

Autism and Hearing Loss

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A group of 199 children and adolescents (153 boys, 46 girls) with autistic disorder was audiologically evaluated. Mild to moderate hearing loss was diagnosed in 7.9% and unilateral hearing loss in 1.6% of those who could be tested appropriately. Pronounced to profound bilateral hearing loss or deafness was diagnosed in 3.5% of all cases, representing a prevalence considerably above that in the general population and comparable to the prevalence found in populations with mental retardation. Hearing deficits in autism occurred at similar rates at all levels of intellectual functioning, so it does not appear that the covariation with intellectual impairment per se can account for all of the variance of hearing deficit in autism. Hyperacusis was common, affecting 18.0% of the autism group and 0% in an age-matched nonautism comparison group. In addition, the rate of serous otitis media (23.5%) and related conductive hearing loss (18.3%) appeared to be increased in autistic disorder. The study emphasizes the need for auditory evaluation of individuals with autism in order to refer those with pronounced to profound hearing loss for aural habilitation and to follow those with mild to moderate hearing loss because of the risk of deterioration.

KEY WORDS: Autistic disorder; hearing loss; serous otitis media; hyperacusis.

INTRODUCTION

The pathophysiology of autistic disorder has been discussed extensively, and there is a solid basis of evidence for an organic background (Gillberg & Coleman, 1992). Different clusters of symptoms characteristic of autism implicate both cortical and subcortical brain areas. Under- or overreactivity to sensory stimuli and a tendency to engage in stereotyped sensory stimulation and/or motor activities are common clinical problems. According to some researchers, these disturbances in sensory modulation are caused by brainstem abnormalities. Studies of autonomic and vestibular

responses have provided support for such abnormalities (see Ornitz, 1987, for review).

Communication in autism is so strikingly impaired that the function of the hearing system has been discussed. In certain instances there is an underlying etiology common both to a hearing deficit and autism, such as in rubella embryopathy (Chess, 1977).

Jure, Rapin, and Tuchman (1991) studied the rate of autism among hearing-impaired children. In a large clinical sample of children with a diagnosis of hearing impairment, they found 5.3% had autism. Among the children with both autism and hearing impairment, neurological findings and/or congenital anomalies were more common than in groups with one diagnosis only. It has been hypothesized that auditory sensory deprivation (especially hearing dysfunction in early childhood) might contribute to the development of autism (Hayes & Gordon, 1977; Konstantareas & Homatidis, 1987; Smith, Miller, Stewart, Walter, & McConnell 1988). In the study by Jure *et al.* (1991), there was no correlation between the severity of hearing impairment

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and of autistic symptomatology. This, and the *relative* rarity of autism among deaf children, was taken to contradict the hypothesis of sensory deprivation as cause of autism. However, possible additive negative effects of simultaneous hearing loss and autistic symptoms, were acknowledged by the authors as was the need for better diagnostic precision in the field of auditory evaluation in autism.

Hearing deficit in autism has been reported to occur at a rate of 0–100%! Student and Sohmer (1978) stated that all of the autistic children they studied had peripheral hearing loss according to auditory brainstem response (ABR) examination. Skoff and his group (Skoff, Mirsky, & Turner, 1980; Skoff *et al.*, 1986) reported that 44–63% of children with autism examined by them had hearing impairment. A similar prevalence (44%) was reported by Taylor, Rosenblatt, and Linschoten (1982). Others have reported very much lower prevalence figures. The present study team has reported a hearing loss prevalence of 13% (Gillberg, Rosenhall, & Johansson, 1983). No hearing loss at all has been reported by Novick, Vaughan, Kurtzberg, and Simson (1980), Rosenblum *et al.* (1980), and Grillon, Courchesne, and Akshoomoff (1989). Klin (1993) reviewed the results of the cited studies, and reported that 33–46% of 170 subjects identified as having autism showed evidence of hearing abnormalities. However, the results are open to methodological critique. The inclusion criteria varied. Some studies excluded children with severe hearing loss and are non-informative with regard to hearing loss in autism. The numbers of subjects were very small in some of the studies, ranging from 6 to 14, and the majority of studies included only a modest number of participants, from 20 to 31. In most of the studies the main aim was to study ABR in autism, reducing the validity of data concerning hearing in autism.

