## SYMPOSIUM ON OFFICE EVALUATION OF INTELLIGENCE AND SPECIAL SENSES

# OFFICE DETECTION OF HEARING DEFECTS IN CHILDREN

**By John E. Bordley, M.D.**

*Department of Otolaryngology, Johns Hopkins University School of Medicine*

STATISTICAL studies undertaken in many communities of this country show rather marked variation as to the percentage of children suffering from significant handicaps in communication. However, none of those making such surveys would feel it an exaggeration if one ventured to state that the instance of handicapping impairments of hearing range somewhere between 1 and 2% in the preschool and early school age children. The number of crippling hearing problems apparently is growing. This growth paradoxically seems to be proportional to the improvement in the obstetrician's and pediatrician's ability to save children, particularly children born after severe trauma, and premature children.

The national picture of the hearing handicapped child is changing. Thirty years ago our deaf schools were filled with children whose hearing impairments were caused by sequelae of meningococcic meningitis or of other infections that involved the ears. This no longer pertains, and we are faced today with a population of children whose hearing handicaps include lesions not localized to the middle or inner ear or even to the auditory nerve. Very few of these children are completely deaf. They suffer either distorted hearing as the result of damage to the end-organ, or, due to some central damage they are unable to understand speech because they cannot properly comprehend or synthesize speech patterns without special training. Often such youngsters suffer from multiple neurologic handicaps.

Many children have an amazing ability to learn communication both from an auditory and a speech point of view, provided that their problem is detected early and carefully evaluated, and that they are placed in a well-designed training program. After the age of 6 or 7 years, children with such injuries can rarely be trained with as good results as are obtained in the preschool group. Early detection therefore is of the utmost importance.

In the main, early recognition of hearing impairments lies in the hands of parents and pediatricians. After detection, these children can be sent to hearing centers associated with many of the great medical centers in this country where definitive studies can be carried out and their educational program can be outlined. It is evident, however, that without early detection on the pediatric front, medical centers with all of their expensive and elaborate equipment will be of little use to help these handicapped children.

Children with hearing handicaps present varying problems to the clinical pediatrician. They may have other neurologic changes which tend to mask their communicative difficulty. They usually seem to be mentally retarded when compared to children with normal hearing. So, in many instances, the diagnosis of mental retardation robs these children of an opportunity for early definitive education. This should always be borne in mind when a youngster is being examined and where aberrance in development could possibly be associated with inability to communicate.

The most powerful tool available to the

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**ADDRESS:** Baltimore 5, Maryland.

pediatrician and to the clinical otologist for the detection of hearing problems in young children is undoubtedly a careful history. The history should be so designed as to inform the examiner of any condition that might predispose toward hearing damage. Table I shows the outline that was developed in our clinic about 10 years ago, when we started to make a systematic study to determine etiologic factors in hearing problems. It is, I am sure, very much the same as that used in most pediatricians' offices. However, the tabulation of items serves to emphasize certain details.

TABLE I

FORM FOR HISTORY*

*Family History*
- Hearing impairment (relationship; age)
- Siblings
- Miscarriages

*Pregnancy*
- Rh incompatibility
- Exposure to children's diseases
- Immunizations
- High fever
- Bleeding (when)
- Toxemia
- Drugs used (when)

*Delivery*
- Hours in labor
- Type of birth
- Instrumentation
- Presentation
- Anesthetic used

*Neonatal Status*
- Appearance
- Weight
- Cyanosis
- Jaundice
- Oxygen therapy
- Transfusions
- Deformities

*General Health*
- No history (adopted)
- Serious illness
- Children's diseases
- Unexplained fevers
- Convulsions
- Serious injuries
- Drugs used

*ENT Health*
- Colds
- Otitis media
- Tonsillitis
- Mouth breathing
- Allergies
- Operations

*Development*
- Age of first sitting
- Age of first walking
- Age of self-feeding
- Age of toilet training

*Speech Development*
- Normal babble at
- Words at
- Phrases at
- Understandable speech at
- Present vocabulary

*Hearing*
- Impairment suspected
  - How observed
  - At what age
- Response to:
  - Conversation
  - Calling
  - Ambient noise
  - Radio
  - Vibratory stimulus

*Maturity*
- Attitude toward children
- Typical play activities
- Tantrums (how controlled)
- Education

* A listing, by groups, of the items about which information should be obtained in taking the history of a child with impaired hearing or other difficulty in communication.

