The Interface of Paediatric ENT and Autistic Spectrum Disorder: a Complex Conundrum for Otolaryngologists

Authors

Miss Susannah Gimlette¹

Miss Emma Stapleton, FRCS²  ORCID 0000-0002-7763-9705

Institutional affiliations

1. The University of Manchester Medical School, Oxford Road, Manchester M13 9PL

2. Department of Otolaryngology, Manchester Royal Infirmary, Manchester M13 9WL

Author responsible for correspondence:

Miss Emma Stapleton

Department of Otolaryngology, Manchester Royal Infirmary

Oxford Road, Manchester, M13 9WL

Email: emmastapleton@doctors.org.uk

Twitter: @otolaryngolofox
ABSTRACT (150 words)

Introduction: Autism spectrum disorder (ASD) is a lifelong neurodevelopmental condition incorporating complex physical and neurological symptoms including complex sensory symptoms. This review explores the interface between ASD and Paediatric ENT.

Methods: Using the PRISMA guideline, a robust literature search and review was conducted by two researchers. 34 papers were filtered into the final review.

Results: Published literature clearly demonstrates potential for ASD to present in the form of auditory and other sensory symptoms, to ENT surgeons and audiologists who may not fully appreciate this complex condition. Despite this well-documented link, auditory symptoms, auditory processing disorders (APDs) and hearing loss within ASD remain poorly understood.

Conclusion: Improved recognition and understanding of ASD by Otolaryngologists, could enable more effective diagnostic and management strategies for autistic children who present with auditory and other sensory symptoms. In light of the current ‘autism epidemic,’ there is an urgent need for further research on this theme.

MeSH KEYWORDS: Autistic disorder, Audiology, Otolaryngology
Introduction

Autism spectrum disorder (ASD) is an umbrella term encompassing various lifelong neurodevelopmental disorders. It has a roughly 5:1 male to female diagnosis rate\(^1\). Autism was first described in 1943, but was considered ‘childhood schizophrenia’ into the 1960s and gained no official description until the 1980s\(^2\). Rates of ASD diagnosis have steadily increased, and currently estimated to be as high as one in fifty-four children\(^2,3\) a three-fold increase since the early 2000s\(^3\). This has led to what some have called, the ‘autism epidemic’\(^4\) and is likely due to better diagnostic tools and identification techniques.

Typically autistic traits present around 3 years of age, but diagnosis is uncommon under age four\(^5\). This can be due to various reasons. ASD can present with many complex physical and neurological symptoms which can delay diagnosis. As a generalization, these symptoms fall into two categories. Firstly, abnormal behaviour: a gain of skill such as highly developed interests or talents in specific subjects; a preference for social isolation and difficulty understanding emotions; or lack of verbal communication\(^6\). Secondly, an abnormality in sensory processing: this can be auditory; visual; or physical. These abnormalities in sensory processing can be hypo or hypersensitive. Auditory system abnormalities are a prominent problem affecting many in the autistic population. These range from hyperacusis and tinnitus to profound bilateral hearing loss, with a prevalence higher than the national average\(^4\). Hearing abnormalities have been estimated in 33-46% of confirmed ASD cases\(^4,7\). Another study\(^8\) showed that 40% of children exhibit symptoms of sound sensitivity, with decreased sound tolerance (DST) having a lifetime prevalence of 50-70% in autistic people\(^9\). These
symptoms can affect autistic people profoundly, from losing sleep to developing anxiety, to severe interference with school or work\textsuperscript{10}. Rosenhall et al showed that auditory symptoms and hearing loss had similar rates throughout the ASD community, regardless of “intellectual functioning”\textsuperscript{11}.

The recommended method to test autistic children for auditory processing disorders is to check for uncomfortable loudness levels, requiring minimal verbal communication, relying on facial expression and body communication\textsuperscript{10}. Although autistic children require more from healthcare systems, particularly in the otorhinolaryngology field, than their non-autistic counterparts, paradoxically they are less likely to have their needs met\textsuperscript{12}. It has also been recognised that if an autistic child has hearing problems, this can delay the diagnosis of both conditions\textsuperscript{3}. Autistic children frequently present to ENT departments with non-auditory symptoms, including otitis media and allergic rhinitis\textsuperscript{2}. Due to differences in communication skills, these conditions usually present later and more severely, leading to more hospital visits and surgeries\textsuperscript{2}. There are numerous theories behind the physiological cause of ASD, with multiple papers contradicting each other. Mathew et al discuss the ‘early closure of neuroplasticity’ in autistic children, proposing that therefore early intervention with hearing loss can be vital to improving the outcome\textsuperscript{3}.

The aim of this review was to explore the interface of ASD and paediatric ENT, with a specific focus on the potential for ASD to present to paediatric ENT clinics with sensory or other symptoms. There are several terms used within this manuscript, which are defined in Table 1.
Materials and methods

A pilot literature search demonstrated a heterogeneity of literature, unsuitable for incorporation into a systematic review. A scoping review approach was therefore adopted\textsuperscript{20}.