Autism and mental retardation are very often comorbid (Nordin & Gillberg, 1996). Hearing deficits are prevalent in population-based studies of adults with mental retardation (Göstason, 1985). In a recent Dutch study, hearing impairment was found in 6.9% of 0- to 19-year-olds with mental retardation living in institutions and in 12.3% in the total mental retardation sample, all ages included (van Schrojenstein Lantman-de Valk, 1997). Severe hearing deficits or deafness were diagnosed in 6–8% of children with severe mental retardation, and in 2–7% of children with mild mental retardation in Swedish studies (review in Hagberg & Kyllerman, 1983).

In summary, there has long been a need for a larger scale study particularly geared to the prevalence of hearing deficits/hearing loss in autism.

METHOD

Participants

Autistic Disorder

Children and adolescents with autistic disorder undergoing a comprehensive neuropsychiatric and medical workup at the Child Neuropsychiatry Clinic in Göteborg, Sweden, were selected for inclusion in the present study. The study was approved by the local ethical committee. No selection of cases occurred. Audiological evaluation was part of the routine procedure and very few (<10%, not possible to determine the exact number) declined this part of the assessment. The sample was clinically based, but previous studies have shown that population-based and clinical samples of children and adolescents with autism in Western Sweden do not differ significantly with regard to neurobiological background (Gillberg, 1992).

The patients were examined clinically by at least two clinical autism experts, a child psychiatrist or child neurologist and a child psychologist, who agreed on the diagnosis of autistic disorder according to the DSM-III-R criteria (American Psychiatric Association, 1987) at a case conference where all available information about individual patients was reviewed. These clinicians had no knowledge of the results of the audiological investigation at the time of the conference. Interrater reliability for the diagnosis of autistic disorder has been shown to be excellent (100% agreement for autism versus nonautism developmental disorders in a twin study comprising 42 individuals; Steffenburg *et al.*, 1989). The study group consisted of 199 children and adolescents, 153 boys and 46 girls. The mean age at the audiological investigation was 7.0 years (range: 1.2–21.3) for the boys and 8.4 years (range: 1.3–20) for the girls. The diagnosis of autistic disorder was confirmed at or after 3 years of age in all cases.

IQ had been estimated using standardized tests (WISC, Griffiths, Leiter) as part of the routine diagnostic evaluation and educational planning. The testing was performed by clinical psychologists, who were well acquainted with testing of children with severe communication and other developmental disorders. Eighty-seven patients (65 boys, 22 girls) had severe mental retardation (SMR, IQ < 50), 56 (42 boys, 14 girls) had mild mental retardation (MMR, IQ 50–70), and 55 (45 boys, 10 girls) had average intellectual capacity (A, IQ > 70). For one boy the test results could not be retrieved.

Comparison Samples

Results obtained in children with autism were compared with those recorded in samples of children from the general population examined in the same clinical setting with the same methods (Axelsson, Aniansson, & Costa, 1987; Axelsson, Rosenhall, & Zackau, 1994, Kankkunen, 1982) as well as in other centers in Sweden using similar methodology (Augustsson, Nilson, & Engstrand, 1990; Gimsing & Bergholtz, 1983; Hirsch, 1988; Sehlin, Holmgren, & Zakrisson, 1990) and abroad (Martin *et al.*, 1981; Parving, 1983; van Rijn, 1989). All the cited studies have epidemiological designs and contain demographic information about the populations covered.

The comparison group for ABR and for the study of hyperacusis consisted of 57 children and adolescents. They were apparently healthy and had an age range of 4–20 years. Thirty-one were boys (mean age: 12.5 years) and 27 girls (mean age: 10 years). They were studied with the same equipment and procedure as the patients.

Procedure

Audiometry/Auditory Brainstem Response Examination

Audiometry was performed with methods considered appropriate for the individual's chronological and developmental age. The following psychoacoustical tests were used: pure tone and speech audiometry, play audiometry, visual reinforcement audiometry, observational audiometry using informal sound sources, reaction to live voice. The audiometers were calibrated according to ISO 389 (International Organization for Standardization, 1991). In standard pediatric audiology procedures, psychoacoustical threshold estimates are routinely repeated and the best value is chosen.

