



## Autism and peripheral hearing loss: A systematic review



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### ABSTRACT

**Objective:** To systematically review the literature describing the relationship between autism spectrum disorder (ASD) and peripheral hearing loss including literature recommendations for audiological assessment and auditory habilitation in cases where peripheral hearing loss and ASD coexist.

**Data sources:** Published studies indexed in MEDLINE (1948–2011).

**Review methods:** The search strategy identified 595 potential studies. After a review of the titles, 115 abstracts were reviewed and 39 articles were retrieved and assessed independently by at least two authors for possible inclusion. 22 articles pertained to children with ASD and peripheral hearing loss, hearing assessment in children with ASD, audiological habilitation for children with ASD or hyper-responsiveness in children with ASD. 17 further studies were garnered from the reference section of the 22 papers.

**Results:** Controversy exists in the literature regarding prevalence of hearing impairment among individuals with ASD. In cases where ASD and hearing impairment co-exist, diagnosis of one condition often leads to a delay in diagnosing the other. Audiological assessment can be difficult in children with ASD and test–retest reliability of behavioural thresholds can be poor. In cases where hearing impairment exists and hearing aids or cochlear implantation are recommended, devices are often fit with special considerations for the child with ASD. Hyper-responsiveness to auditory stimuli may be displayed by individuals with ASD. Evidence or the suspicion of hyper-responsiveness may be taken into consideration when fitting amplification and planning behavioural intervention.

**Conclusions:** Prevalence rates of hearing impairment among individuals with ASD continue to be debated. At present there is no conclusive evidence that children with ASD are at increased risk of peripheral hearing loss. A complete audiological assessment is recommended in all cases where ASD is suspected so as not to delay the diagnosis of hearing impairment in the event that hearing loss and ASD co-exist. Objective assessment measures should be used to confirm behavioural testing in order to ensure reliability of audiological test results. Fitting of hearing aids or cochlear implantation are not contraindicated when hearing loss is present in children with ASD; however, success with these devices can be variable.

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### 1. Introduction

Autism is a pervasive and complex developmental disorder characterized by impaired social interaction, atypical communication and restricted, repetitive and stereotypical patterns of behaviours, activities and interests [1]. Individuals with autism differ markedly in the number and severity of symptoms displayed [2]. Typical signs of autism include, but are not limited to; speech

and language delay, regression of developmental milestones at 18–24 months of age, avoidance of eye contact, tactile defensiveness and engagement in repetitive and self-stimulating behaviours. See Table 1 in Egelhoff et al. [2] for a more comprehensive list. Approximately 80% of children with autism have some degree of cognitive impairment [3].

Autism is currently classified as a spectrum disorder, which includes autistic disorder, Asperger's disorder, Rett's disorder, childhood disintegrative disorder and pervasive developmental disorder not otherwise specified [4]. Prevalence estimates of autism spectrum disorder (ASD) are controversial. A review by Frombonne [3] reported ASD prevalence rates ranging from 0.7/10,000 to 21.1/10,000, with a median value of 5.2/10,000 (equivalent to: 1/1925) and that prevalence rate increased with study

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**Table 1**  
Summary of studies that evaluate the prevalence of hearing loss in children with ASD.

Study and year	Subject group	Cognitive function	Audiological measures	Audiological results
Rosenhall et al. [8]	199 children and adolescents with ASD	Range	Behavioural audiometry, ABR	Hearing loss at a prevalence rate of 3.5% among ASD population, higher than within general population
Tas et al. [9]	30 children with ASD	Not specified	OAE, ABR	Possible increased hearing loss prevalence among a population with ASD
Jure et al. [10]	61 cases of ASD and hearing loss from 1150 cases of hearing loss	Range	Not available: retrospective review of children with ASD and known hearing loss	Prevalence estimates not possible; co-occurrence ASD and hearing loss may be increased
Gravel et al. [11]	37 children with ASD	High functioning	Behavioural audiometry and computer-assisted threshold measurement, tympanometry, OAE, MEMR	All had normal peripheral hearing and normal peripheral auditory system function.
Tharpe et al. [12]	22 children with ASD	Range	Behavioural audiometry, ABR, OAE, MEMR, tympanometry	All had normal peripheral hearing

ASD: autism spectrum disorder; OAE: otoacoustic emissions; ABR: auditory brainstem response; MEMR: middle ear muscle reflex.

publication year. A review of published ASD prevalence rates in studies since 2000, and primarily since 2006 revealed a median value of 62/10,000 (equivalent to: 1/161) [5]. As ASD prevalence rates over the past decade have shown a rapid increase [4], many argue that the recent increase is not directly due to an increased prevalence of the disorder, but is due to an increased awareness of the disorder and broadening of its diagnostic criteria [6].