The child suffering from hearing impairment as a result of middle ear infection offers very little difficulty from a diagnostic point of view. The history of earache, a discharging ear, or the story of fluctuation of hearing during and after upper respiratory infections, coupled with the otoscopic evidence of changes in the tympanic membrane, or fluid in the middle ear is evidence enough to call for a consultation with the otologist. The serious diagnostic difficulty lies in those cases suffering from some less-apparent hearing problems. Children born with or early acquiring some damage to the neurologic portion of their hearing apparatus frequently present a very baffling picture. The diagnosis of these children can be greatly facilitated by a careful history.

A review of some 1,100 consecutive cases in the Johns Hopkins Hearing and Speech Clinic (Table II) has shown that the three most critical periods to inquire into are the prenatal period, the paranatal period, and the first year of life. Therefore, one should start by looking into the length of the pregnancy. One should also realize that prenatal viral infections are not limited to rubella, but include other children's diseases, and even severe epidemic viral infections without rash. In our review erythroblastosis resulting in jaundice and kernicterus has been associated with a large group of children with severe hearing problems.

In taking a history of the delivery, points of special interest should be the type of anesthesia, the presentation of the infant, and any complications such as prolapse of the umbilical cord or knotting of the cord. Cyanosis, its severity and duration, should be noted, and the details obtained of any oxygen therapy administered. Actual birth injuries also must be listed.

In the first year of life, the diseases of childhood such as rubella, varicella and rubeola have shown a greater tendency to

TABLE II

A DIAGNOSTIC EVALUATION, WITH RESPECT TO HEARING, OF 1,152 YOUNG CHILDREN*

| Etiologic Factors | Under 6 yr | 6 yr and Over | Etiologic Factors | Under 6 yr | 6 yr and Over | Etiologic Factors | Under 6 yr | 6 yr and Over | Etiologic Factors | Under 6 yr | 6 yr and Over |
|---|---|---|---|---|---|---|---|---|---|---|---|
| Rh factor | 53 | 22 | Cyanosis | 19 | 2 | Varicella | 10 | — | Poliomyelitis | 1 | — |
| Prenatal rubella | 29 | 17 | Cerebral palsy | 21 | 11 | Scarlet fever | — | 2 | Diabetes | 1 | — |
| Prenatal immunization | 3 | 0 | Cerebral degeneration | 15 | 9 | Jaundice | 4 | — | Diarrhea | 2 | — |
| Infantile infection | — | — | Meningitis | 48 | 16 | Pneumonia | 2 | — | Conductive | 36 | 27 |
| Toxemia in pregnancy | 13 | 2 | High fever | 44 | 12 | Neurothropic chemicals | 3 | 1 | Undeterminable | 253 | 85 |
| Developmental anomaly | 18 | 8 | Rubeola | 27 | 11 | Pertussis | 8 | 2 | Normal | 201 | 62 |
| Familial | 24 | 4 | Epidemic parotitis | 4 | 2 | Typhoid fever | — | 1 | Premature | 2 | — |
| Birth injury | 14 | 1 | | | | | | | | | |

* Consecutively examined both otologically and in the Hearing and Speech Clinic of the Department. Only 297 of these children were 6 years of age or older; 855 of them were less than 6 years old.

impair the hearing than in later life. Other viruses also seem to play a similar role. In our review a large group of children with handicapped hearing have been observed, where the only positive finding was a history of high fever, with or without convulsions, during the first 12 months of life; this illness was diagnosed by the pediatrician as a virus infection. This group has shown a high incidence of central auditory damage. Meningitis or encephalitis account for a significant number of hearing problems still, but tuberculosis and influenza have taken the place formerly held by meningococcus infections.