Auditory brainstem response was performed in 192 of the 199 cases (97%). In most cases the standard level 80 dBnHL was used, but ABR threshold estimation was performed in 30 cases (20 to a level of 20 dBnHL, and 10 to a level of 40 dBnHL). Most children were awake during the ABR testing; only 3 children underwent general anesthesia.

Otomicroscopy/Tympanometry

Otomicroscopy and/or tympanometry was performed in 162 of the cases (81%).

Testing for Hyperacusis

It has been suggested that autism may be associated with "supernormal" hearing as well as with difficulties to tolerate even moderately intense sound levels (Grandin,

1992; review in O'Neill & Jones, 1997). In the present study, it was possible to evaluate the tolerability of a loud sound using the ABR presentation. In ABR a series of loud broadband clicks, presented at a level of 80 dBnHL, is acceptable to healthy subjects but the clicks are often perceived as metallic and slightly unpleasant. Individuals with hyperacusis have difficulties tolerating loud noise, and the clicks must be presented at a lower intensity level, often 70 dBnHL. A majority of the children in the present study were tested with ABR. Those who had normal hearing thresholds were included in a study of hyperacusis. Some individuals with cochlear hearing loss were excluded since they often have recruitment of loudness, a phenomenon similar to hyperacusis.

Degree of Hearing Loss

The extent of hearing loss was defined by the pure tone average (PTA) of the frequencies 0.5, 1, and 2 kHz. The commonly used PTA 0.5–4 kHz was not used here since in some cases with hearing loss the frequency 4 kHz could not be tested reliably. *Mild to moderate hearing loss* was the term applied to those who had a PTA of 20–40 dB HL. *Pronounced hearing loss* was diagnosed in cases with PTAs of 40–70 dB HL. *Severe to profound hearing loss* was diagnosed when the PTA was > 70 dB HL. The latter two groups had hearing loss of such magnitude that aural habilitation was needed. When estimating the prevalence of hearing loss among autistic children, we included only those with permanent hearing loss, sensorineural or conductive. Hearing loss related to serous otitis media (SOM) is generally not permanent and was not included.

Methodological Concerns

The investigation took place during a 12-year period, from 1979 to 1991. During these years, consecutive cases of unequivocal autism were studied. The test procedures were comparable during the entire span of the study. The ABR equipment was exchanged towards the end of the period, but precautions were taken to secure that the procedure was performed in a comparable way. Most of the tests were performed by three audiology technicians who had vast experience in testing children, including children with autism.

One problem inherent in a study of the prevalence of hearing loss in autism is the difficulty associated with testing individuals with the disorder. Problems encountered were deficient cooperation, attention deficits, and cognitive dysfunction. Lack of cooperation may preclude psychoacoustical test procedures. Poor attention and cognitive problems make it difficult to perform re-

liable threshold estimates. The quality of the results of the hearing assessments vary considerably, from very reliable to relatively uncertain. The test procedures used here and the interpretation of the results took this fact into account. If possible, the test was repeated for those children who were difficult to test, and the test result considered most reliable (i.e., with the least variability on the same test occasion) was chosen for the analysis. A majority of the children were tested at one or two occasions. Seventeen of the patients were tested at three different occasions or more. In most instances the children were tested with psychoacoustical tests and ABR at the same test session. This combination allowed comparison of results obtained with completely different methods. The participants were divided into two groups according to the reliability of the testings:

Reliably Tested Group. The first group consisted of 126 children and adolescents who could be assessed reliably. Most of these participants cooperated well and threshold measurement at discrete frequencies (at least above a screening level of 20 dB HL) was achieved. Twenty of the children were assessed with ABR threshold measurements. The conditions when these tests were performed were favorable, allowing estimations down to a level of 20 dBnHL. The cases included in this group constituted the basis for the analysis of prevalence of mild to moderate hearing loss (pure tone audiometry average [PTA] at 0.5, 1, and 2 kHz), isolated high frequency loss, and unilateral hearing loss.