Publications were identified through PubMed, Ovid MEDLINE, and Google Scholar, using the search terms: autism spectrum disorder; autism; autistic; neurodiverse; non-verbal; paediatric; childhood; early; ENT; otolaryngology; hyperacusis; tinnitus; auditory processing disorder; auditory hypersensitivity; deafness. The reference lists of manuscripts identified in the initial search were also screened. There was no limit on the publication dates of studies.

104 papers were initially identified with 34 selected for the final scoping review (figure 1). Inclusion criteria were papers which addressed the themes of both ASD and ENT issues in children. Of those initially excluded, 48 were irrelevant and two were case studies. On inspection of the full papers, 18 were deemed irrelevant, and one was a duplicate.

The included papers were analysed and categorised thematically. These formed the basis for the scoping review.
Results and analysis

Of 34 papers included, there was a variety of study design. Five were descriptive reviews, two retrospective case series, eight prospective research, eight case-control studies, eight systematic reviews, four qualitative research, and one population study (Figure 2).

Classified thematically, manuscripts were classified into exhaustive, exclusive categories. Three addressed the concept of pre-existing ASD in paediatric ENT; fourteen explored ASD and auditory symptoms; ten addressed ASD and auditory processing; five came under the theme of ASD and hearing loss; two addressed ASD and cochlear implantation (Figure 3).

PRE-EXISTING ASD IN PAEDIATRIC ENT

First described by Kanner in 1943\textsuperscript{21}, ASD has gone through various stages of recognition until the latest by NICE in 2013\textsuperscript{22}. With the rising incidence of ASD and studies consistently demonstrating an estimated 40\% or more of diagnosed children showing symptoms of sound sensitivity\textsuperscript{8}, it is key for Otolaryngologists and Audiologists to recognise the condition.

Autistic children have also been shown to exhibit a higher prevalence of anxiety, which can be particularly challenging in healthcare settings\textsuperscript{12}. Sensory symptoms can become a serious challenge for autistic children and their carers\textsuperscript{2}. A study by Fahy et al in a paediatric ENT setting, concluded that healthcare professionals considered parents to be the ‘experts’ when it came to managing and understanding their autistic children’s needs\textsuperscript{2}.
ASD AND AUDITORY SYMPTOMS

A paper by Law et al.\textsuperscript{23} found that the lifetime prevalence of auditory sensory issues (tinnitus, hyperacusis, misophonia and phonophobia) may be as high as 86.6\% in autistic populations.

Hyperacusis is common in autistic populations\textsuperscript{10} and can affect individuals in different ways, from causing anxiety to losing sleep and trouble concentrating\textsuperscript{10}. There are numerous papers theorizing the aetiology of hyperacusis in ASD, from increased neural synchrony\textsuperscript{10} to a small or absent superior olivary complex\textsuperscript{24}. Despite the evidently high prevalence of hyperacusis in ASD, the link remains poorly understood\textsuperscript{25}. Studies conclude that the prevalence of hyperacusis in autistic children is generally around 40\%\textsuperscript{26} (range 29-69\%\textsuperscript{4,10,26,27}) with a lifetime prevalence of DST in ASD of 70-86.6\%\textsuperscript{9,27}. The general population incidence of hyperacusis is between 3.2\% and 17.1\%\textsuperscript{10}. One study found that of 61 children with a diagnosis of ‘troublesome hyperacusis’, autism was subsequently recognised in 13\%\textsuperscript{28}, which is higher than the current national average of around 1.9\%\textsuperscript{2,3}.

Sheerer et al.\textsuperscript{29} discuss how hyperacusis in autistic children can be associated with multisensory integration. This may be due to sensory modulation differences, the brain becoming overwhelmed and unable to filter and process the intensity of the auditory stimulus\textsuperscript{29}. This correlates with the recognised autistic trait of becoming overwhelmed in busy places\textsuperscript{10}. It has been demonstrated that a majority of autistic children (59\% in a survey by Wilson et al.\textsuperscript{26}) are startled by loud noises, in comparison to 15\% of neurotypical counterparts\textsuperscript{29}. Hyperacusis can cause lead to maladaptation in social and academic situations for autistic children, where avoidance and isolation can lead to worsening social skills and anxiety, and difficulties with school work in a loud environment\textsuperscript{26}.  

\url{https://doi.org/10.1017/S0022215122001980} Published online by Cambridge University Press
Tinnitus, an audiological problem causing a patient to hear sounds without the presence of an external auditory stimulus, is estimated to have a lifetime prevalence of 10-15% in the general population. A survey of autistic children demonstrated that 35% experienced tinnitus, about half experiencing it in both ears. This survey used the Tinnitus reaction questionnaire (TRQ) which assesses the associated psychological distress caused by tinnitus. The average TRQ score among autistic people with tinnitus was 27 (formal treatment is recommended for a score higher than 17) demonstrating a significant issue. Interestingly, in this study, 11% of patients reported tinnitus even in the presence of background noise, which is not a classic feature of tinnitus.

