HEARING-IMPAIRED AUTISTIC CHILDREN

R. Jure
I. Rapin
R. F. Tuchman

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The four most prevalent conditions to consider in the differential diagnosis of children with inadequate language development are hearing loss, mental deficiency, developmental language disorder (dysphasia) and autism. These diagnoses are not mutually exclusive; they may share a common etiology or occur independently. Because of the unavailability of biological markers, correct diagnosis of the developmental disorders of early childhood rests entirely on behavioral criteria. Diagnosis is particularly difficult in multiply disabled children in whom two or more of these conditions co-exist.

There is little information about children with both peripheral sensorineural hearing-loss and autistic symptoms. Since each of these disorders has a profound effect on the development of language and communication, their joint effects are likely to be cumulative and devastating. Early identification of both disorders is essential in order to provide appropriate habilitation at the language-learning age, when intervention is known to be most effective (Ruben and Rapin 1988). The purpose of this study is to describe the clinical features that characterize a sample of 46 hearing-impaired autistic children evaluated by one of the authors.

Material and method

All patients in this study were assessed by one child neurologist (I.R.) with a long-term interest in the diagnosis and care of autistic children and of deaf children. Over the years she has systematically reviewed, updated and categorized her patients by diagnostic codes. During the past 10 years she has compiled a computer database, from which 1150 children with a diagnosis of hearing impairment were identified. This population included 387 children evaluated at St. Joseph's School for the Deaf in the Bronx, a population biased toward 'normal' severely to profoundly deaf children, since the school does not accept children with mild or moderate sensorineural hearing-losses, severe or profound mental deficiency, or overt neurological or social deficits. It also included 277 children undergoing private neurological consultation, a population biased by the particular neurologist's interest in communication disorders and autism, and by the presence of many children with complex conditions. Both groups were evaluated from May 1966 to May 1988. Finally, the population included 486 children referred to the Auditory Evoked Response (AER) Laboratory for audiological testing from 1963 to 1972. This third group was biased toward very young and multiply disabled

behaviorally untestable children, since the Laboratory was the first facility in New York City to provide electrophysiological (cortical) audiometry.

From the total population of 1150 hearing-impaired children, 46 (4 per cent) met the criteria for autism, and they form the basis of this report. 28 of these 46 children also belong to a cohort of 314 autistic children evaluated by the neurologist and described elsewhere (Tuchman *et al.* 1991*a*). A further 15 children who had been coded for both hearing impairment and autism but who had inadequate clinical and/or audiological data or were deaf-blind were excluded. They would have brought the total to 5·3 per cent of the 1150 children.

The charts of the 46 children were reviewed and data on perinatal history, medical history, hearing loss, neurological evaluation, language and cognitive function, diagnostic studies and educational experience were extracted from the medical records. There were 30 boys and 16 girls (male:female ratio of 1·86:1). Eight of the children were seen at St. Joseph's School for the Deaf, 20 at the neurologist's practice and 18 came from the AER Laboratory.

Autism is a behaviorally defined developmental syndrome of brain dysfunction, with core symptoms of variable severity. Its etiology is unknown in most cases. Core symptoms in childhood include impaired and atypical socialization, impoverished play, a language disorder affecting comprehension, expression, and non-verbal communication, inadequately modulated affect, stereotypies and a narrow range of interests. Other associated symptoms may include motor abnormalities such as hypotonia, clumsiness and toe-walking, abnormal responses to sensory stimuli, sleep disorders, aberrant attention, and uneven cognitive skills (Rapin 1988, 1991; Tuchman *et al.* 1991*a*). The diagnostic criteria for autism used for inclusion in the study were consistent with those of the DSM-III-R (American Psychiatric Association 1987).

The neurologist who saw all the children reviewed their charts and scored their behavior as either mildly to moderately autistic, *i.e.* capable of some social interaction and testable to some degree; or as severely autistic, *i.e.* severely withdrawn, interacting briefly or not at all and often untestable, except indirectly through questionnaires to parents or caretakers such as the Vineland Adaptive Behavior Scales (Sparrow *et al.* 1984). The first group encompasses the 'pseudo-social' and 'inappropriately interactive' subtypes of Allen (1988) and the 'interactive but odd' and 'passive interaction' of Wing and Gould (1979). The second group includes Allen's 'socially remote' and 'unavailable' subtypes and Wing and Gould's 'aloof' group.