Difficult to Evaluate Group. The second group consisted of 73 children and adolescents who were difficult

to evaluate. The assessments performed allowed a crude estimate of the hearing. Those with severe hearing loss could be identified, and also those with deafness, but not those children who had mild to moderate hearing loss.

RESULTS

Permanent Mild to Moderate Hearing Loss

Mild to moderate permanent hearing loss (PTA 20–40 dB HL or isolated high frequency hearing loss), in all cases sensorineural, was demonstrated in 10 out of 126 cases (7.9%; 95% confidence interval [CI] 4.1–14.5; Fleiss, 1981) (Table I). Slight high frequency loss affecting the frequencies 4–8 kHz was seen in 5 of these cases (1 case, H17, had slight hearing loss in the low as well as in the high frequencies in the left ear only). A falling curve, involving speech as well as high frequencies, was demonstrated in 4 cases (Figure 1). One case had a flat hearing loss in both ears.

Permanent, Severe Unilateral Hearing Loss or Deafness

Two of the participants had pronounced unilateral hearing loss and mild hearing loss in the contralateral ear (2/126 cases, 1.6%; 95% CI 0.3–7.4). One of these cases, a 13-year-old girl, had unilateral conductive hearing loss (Figure 2). The diagnosis was possibly otosclerosis, a disease which her mother had. The other case had unilateral deafness.

Table I. Children with Autism and Slight to Moderate Hearing Loss or Unilateral Hearing Loss^a

Case ^b	Age (years)	Hearing loss, dB HL					
		Right ear			Left ear		
		PTA	4 kHz	6–8 kHz	PTA	4 kHz	6–8 kHz
1 H17 B	10	10	15	30	20	30	20
2 J11 B	2	17	25	—	20	30	—
3 J18 B	15	13	25	15	15	20	35
4 H29 B	16	0	0	40	3	15	25
5 H62 B	3	20	35	—	20	30	—
6 K37 G	6	30	35	—	30	30	30
7 H 9 B	3	17	25	35	23	50	—
8 H79 B	4	28	35	—	23	40	—
9 K12 G	3	27	45	75	22	30	45
10 J 9 B	9	27	50	—	17	30	—
11 K21 G	13	65	45	30	22	10	10
12 H52 B	7	nr	nr	—	22	20	—

^a PTA (pure tone average, 0.5, 1, and 2 kHz) values are given in dBHL, right and left ear. Threshold values at 4 kHz and 6 or 8 kHz are also given. nr = no response, — = not measured.

^b B = boy, G = girl.

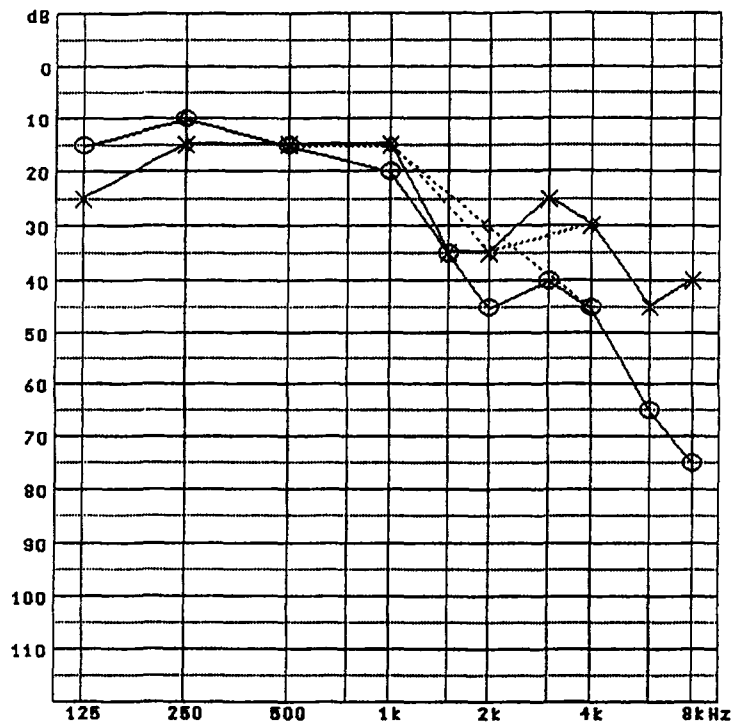


Fig. 1. 10-year-old girl (K12) with mild, sensorineural, slightly asymmetrical hearing loss in the high and mid-frequencies.

