all general and special educators, therapists, parents, and the child, if appropriate) meets monthly after
school to address larger issues. A “core” team (which, for example, may consist of the parent, general education teacher, special education teacher, speech and language pathologist, and/or aide) meets weekly to look at curricular changes and devise lesson plans and materials for the aide to use with the child during the upcoming week. An audiologist can be an integral member of either of these groups, and can help to ensure that the child’s educational and social objectives are facilitated. The following is a listing of some of the contributions that an audiologist might make:

• Provide general training to the team and to the child’s classmates on hearing impairments and on how critical it is that the child hear as well as they can.
• Conduct simulations of the individual child’s hearing ability for family, staff, and classmates.
• Provide consultation to appropriate team members on hearing aid care, hearing aid checks, the uses of FM systems, and auditory training strategies.
• Train staff and classmates in the proper use of an FM system.
• Provide suggestions for ways to use an FM system to facilitate social interaction with peers (i.e., passing the microphone around during class discussions or recess; having the teacher wear the microphone while reading to the children).
• Provide consultation to the family regarding proper hearing aid care and checks.
• Check to see that hearing aids are in good working order and that ear molds have a proper fit.
• Facilitate a choice of earmold or aid that fits in with acceptable fashion with the child’s classmates (i.e., neon colors, camouflage).
• Suggest ways to facilitate communication between the child and peers.
• Attend team meetings regularly and offer input into educational plans.

While audiologists may not generally be employed by school districts as members of the special education support staff, it is imperative that these experts in hearing be involved in the educational teams of children with CHARGE. Within inclusive educational settings or not, the audiologist needs to ensure the child is making use of whatever hearing he or she has in a functional and meaningful way which is integrated into all curricular areas.

Conclusion
Children with CHARGE have problems of extraordinary complexity. Their problems are just beginning to be understood by the professionals whose help they need. Audiologists have a key role because many aspects of development depend critically on hearing. With the information presented in this article, audiologists hopefully will have a better basis to assist children with CHARGE in reaching their full potential.

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