Severity of hearing impairment was categorized as mild (25 to 44dB), moderate (45 to 69dB), severe (70 to 89dB) or profound (>90dB) (average threshold for 0·5, 1·0 and 2·0kHz in the better ear). The diagnosis was based on both pure-tone audiometry and AERs for 37 children, by AERs only for five patients and by audiometry only in four.

Data on severity of hearing impairment, age at diagnosis of hearing loss and age at diagnosis of autism were collated. When exact age at diagnosis of hearing loss was unavailable, we used the age when hearing aids were prescribed. If there was no exact information about age at diagnosis of autism, we used the date of the evaluation at which the symptoms fulfilling the diagnostic criteria for autism were described.

Valid assessment of cognition is difficult even in hearing autistic children, and there is no validated instrument to test hearing-impaired autistic children. The child neurologist routinely assesses each child's ability to perform age-appropriate tasks as part of her neurological examination. We used this assessment, and the parents' report of the child's ability to carry out activities of daily living and any available test data to define three levels of functional intelligence. These were: (1) normal to near-normal intelligence—children estimated to have age-appropriate functional skills or who had a performance IQ of 80 or above; (2) mild to moderate mental deficiency—children with a performance IQ of 50 to 79, or who were below-average but had achieved some developmental skills (in this group were also included children

TABLE I
Diagnosis of hearing impairment and autism

Age at diagnosis (N=46) *Diagnosis*	*Median (mths)*	*Range (mths)*
Hearing loss/autism	24	3-115
Hearing loss	24	2-144
Autism	49	6-214

Delay in diagnosis (N=45) *Order*	*N*	*%*	*Interval* *Mean*	*Range*
Both within 18 mths	19*	42	7	0-18
HL >18 mths before autism	21*	47	53	20-147
Autism >18 mths before HL	5**	11	42	19-72

*Diagnosis of hearing loss in five children later discarded for several years.
**One boy with possibly progressive hearing loss diagnosed at 12 years omitted.

TABLE II
Severity of hearing loss

Threshold in better ear (dB)	*N*	*%*
25-44	1	2·2
45-69	8	17·4
70-89	14	30·4
≥90	23	50·0

for whom there were enough data to state that they were not severely retarded but for whom there was inadequate information to state that they were of normal or near-normal intelligence); or (3) severe to profound mental deficiency—children with few functional skills, none of whom had been tested with standard instruments.

Results

Age at diagnosis

Mean age at first evaluation by the neurologist was 5 years 6 months (range 10 months to 17 years 10 months). 24 children were seen on two or more occasions; their mean age at the most recent visit was 10 years 7 months (range 3 years 5 months to 22 years 7 months). Mean age at last contact for the entire group was 8 years 6 months (range 1 year 9 months to 22 years 7 months).

Ages at diagnosis of hearing loss and autistic features gathered from available records admittedly are imprecise, but informative. Because of extreme outliers, median ages rather than means were computed. Table I shows that while median age for diagnosis of hearing loss was two years, it was four years for autism. This means that for half the 46 children, correct diagnosis was unduly delayed, in some cases for many years. While extreme delay is understandable for a 10-year-old girl who came from a foreign country, it is not easily explicable for three American children in whom neither hearing loss nor autism were diagnosed until five years of age. They had been diagnosed mentally deficient, although only one was severely so.

Both disorders were detected within 18 months of each other in 19 of the 46 children in the study (Table I), although for five children, three of them profoundly deaf, the diagnosis of hearing loss had been subsequently erroneously discarded for several years. A lag longer than 18 months, averaging four years, occurred before autism was diagnosed in 21 of the hearing-impaired children, while deafness was overlooked in five autistic children for up to six (mean 3·5) years. There were seven autistic children whose loss was not detected until they were five years or older and 16 hearing-impaired children whose diagnosis of autism was not made until after that age.