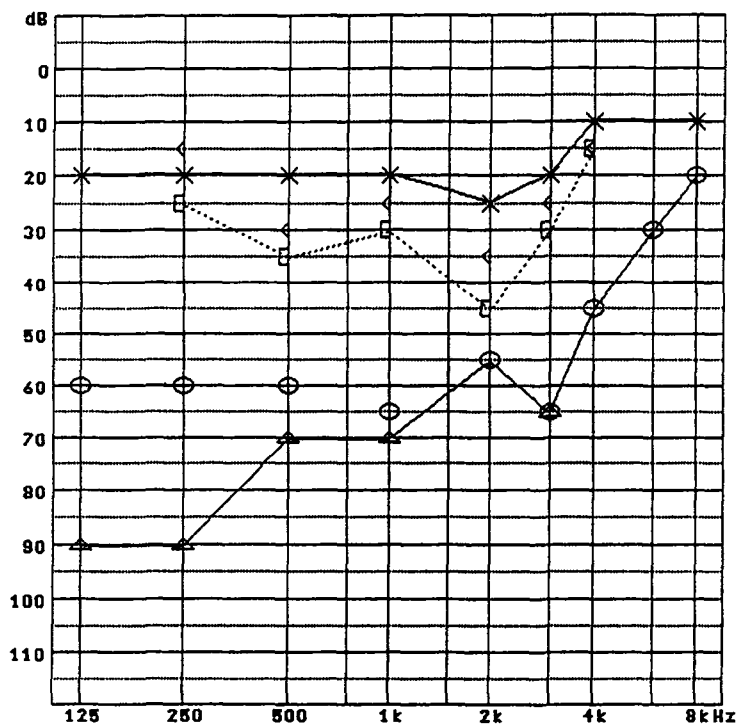


Fig. 2. 13-year-old girl (K 21) with unilateral conductive hearing loss of the right ear, possibly caused by otosclerosis.

Permanent Pronounced Bilateral Hearing Loss or Deafness

Permanent pronounced to profound hearing loss or deafness in both ears was demonstrated in 7 of the entire study group of 199 cases (3.5%; 95% CI 1.6–7.4), all of them boys (Table II). In most of these cases reliable threshold measurements could not be achieved; complete threshold estimates including PTA could only be performed in one case (14). Pronounced hearing loss in both ears (40–70 dB HL) was demonstrated in 3 instances; 2 of these cases had sensorineural hearing loss, and 1 mixed hearing loss (sensorineural and conductive loss). Severe to profound hearing loss (<70 dB HL) was present in 4 cases.

In 3 cases (13–15) hearing residuals could be demonstrated. In another 4 cases (16–19) profound deafness or very uncertain hearing residuals was demonstrated. One case with bilateral sensorineural hearing loss is shown in Figure 3.

Hearing Loss and IQ Level

In the total group of 19 cases with permanent hearing deficits, SMR was found in 8, MMR in 7, and normal intellectual capacity in 4 cases. The 3 boys with bilateral deafness all had SMR; 2 of these boys (cases L8 and L10, Table II) had rubella embryopathy.

The ratio of hearing loss among children with MR was 15/143 (10.5%; 95% CI 6.2–17.0) and among children without MR 4/55 (7.3%; 95% CI 2.4–18.5); that is, there was no significant association between MR and hearing loss, $\chi^2(1) = 0.18$.

Serous Otitis Media

The prevalence of serous otitis media was estimated from the 162 children who were examined with otomicroscopy and/or tested with tympanometry. Thirty-eight of these children (23.5%) had SOM. Twenty-four of them had conductive hearing loss (air conduction PTA >20 dB HL) up to a PTA of 43 dB HL. These children do not include the 19 with permanent hearing loss already mentioned. At the time of the testing, 12 children were treated with transtympanic plastic tubes, and half of these children had hearing loss. Of those whose hearing was tested reliably 23 of 126 (18.3%; 95% CI 12.2–26.4) had SOM with hearing loss.