This review sought to identify whether a relationship exists between peripheral hearing loss and ASD. Due to the extent and variability of the literature reporting on central issues (primarily central auditory processing) in individuals with ASD and the degree to which input from other modalities integrates with the auditory system at the central level, only the peripheral auditory system was considered.

## 2. Methods

### 2.1. Search strategy

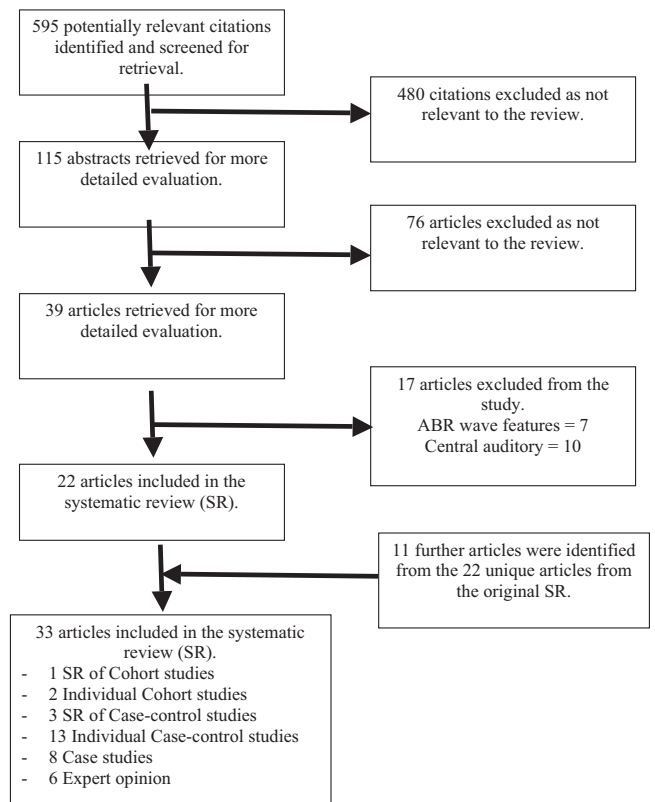
Original published studies on autism and hearing loss were identified with a comprehensive search strategy in MEDLINE (from 1948 to 2011). The search terms used for autism were “autistic disorder [MeSH]”, “autism” and “schizophrenia, childhood [MeSH]”. The search terms used for hearing loss were “hearing loss [MeSH]”, “deafness”, “hearing loss, bilateral [MeSH]”, “hearing loss, high-frequency [MeSH]”, “hearing loss, sensorineural [MeSH]”, “hearing loss, central [MeSH]”, “hearing loss, unilateral [MeSH]”, “hearing impairment”, “deaf”, “auditory perception [MeSH]”, “cochlear nerve [MeSH]”, “cochlea [MeSH]”, “tinnitus [MeSH]”, “vestibulocochlear nerve [MeSH]”, “auditory cortex [MeSH]”, “auditory pathways [MeSH]”, “auditory system”, “auditory processing” “hyperacusis”, “evoked potentials, auditory brain stem [MeSH]”, “hearing [MeSH]”, “cochlear diseases [MeSH]”, “cochlear implants [MeSH]”, “cochlear implantation [MeSH]”, “auditory threshold [MeSH]”, “otoacoustic emissions, spontaneous [MeSH]” and “hearing aids [MeSH]”. These terms were combined with Boolean operators. Reference lists from relevant articles, including other reviews, were searched. Search was limited to the English language and humans (see Fig. 1).

### 2.2. Purpose of study

To determine the relationship between ASD and peripheral hearing loss, hearing assessment in children with ASD, audiological habilitation for children with ASD, or hyper-responsiveness in children with ASD.

The identified studies were assessed for eligibility and were included only if they explored an association between autism and

peripheral hearing loss, audiological assessment or audiological management (deafness, cochlear implantation, hearing aids, or hyper-responsiveness to auditory stimuli). The included studies comprised; 9 Expert opinion, 8 Case series, 16 Individual Case-control series, 3 Systematic Review of Case-control series, 2 Individual Cohort studies and 1 Systematic Review of Cohort studies. The main purpose of this study was to review and collate the English literature relating ASD and peripheral hearing loss. Due to the variability between studies, subject inclusion, test methodologies and outcomes measured, it was not possible to perform meta-analysis of the data. At least two authors independently assessed the titles and abstracts of all studies identified by the searches. The full articles were obtained where they appeared to meet the inclusion criteria (i.e. an association between ASD and



**Fig. 1.** Flow chart of systematic review.

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