Severity of hearing loss

Only one child had a mild, possibly progressive hearing-loss, but records were inadequate to decide whether the loss was progressive in the other children. Eight children had thresholds in the moderate range in the better ear, 14 were severely impaired and 23 had essentially no hearing (Table II). Thus 37 children (80 per cent) had hearing losses severe enough to preclude the learning of language without special education.

Historical data and etiology

Histories of multiply disabled children often suggest several possible etiologies. This was the case for the present sample, and we are aware that our etiological assignments are tentative and perhaps

arbitrary. The etiology of autism was tentatively considered to be known for 23 children (50 per cent) with a history suggesting an encephalopathic insult, such as congenital rubella, bacterial meningitis, profound immaturity or hypoxia, and for those with malformation syndromes or 'hard' neurological abnormalities. It was considered to be unknown for the remainder, including four children with minor anomalies and six with otherwise uncomplicated genetic hearing-losses. Table III summarizes mutually exclusive conditions that we deemed to constitute the most likely etiology for the child's hearing loss, and in some cases for the autistic symptoms as well.

Six children (13 per cent) had one or more hearing-impaired sibling, none of whom was autistic, and one girl had an autistic second-cousin who was not hearing-impaired. These findings suggest that in these children the hearing loss and autism probably had different etiologies. Three other children, one of whom had multiple anomalies, had distant relatives with hearing losses.

Eight children (17 per cent) had major malformation syndromes (Jones 1988). These included two severely hearing-impaired children with the CHARGE association (Pagon *et al.* 1981), one child with Goldenhar syndrome (Gorlin *et al.* 1963), one with a diaphragmatic hernia repaired at six months after a severe hypoxic episode, one stigmatized child with a coloboma of the retina, one with a cleft of the soft palate and congenital hypoparathyroidism, and one with an excentric pupil and congenital cataract in one eye, a pre-auricular skin-tag and unusual facial features. One child with profound hearing-loss and multiple anomalies (small stature, hypospadias, microcephaly, unusual facial features, transverse palmar crease and malformed pinnae) later developed retinitis pigmentosa and thus may also have had Usher syndrome (Kloepfer *et al.* 1966).

TABLE III
Presumed etiology or associated features*

Presumed etiology	*N*	*%*
Unknown**	17	37·0
Genetic hearing-loss	6	13·0
Congenital rubella	6	13·0
Preterm birth, perinatal problems	6	13·0
Bacterial meningitis	2	4·4
Malformation syndrome†	8	17·4
Congenital hemiparesis	1	2·2

*Mutually exclusive diagnostic assignments.
**Four with minor anomalies.
†One child also had Usher syndrome.

TABLE IV
Neurological findings*

Neurological findings	*N*	*%*
'Hard' signs	11/46	23·9
'Soft' signs	26/46	56·5
Normal	9/46	19·6
Microcephaly	10/42	21·7
Epilepsy	8/46	17·4
EEG epileptiform	7/23	30·4
EEG non-specifically abnormal	2/23	8·7
EEG normal	14/23	60·9
Imaging abnormal	5/11	45·4

*Findings not mutually exclusive.

Neurological findings

Eleven children had 'hard' neurological findings (Table IV), including spasticity, profound hypotonia, ataxia and cranial nerve anomalies. The etiologies were: malformation syndromes (four), rubella (two), perinatal problems (two), genetic deafness associated with profound hypotonia (one) and unknown (two). Only nine children had no 'hard' or 'soft' signs on neurological examination. The charts of the remaining 26 children contained notations of clumsiness, hypotonia, brisk reflexes and toe-walking—'soft' signs that are frequent in autism—although hypotonia in deaf children may also result from vestibular dysfunction (Rapin 1974), which existed in an unknown proportion of the sample.