Hyperacusis

Of the 192 children who were tested with ABR, 21 could not tolerate the standard click level of 80 dBnHL. All of them could be tested at 70 dBnHL. One of these children had a sensorineural hearing loss, which might result in recruitment. Including only those 111 cases who were tested reliably with ABR and who had normal hearing, the prevalence of hyperacusis was estimated at 18.0% (95% CI 11.6–26.8). All comparison cases tolerated the level 80 dBnHL.

DISCUSSION

There are some important problems associated with the evaluation of hearing in individuals with autism, given that they are often notoriously difficult to test. There is

Table II. Children with Autistic Disorder and Pronounced to Profound Hearing Loss^a

Case	Age (years)	Hearing loss, dB HL					
		Right ear			Left ear		
		PTA	4 kHz	ABR	PTA	4kHz	ABR
13 H24	2	—	—	45	—	—	50
14 J 2	6	42	45	40	43	80	40
15 L 2	16	~50	—	—	~45	—	—
16 L 3	6	—	—	nr	—	—	~70
17 L 8	9	nr	—	nr	nr	—	nr
18 L 9	20	nr	—	—	nr	—	nr
19 L 10	15	~90	—	nr	nr	—	nr

^a This group (all boys) is important as aural habilitation is necessary for children with this degree of hearing loss. PTA values and the hearing at 4 kHz are given in those cases that could be psychoacoustically tested. Results from ABR measurements are also given; nr = no response of ABR tests, indicating pronounced to profound hearing loss; — = not measured.

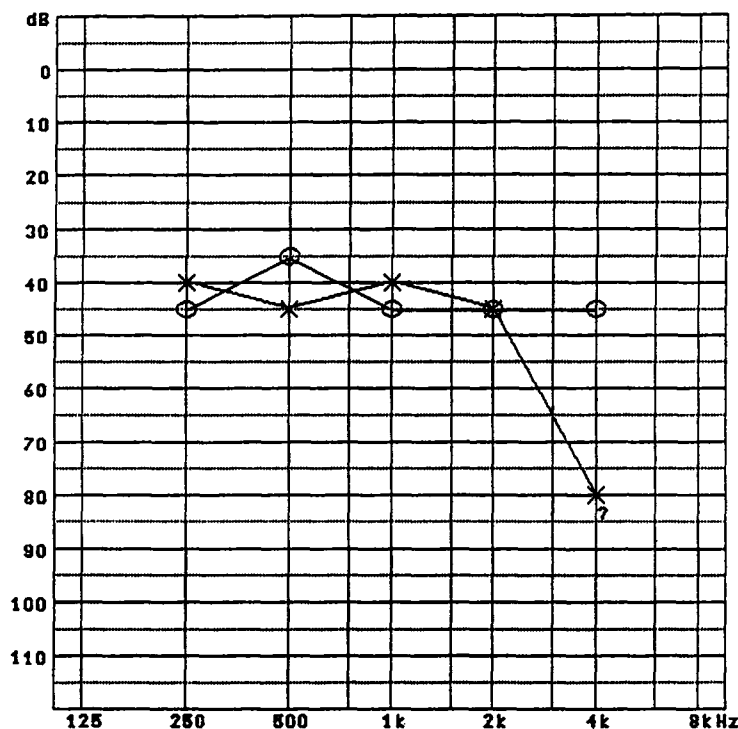


Fig. 3. 6-year-old boy (J2) with bilateral, moderate to severe hearing loss.

a risk for overestimating the prevalence of hearing loss because of difficulties to obtain pure tone threshold at normal levels. By the same token, mild hearing loss can easily remain undetected. Many of the most profoundly autistic children cannot be tested with psychoacoustical methods. The alternative neurophysiological method chosen by most researchers is ABR. With this method it is possible to detect pronounced peripheral hearing loss, but slight hearing loss is difficult to diagnose. One example is hearing loss involving only low frequencies, which is impossible to detect with ABR. Another situation which may complicate hearing assessment with ABR is a concomitant brainstem lesion.