Speech development is the result of a complicated chain of actions and reactions. To develop speech normally a child must have a completely intact auditory system. The anatomy of this system is illustrated in Figure 1. It is easy to see why the child suffering from an atresia or agenesis of the external auditory canal is deaf, or why a child whose middle ear is filled with pus has a hearing loss of a conductive type. The child with a familial deafness may well have partial atrophy of the organ of Corti or of the nerve fibers within the cochlear structure or spinal ganglion. The child with a history of a severe viral infection may have damage almost anywhere in the neuroauditory pathways. The child with erythroblastosis and kernicterus will show damage to the neural elements, particularly in the region of the dorsal and ventral cochlear nuclei and in the brain stem. The child with severe damage from anoxia will frequently show damage to the cortex where there is no demonstrable damage to the organ of Corti; and this has been proven experimentally many times. To hear, one must have a continuity of function from the external ear to the cortex; and hearing, in the broadest concept, means that there must be a comprehension of the multiple sound stimuli presented to the ear. Speech, which develops as a direct result of hearing, can be distorted, retarded or absent as a result of damage to the ear, the auditory pathways or the auditory cortex. It is possible to have a normal middle and inner ear, but still have many of the signs of deafness when the damage lies in the brain.

When reviewing speech development (Fig. 2) it is well to remember that the normal child first cries, second makes play sounds (such as cooing) and then purposeful sounds, especially around feeding time. The child then begins to imitate sounds, which we call the beginning of language, and finally when these imitated sounds bring rewards the child uses them with intent, and that is the beginning of speech. Speech development may be delayed somewhat in a normal child.

In testing the hearing of children two cardinal sins should always be avoided: 1) never confuse vibratory stimuli with sound stimuli; 2) never do a test in such a situation that the child's visual attention can be