Microcephaly, *i.e.* head circumference below the second centile on the Nellhaus chart (Nellhaus 1968), was present in 10 of 42 children with available data (Table IV). All but two had a presumed etiology, such as a malformation syndrome (two), congenital rubella (three), meningitis (one) and preterm birth (two, one of

TABLE V
Visual findings (N = 46)*

Visual findings	*N*	*%*
Cataracts (1 unilateral) + rubella retinopathy	4	8·7
Excentric pupil + cataract (unilateral)	1	2·2
Coloboma of retina (+ iris in 1)	2	4·4
Pendular nystagmus	4	8·7
Oculomotor palsies	1	2·2
High myopia	2	4·4
Heterochromia of iris	1	2·2
Retinitis pigmentosa	1	2·2

*Findings not mutually exclusive.

TABLE VI
Estimated severity of autistic behaviors *vs.* cognition (N = 45)*

Autism	*Cognition*		
	Normal/ near-normal	*Mild/moderate deficiency*	*Severe deficiency*
Mild/moderate	8	19	0
Severe	0	9	9

χ^2 $p = 0{\cdot}001$.
*One child had inadequate cognitive data.

whom was found on CT scan at age nine years to have an undefined leukodystrophy after he developed progessive neurological deterioration). Data on visual and other ophthalmological abnormalities are summarized in Table V.

Seizures and EEG data
Epilepsy (two or more unprovoked seizures beyond the neonatal period) occurred in eight of the 46 children. EEG data were available for 23 children: seven were epileptiform, two non-specifically diffusely or focally abnormal and 14 normal (Table IV). Only one of nine children without 'hard' or 'soft' neurological findings had seizures and a paroxysmal EEG. Six of the eight children with a history of epilepsy had paroxysmal EEGs, but two children with paroxysmal EEGs were not reported to have epilepsy. The paroxysmal EEG abnormalities were sharp waves or spike-wave discharges in focal or bilateral distribution, a type of abnormality frequently seen in acquired epileptic aphasia or Landau-Kleffner syndrome (Landau and Kleffner 1957). In the authors' experience, this syndrome is often associated with autistic symptomatology (Tuchman *et al.* 1991*b*). However, none of the five of 38 children with data in whom regression was reported was among those with seizures or an abnormal EEG.

Imaging data
Eleven children had undergone a CT scan; five were abnormal (Table IV). The following abnormalities were noted: a cystic area in the right cerebellum of the child with Goldenhar syndrome; an acute left-sided subdural effusion following *Haemophilus influenzae* meningitis, which was no longer seen on a subsequent MR scan; abnormality of the white matter suggesting a leukodystrophy in a preterm child with microcephaly; a small arachnoidal cyst of the left middle fossa in a child with cleft soft palate and congenital hypoparathyroidism; and minimal focal dilatation of the right ventricle in a child with severe neonatal problems.

Cognitive, behavioral and social findings
Estimates of intelligence must be interpreted cautiously, since performance IQs were available for only 11 children, and since the validity of even these quantitative data for hearing-impaired autistic children is dubious. Only eight of the 45 children for whom data were available were considered to have normal or near-normal intelligence, while nine were severely mentally deficient (Table VI). Estimates of cognitive potential for the remaining 28 are less certain: this group comprises 13 children considered to be mildly to moderately impaired and 15 for whom available but inadequate data suggested less than severe mental deficiency. It is worth stressing, however, that three children had PIQs over 90, one of whose was 115.

Seventeen of the children were reportedly hyperactive as well as autistic. Autistic behaviors were assessed as mild to moderate in 27 children and severe in 18. There was a clear relationship between the severity of the autistic and cognitive

deficits (Table VI): the nine severely mentally deficient children were all severely autistic, while of the eight estimated to have normal or near-normal non-verbal cognitive skills and functional intelligence, none was judged to be severely autistic. The social skills of five of these eight improved significantly with age and appropriate intervention, including the teaching of sign language, although complete remission of autistic behaviors did not occur. Significant behavioral improvement was reported in only one of those with mild to moderate mental deficiency, and in none of the severely autistic children.