Mild sensorineural high frequency hearing loss is not uncommon in teenage boys. In a study of 18-year-old conscripts, Axelsson *et al.* (1994) found that 14% had hearing impairment, in most cases slight high frequency loss. Since a considerable number of the boys with autism in the present study were teenagers, the prevalence figure of 8% is not remarkably high. However, this figure might be an underestimate since, in spite of all the precautions taken, some children, particularly those with severe autism, could not be tested with the precision needed. It is possible that the prevalence of hearing loss may be higher among children with severe autism, and the total prevalence could therefore be higher

than estimated in this study. The risk of inflating the estimate is small. Five of the 10 boys with slight hearing loss had involvement of only the high frequencies. The 5 remaining children with mild to moderate hearing loss had involvement of the mid and/or the low frequencies as well, which is an uncommon finding in healthy teenagers. Moreover, all autistic children with mild to moderate hearing loss need to be followed to make sure that the hearing loss is not progressive.

The prevalence figure of pronounced to profound hearing loss or deafness (3.5%) is higher than expected in a normal population. In an unselected population of children, profound hearing loss is found in about 0.1–0.2% (Braden, 1994; Davis, Wood, Healy, Webb, Rowe, 1995; Kankkunen, 1982; Martin *et al.*, 1981; Parving, 1983; Sehlin *et al.*, 1990; van Rijn, 1989). Hearing disorders are considerably more prevalent in populations with neurological and developmental dysfunctions (Hagberg & Kyllerman, 1983; van Schrojenstein Lantman-de Valk, 1997). Hearing deficits in autism occurred at similar rates at all levels of intellectual functioning. Thus even though mental retardation is also associated with hearing loss, it does not appear that the covariation with intellectual impairment per se can account for all of the variance of hearing deficit in autism. A need for aural habilitation is considered to arise at an

approximate hearing level of about 40 dB HL, better ear. According to the present study, some of the children with autism have the combination of autism and hearing loss, making the communication problems still more pronounced than either impairment separately. It is therefore very important to assess the hearing of autistic children and to initiate the necessary aural habilitation when hearing is compromised. In most of the autistic boys in the present study such habilitation had already been initiated, applying communication suitable for the children, including sign language.

SOM, resulting in fluctuating conductive hearing loss, has been reported to be common among autistic children (Kontantareas & Homatidis, 1987; Smith *et al.*, 1988). In epidemiological studies of Swedish children from different regions, the prevalence of SOM-related hearing loss has been estimated to be between 4–12%, most often around 5–7% (Augustsson *et al.*, 1990; Axelsson *et al.*, 1987; Gimsing & Bergholtz, 1983). The prevalence of SOM-related conductive hearing loss in this study, 18.3%, therefore seems to be greater than anticipated from the studies cited. The conclusion is supported by the fact that the mean age of the present autism study group was higher than in the studies cited (SOM is more common among preschool children than in school children). Konstantareas and Homatidis (1987) reported that a minor anomaly, low-set ears, is a frequent finding in children with autism. This anomaly could possibly result in eustachian tube dysfunction. However, this observation cannot be confirmed in this study since we did not systematically evaluate the anatomical position of the ears.

Hyperacusis has long been regarded as a very frequent symptom in autism (Coleman & Gillberg, 1985). This study provides preliminary, systematic evidence that almost one in five of individuals with autism and “normal hearing” have hyperacusis, a phenomenon that is rare among children and adolescents who do not have autism.

Conclusions

Severe and profound hearing loss is uncommon among children with autistic disorder. However, the prevalence (3.5%) is at least 10 times that reported in epidemiological studies of children from the general population. Although mental retardation is also associated with hearing loss, it does not appear that the covariation between autism and intellectual impairment per se can account for all cases of hearing deficit in autism. The prevalence of mild hearing loss, affecting low and mid frequencies as well as high frequencies, might also be raised in individuals with autism, although

the findings in this respect were less unequivocal. Hyperacusis was more common than in a comparison group, a finding indicating a need for further research into neural mechanisms underlying this phenomenon. Finally, the prevalence of SOM is probably increased in children with autism, possibly reflecting anatomical anomalies affecting the middle ear. The study highlights the need for audiological evaluation and implementation of audiological habilitation services whenever indicated. It is important to establish ways to integrate habilitation efforts for these complicated cases.

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