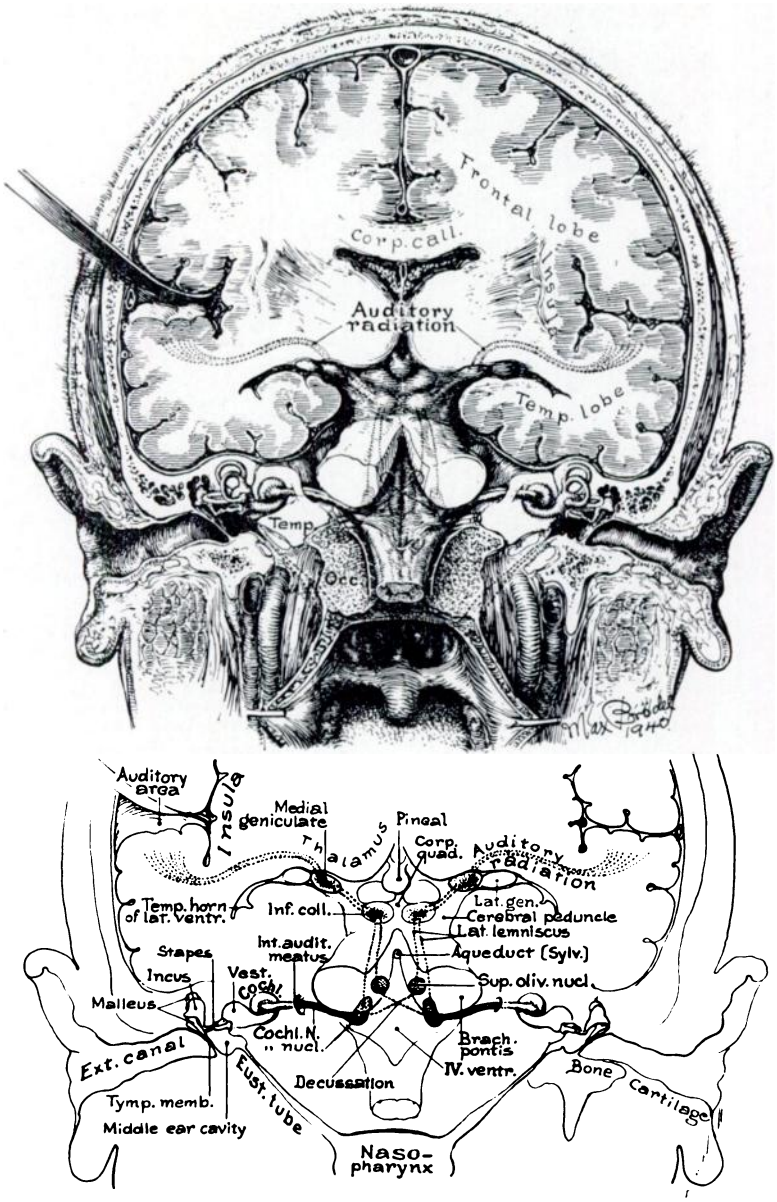


FIG. 1. This drawing by the late Max Brödel shows, in a coronal section of the head, the location of the peripheral organ of hearing and of the principal central connections concerned with audition. (From the 1940 *Year Book of the Eye, Ear, Nose and Throat;* The Year Book Publishers, Chicago.)

attracted by some physical movement. Testing should be done with some reference to the child's age. Here again, it is well to remember that: 1) shortly after birth a child will give a Moro response to sudden sounds; 2) at 3 months of age the normal child will show a "visual interest" in certain environmental sounds, such as the preparation of a feeding, and may also show a response to a tuning fork; 3) by 9 months interest can be aroused by vocal sounds, imitation of these sounds may be attempted, and the baby will also begin about that time to note distant sound; 4) by 12 months there are many

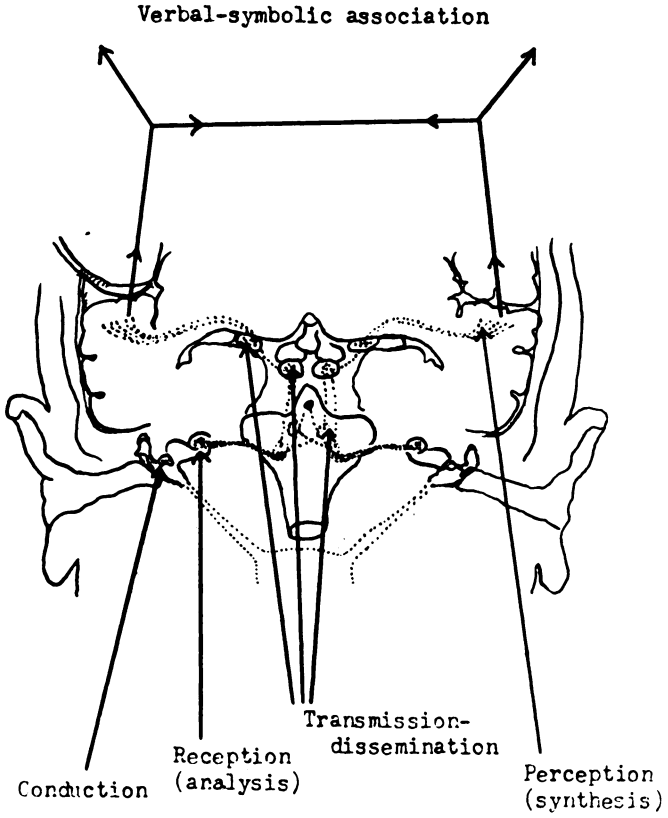


FIG. 2. A schematic representation, based on the anatomy shown in Figure 1, of the functions of the various parts of the auditory system. (From the *Volta Review*, September, 1956.)

evidences of awareness of sound.