There was no clear relationship between sociability and severity of hearing loss: of the 27 children with mild to moderate autistic deficits, five were mildly to moderately hearing-impaired and 22 were severely to profoundly so, while the proportions were four mildly to moderately hearing-impaired and 15 severely to profoundly so among the 19 severely autistic children.

Language and education

Two children could speak: both had mild to moderate hearing losses, one of them possibly progressive. Three other children, none of whom was profoundly hearing-impaired, could speak single words or simple phrases. The other 41 children (89 per cent) did not speak.

Twenty-seven children had received some sign-language training (Table VII). Comprehension of sign language was difficult to judge, but was considered marginally adequate for a hearing-impaired child in only seven cases, none of whom was either severely autistic or mentally deficient. No child was a fluent signer. Of the six children who did not sign, two were severely autistic and mentally deficient, and the remainder moderately so. Four of seven severely to profoundly hearing-impaired children with normal or near-normal estimated intelligence signed phrases, the other three signed words. Echolalia in sign was reported for five of the 21 signers.

There were no data on reading skills for 26 children who were either below age seven years or not in school (Table VII). Of the remainder, eight were non-readers, nine could read words, and only three, two of whom were neither severely hearing-impaired nor severely autistic, could read sentences. The third child was a profoundly deaf, intelligent, mildly autistic boy who attended a school for the deaf.

TABLE VII
Communication skills

	N	*%*
Children exposed to sign language	27	56·5
Do not sign	6	22·2
Sign words only	14	51·9
Sign phrases	7	25·9
Fluent signers	0	
Reading data available	20	43·5
Do not read	8	40·0
Read words only	9	45·0
Read sentences	3	15·0

TABLE VIII
Schooling experience (N = 46)

Educational setting	*N*	*%*
No data	5	10·9
Not yet in school	4	8·7
Hearing program for learning-disabled	1	2·2
School for the deaf	12	26·1
Program for rubella deaf-blind	2	4·3
Program for autistic	5	10·9
Program for multiply disabled	8	17·4
Disastrous experience in many programs	9	19·5
Residential program	6	13·0

Information regarding the type of school attended was available for 41 children (Table VIII). There are no programs specifically designed for autistic hearing-impaired children. Four children were not yet in school, including a girl of nine years who came from another country and who had yet to be assessed. One of the two speaking children was in a program for the learning-disabled, the other was in a program for autistic children. Among the 12 children in schools for the deaf, admission of five had been delayed by two years or more because of diagnostic uncertainty.

Of the eight children with normal or near-normal intelligence, four were in

schools for the deaf and four were in programs for multiply disabled hearing children, where they were receiving more or less intensive training in sign language. 23 children were not in programs for the hearing-impaired. Six children, three of whom were severely mentally deficient, were living in residential programs for the multiply disabled.

The educational experience of nine children was disastrous because of errors in diagnosis and/or the lack of appropriate classes. One boy had attended five different programs by age seven. A diagnosis of severe to profound hearing-loss, made early in three children, was subsequently discarded for several years because of erroneous behavioral tests of hearing; the children were transfered to programs for autistic children where some made behavioral progress, but little or no cognitive progress because of their inability to communicate. Schools for the deaf, lacking experience with even moderately autistic children, discharged deaf autistic children or refused to admit them because they found them impossible to teach. Children whose deafness sometimes was entirely overlooked were admitted to programs for the multiply disabled, where no-one could communicate with them and where they made no progress behaviorally or intellectually and remained languageless. For example one girl was not recognized as being deaf as well as autistic until she was eight years old, despite the fact that she had a younger deaf sister and three hard-of-hearing brothers.

Some children had better experiences. Two teachers of intelligent girls who had been unmanageable in several programs for the autistic and the multiply disabled started to use sign language without knowing that the girls were significantly hearing-impaired. The girls' behavior improved so drastically and they learned to sign so rapidly that they became fully educable, although still autistic. However, another girl whose teachers emphasized her good memory and intellectual potential remained unable to function in a classroom.