For the office, the test equipment can be very simple. The human voice, the human whistle, a cup and a spoon (to simulate house sounds), two or three tuning forks (ranging from 500 to 4,000 cy/sec), and a few noise-making toys (such as squeakers, a drum, a few rattles of different pitch) make a very adequate armamentarium.

One should always remember that the test is to determine aberrant responses or absence of response to sound stimuli. These tests cannot be definitive; they are merely a warning. Remember also that an infant often responds better to a moderate intensity than a large one. An infant will not repeat responses for any length of time and will show very little interest after a few repetitions. Keep in mind that even your gentle cultured voice will create vibratory stimuli when delivered with a shout at 2 inches from the pinna. A bunch of keys hurled to the floor can even cause some vibration in a nearby crib.

In summary, therefore, it might be well to restate briefly some of the more pertinent things to remember:

A child exposed to factors that may result in communicative disorders should always be suspected of hearing damage.

A child with delayed speech development should be watched very carefully.

A child with speech impairment should always have a hearing test.

A child may be deaf as a result of changes in: 1) the external auditory canal; 2) the middle ear; 3) the inner ear; 4) the auditory pathways; and 5) the cerebral cortex. These lesions result in different types of responses to sound stimuli. The child with a normal ear and with a central injury may soon re-

ject sounds, because they are meaningless; his responses at best will be very "in and out," so average test results will be useless. This "in and outness" usually reflects itself in family contention, as to whether the child can hear or whether he is a behavior problem.

Where history and simple office tests cause suspicion of hearing impairment, a definitive examination should always be sought. Delay may mean losing an opportunity to start the rehabilitation early, at the time when the best results can be obtained.

# OFFICE EVALUATION OF VISION IN CHILDREN

**By Frank Duncan Costenbader, M.D.**

I AM PLEASED to be included on a panel discussing the special senses in conjunction with intelligence and certain skills. It would seem that in so grouping these subjects, the eyes, the ears and the other senses have no longer been considered isolated phenomena but as parts of an integrated whole.

It seems important at this early point in the discussion to emphasize the fact that the term "vision" is frequently misused, usually only connoting visual acuity. It should be emphasized that vision in its broadest sense includes visual acuity, the extent of the fields of vision, the normality or abnormality of binocular vision, and the adequacy of the visual associations such as recognition, identification and memory. It seems superfluous to point out that excellent visual acuity, if seen through a gun barrel, is by no means satisfying. Also, that a full, wide field of vision, when the object of interest is blurred, is most unsatisfactory. Having two eyes, each of which is a perfect unit, but not seeing together well and comfortably, is most annoying and handicapping. Finally, referring back to the brain a perfect visual image, which cannot be properly recognized and identified and then correlated with similar images previously received, is a totally frustrating experience.

Thus, for a child "to see well" he must see clearly the thing he looks straight at, he must see widely the things about him, as well as the object of interest, he must have his two eyes properly co-ordinated, and he must be able to recognize, identify and associate this image with related images and activities of the past.

## TELL PARENTS ABOUT EYES

In the practice of pediatric ophthalmology, it has long been apparent that even though an adequate examination is carried out and proper treatment prescribed, the end result is not attained unless the parents understand something of the nature of a child's eyes and their child's specific difficulty. In order to properly clarify such an explanation, I frequently liken the eye to a camera. The aperture of the camera corresponds to the pupil of the eye, regulating the amount of light admitted. The lens of the camera is like the lens of the eye with the exception that the lens of the eye may change in shape, thus changing the focus of the eye, while the lens of the camera cannot. On the other hand, the length of the chamber, which in a camera *is* adjustable to help focus, is of fixed amount in the eye at any one time, and thus is not adjustable. The film or photographic plate of the camera corresponds to the retina and "records" the picture.

Here, however, the analogy breaks down. The camera film is equally sensitive both

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**ADDRESS:** 1605 22nd Street, N.W., Washington 8, D.C.

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