Discussion

There are no data on the prevalence of autism in the hearing-impaired population. The fact that a single neurologist found 61 cases of autism and hearing impairment (5·3 per cent) among 1150 cases of hearing-impairment suggests that autism and deafness co-exist more often than by chance alone, since estimated prevalence figures of 1/1000 are quoted for congenital hearing-loss (Chong 1988) and 0·4 to 1·6/1000 for autism (Wing and Gould 1979, Ritvo *et al.* 1989, Sugiyama and Abe 1989). Unfortunately this sample, drawn from three biased populations, does not provide data on the prevalence of autism among hearing-impaired individuals.

The hypothesis that sensory deprivation—including the mild one resulting from middle-ear effusion—may be responsible for autistic symptomatology is unlikely, despite several reports to the contrary (Hayes and Gordon 1977, Konstantareas and Homatidis 1987, Smith *et al.* 1988). A strong argument against this hypothesis is the rarity of autism among the deaf and hard-of-hearing. Another is that there was no correlation between the severity of hearing impairment and of autistic symptomatology in this sample of 46 children.

Etiology is probably genetic in most children with unexplained sensorineural hearing loss (Bodurtha and Nance 1988). Genetics may also play a significant role in the etiology of autism (Ritvo *et al.* 1989, Folstein and Piven 1991), although the prevalence of genetic autism may be lower than that of genetic hearing loss. Hearing loss was definitely genetic in seven of the 46 children and may have been genetic in some of the 17 with an unknown etiology. There are no reliable epidemiological data to determine whether these figures deviate significantly from those for deaf children without autism. The fact that none of the deaf siblings of the six deaf-autistic children in the study was autistic strongly suggests that the two disorders had separate etiologies in these six children. A single child had an autistic second-cousin and no child had a relative who was both hearing-impaired and autistic.

Etiology is unknown and probably diverse for the majority of children with

congenital hearing losses and for those with autism. In contrast, etiology was completely unknown for only 18 of the 46 children with both disorders. Autism denotes brain dysfunction or damage; the more severe and diffuse the brain dysfunction, the more likely it is to produce autistic symptoms. A number of conditions can damage both the ear and the brain and thus be responsible for both impairments.

The best known is congenital rubella, which can cause vasculitis resulting in meningo-encephalopathy (Rourke and Spiro 1967) with microcephaly, as well as endolabyrinthitis, cataract, retinopathy and other malformations. This study spanned the 1964 rubella epidemic. As a result, nearly one-fifth of the children tested in the AER laboratory had congenital rubella, as did six of the 46 children in the present study. In a longitudinal study conducted on 243 children with congenital rubella, Chess *et al.* (1971) found 18 children (7 per cent) with classical autism or autistic symptoms. On follow-up, the autistic symptoms of six of the 18 children had abated (Chess 1977). Chess postulated that autism in these children was caused by chronic infection of the CNS by the rubella virus and was not the consequence of sensory deprivation or mental retardation. She pointed out that there was no difference between autistic and non-autistic children with rubella in the number and severity of their sensory deficits (many were visually impaired as well as deaf), and that the youngsters whose autistic symptoms improved retained their sensory pathology. In our sample, one of six children whose autistic symptomatology improved significantly was a child who had had rubella.

Congenital cytomegalovirus (CMV) infection has also been cited as a possible cause of hearing impairment (Reynolds *et al.* 1974) and of autism (Markowitz 1983). Unfortunately, viral cultures at birth and serological studies were not carried out on the two children with hearing loss, autism and unexplained microcephaly without clear etiology, a clinical picture consistent with possible intra-uterine CMV infection.

Mental deficiency, which also denotes diffuse brain dysfunction, is common in autistic children. This sample indicates that when deafness and autism are associated, there is a higher than expected prevalence of 'hard' neurological findings, congenital anomalies and presumed etiologies than in the typical deaf or autistic child. The etiologies in Table III must be considered tentative, however, since several children had more than one possible etiology, *e.g.* preterm birth, meningitis and hydrocephalus, multiple malformations and Usher syndrome.

Deafness, autism and mental deficiency have been reported in some children with ear malformations, especially the CHARGE association (Rapin and Ruben 1976, Wiznitzer *et al.* 1987). Five of eight children with major malformations had malformed ears. The one with Goldenhar syndrome (Gorlin *et al.* 1963) and the two with the CHARGE association (Pagon *et al.* 1981) were severely to profoundly retarded. It is easy to overlook autistic features in such children who have been admitted to hospital frequently and are so obviously disabled. We stress, however, that a pessimistic attitude during infancy is not warranted for children with ear and branchial-arch malformations, many of whom are hearing-impaired, inasmuch as many are neither mentally deficient nor autistic and have a much better long-term outlook than one might have assumed in infancy (Rapin and Ruben 1976, Wiznitzer *et al.* 1987).

The role of perinatal problems that do not severely damage the brain is controversial. A recent review of the epidemiology of autism (Nelson 1991) concludes that prenatal factors are probably more important than perinatal ones. Evidence for an etiological role of perinatal problems in hearing loss is somewhat more convincing (Clarke and Conry 1979, Stein *et al.* 1983). Five of the six children in the present sample in whom perinatal problems were listed as probably etiological weighed <1500g at birth. Very low-birthweight infants are at high risk for both brain damage (Stewart *et al.* 1981) and deafness (Sanders *et al.* 1985). Very low birthweight is a plausible but unproven cause for hearing loss combined with autism.

Bacterial meningitis, another plausible cause, was reported in two children in the

study. Meningitis can damage the brain (Swartz 1984) and also cause purulent endolabyrinthitis, a well established etiology for profound hearing-loss with vestibular impairment (Landthaler and Andrieu-Guitrancourt 1975, Dodge *et al.* 1984). Its causative role in autism in the non-retarded child with a normal MRI image is conjectural. The other child with bacterial meningitis had multiple potential etiologies, since he was profoundly preterm and hydrocephalic.

This study demonstrates that recognizing hearing impairment and autism in a child with other disabilities is not easy. Yet early diagnosis of both is essential for optimal habilitation. The diagnosis of autism was particularly late, lagging behind that of hearing loss by at least 18 months (four years on average) in almost half the sample.

On the other hand, five children's hearing loss—probably present since early life—was overlooked, and it was falsely discarded in a further five, presumably because lack of language and reaction to sound were attributed to autism. Attributing abnormal behaviors of deaf children to hearing loss and missing the diagnosis of hearing loss when autistic symptoms are prominent are serious clinical errors that highlight the difficulty clinicians often encounter when differentiating hearing loss from autism, especially in children with mental deficiency and other disabilities.

The lesson is that reliable hearing tests must be carried out on all children with inadequate language, even when mental deficiency or autistic behaviors seem to provide an adequate explanation. No child is too young or too disabled to undergo definitive, quantitative assessment of hearing, using brainstem auditory evoked responses. Contrary to some reports, brainstem auditory evoked response audiometry is reliable for detecting hearing loss in autistic children (Klin 1991). In older, co-operative children a standard behavioral test by a pediatric audiologist is most informative. Impedance audiometry should also be carried out to evaluate middle-ear function. Tests should be repeated diligently until there is no doubt about hearing sensitivity; it is never enough to rely on the parents' impression of normal hearing or on the physician's screening in the office.

Deaf children acquire language laboriously unless they belong to the small minority who have signing deaf parents. Autism added to deafness makes the acquisition of language even more problematic, since all autistic preschoolers, including those who have normal or reasonably normal intellectual skills, have a communication disorder (Allen 1988). Sign language (*i.e.* language by eye) has been recommended for hearing autistic children (Schaeffer 1978, Bonvillian *et al.* 1981). Unfortunately, not all autistic children do well with signs because their pragmatic deficits interfere with the communicative use of gestures. Of interest were five of the children who received sign-language education and who had echolalia in signing. Echolalia, which is common in young hearing verbal autistic children, suggests good verbal memory and poor comprehension, but can be used to improve children's communicative abilities (Prizant and Schuler 1987).

This study emphasizes that school placement is a severe problem for deaf autistic children because of the lack of adequate programs. Custodial care is inappropriate because some may not be as severely disabled as first appears. Ideally, these children should be placed in a special class at a school for the deaf that provides not only sign language but also a program of behavioral management.

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Authors' Appointments
R. Jure, M.D.;
*I. Rapin, M.D.;
R. F. Tuchman, M.D.;
Saul R. Korey Department of Neurology, Department of Pediatrics, and the Rose F. Kennedy Center for Research in Mental Retardation and Human Development, Albert Einstein College of Medicine, Bronx, NY.

**Correspondence to second author* at Room 807 Kennedy, Albert Einstein College of Medicine, 1410 Pelham Parkway South, Bronx, NY 10461.

SUMMARY
The charts of 46 children diagnosed as deaf and autistic were reviewed. Nearly one-fifth had normal or near-normal non-verbal intelligence and only one-fifth had severe mental deficiency. The severity of the autistic behavior was related to the severity of the mental deficiency, but not to that of the hearing loss. In 11 of the 46 children, autism went unrecognized for over four years after the diagnosis of hearing loss, and in 10 the hearing loss went unrecognized for several years after the diagnosis of autism. The educational experience of some children was generally disastrous because of the frequently late and incorrect diagnoses and the lack of specialized facilities for hearing-impaired autistic children.

RÉSUMÉ
Enfants à la fois sourds et autistes
Les dossiers de 46 enfants portant à la fois le diagnostic de surdité et celui d'autisme ont été analysés. Près d'un cinquième des enfants avaient une intelligence non verbale normale ou proche de la normale, et seulement un cinquième des enfants avaient une intelligence non-verbale normale ou proche de la normale, et seulement un cinquième avaient une déficience mentale grave. La sévérité de l'autisme était liée à celle de la déficience mentale mais pas à celle de la surdité. Chez 11 des 46 enfants, l'autisme fut despisté plus de quatre ans après la surdité, tandis que chez 10 enfants c'est la surdité qui resta méconnue plusieurs années après le diagnostic de l'autisme. L'expérience éducative de ces enfants fut généralement désastreuse du fait du dépistage tardif et des erreurs de diagnostic, ainsi qu'à cause du manque presque total de programmes scolaires spécialisés pour enfants à la fois sourds et autistes.

ZUSAMMENFASSUNG
Hörgestörte autistische Kinder
Von 46 tauben und autistischen Kindern wurden die Krankengeschichten kontrolliert. Fast ein Fünftel hatte eine normale oder nahezu normale non-verbale Intelligenz und nur ein Fünftel hatte eine schwere geistige Behinderung. Der Schweregrad des autistichen Verhaltens stand in Relation zum Schweregrad der geistigen Behinderung, nicht aber zu dem des Hörverlustes. Bei 11 der 46 Kinder wurde der Autismus erst vier Jahre nach der Diagnose des Hörverlustes festgestellt und bei 10 wurde der Hörverlust erst mehrere Jahre nach der Diagnose des Autismus erkannt. Die Schulbildung dieser Kinder war im allgemeinen schlecht wegen der naurig verspäteten und ungenauen Diagnosen und wegen fehlender spezialisierter Ausbildungsmöglichkeiten für hörgeschädigte autistische Kinder.

RESUMEN
Niños autísticos con alteración de oído
Se revisaron los protocolos de 46 niños diagnosticados como sordos y autísticos. Casi una quinta parte de ellos tenían una inteligencia no verbal normal o casi normal y sólo una quinta parte tenia un deficit mental grave. La gravedad del comportamiento autístico estaba en relación con la gravedad de la deficiencia mental, pero no con la de la pérdida de la audición. En 11 de los 46 niños el autismo no fue diagnosticado en el curso de los cuatro años siguientes al diagnóstico de la pérdida de audición y en 10 casos la pérdida de la audición no se diagnosticó hasta varios años después del diagnóstico de autismo. La experiencia educativa de los niños en general fué desastrosa, pues el diagnóstico era con frecuencia tardío e incorrecto y había falta de facilidades especiales para los niños autísticos con alteración auditiva.

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