ASD AND AUDITORY PROCESSING

Abnormal auditory processing in ASD has been previously proposed, though no definitive explanation has been identified. Bouvet et al. propose that autistic people have more sensitive hearing and are therefore able to detect changes in notes and pitch in music more easily than their neurotypical counterparts. This pattern of cortical processing is thought to influence vision as well as interests, allowing autistic people to hyperfocus on tasks and hobbies, a well-documented autistic trait.

A problem autistic people often face is trouble hearing and understanding speech with background noise. This could be due to concurrent hearing problems, but might also be a result of the functioning aspect of the autistic mind. “Failure of selective attention” is documented well in the literature on autism, and can often be due to sensory overload. In a paper by Alcántara et al. they refer to “auditory scene analysis” where, often in the autistic

https://doi.org/10.1017/S0022215122001980 Published online by Cambridge University Press
mind, different auditory inputs are grouped in the mind into different auditory ‘objects’. This consequently becomes difficult for the mind to focus on just one of these auditory inputs\textsuperscript{35}. This can become distressing for an autistic child, particularly in a loud classroom environment, putting them at an educational disadvantage\textsuperscript{35}.

A paper by Remington et al\textsuperscript{34} attempts to contextualise hyperacusis with neural pathways, proposing that autistic people have an “increased perceptual capacity relative to neurotypical individuals”, allowing autistic people to be able to process information to a higher degree, making it easier to become overwhelmed\textsuperscript{34}. This would coincide with a paper by Myne et al\textsuperscript{36}, where autistic people were found to have a higher acuity for musical notes. Myne et al discuss M100 latency delays in autistic children being largely in the right hemisphere, which commonly processes sound and music\textsuperscript{36}. They found that around 10% of autistic children experienced right-hemisphere 10ms M100 latencies compared with neurotypical controls\textsuperscript{36}. M100 latency delays can be indicative of a disruption in the pathway encoding simple sensory information\textsuperscript{37}. Another paper which supports the theory of right-sided hemisphere bias in autistic people is by Matsuzaki et al\textsuperscript{37}. They used magnetoencephalography to attempt to measure the neurophysiological mechanisms in use for auditory language discrimination. They demonstrate auditory mismatch fields (MMF) amplitudes and latencies, largely in the right hemisphere\textsuperscript{37}. The significantly increased rightward MMF amplitude lateralization they noted in autistic adults is consistent with the paper by Myne et al\textsuperscript{36}, as well as other papers noting the structural and functional abnormal hemispheric asymmetry in paediatric ASD. This indicates a differential path of maturation in the cortical auditory systems in autistic children\textsuperscript{37}.
**ASD AND HEARING LOSS**

Hearing abnormalities are estimated to be present in 33-46% of autistic children, of both sensorineural and conductive origin. The link between hearing loss and ASD, however, has sparse evidence to support it. Some studies estimate the hearing loss in ASD to be ten times higher than in the neurotypical population, others documenting similar rates.

Some of the stereotypical signs of autism, for example, toe walking, hand flapping or ‘sustained odd play’ can result in an earlier diagnosis, but it has been shown that if a child has a hearing impairment, this can delay the diagnosis of autism. In a study by Mandell et al. 382 autistic children and young adults under age 21 were included. The average age of autism diagnosis was 3.1 years. They went on to show that if that child had hearing impairment, their average age of diagnosis was 4.1 years, and was the symptom most likely to delay diagnosis.

Identifying a true dual diagnosis of ASD and hearing loss can be challenging, due to the nature of the symptoms of both conditions. Each condition complicates diagnosing the other. Both conditions independently cause delays in language development, and when concurrent this can worsen the problem, hence early identification being key. In a paper by Gilbert et al., it is proposed that if deafness is suspected under the age of three years, this should also be highly indicative of autism.

**ASD AND COCHLEAR IMPLANTATION**

Diagnosing a child with severe autism with profound sensorineural hearing loss (SNHL) can pose a challenge in determining whether the result of a hearing test is due to true loss of hearing, or communication differences due to autism.
In the 20th century, it was generally thought that autistic children wouldn’t benefit from cochlear implantation, but in the past 20 years evidence has emerged that there is some benefit to be gained. A review article3, showed that out of the 9 studies analysed, 8 demonstrated that autistic children had an overall improvement in their speech expression following cochlear implantation, although the extent of this was highly variable. It was also concluded that, in general, non-compliance was usually related to the severity of ASD3.

There is a move towards cochlear implantation around the age of twelve months. ASD diagnoses are usually made after age three, and hearing problems in ASD are prevalent. It is therefore essential for the otolaryngologists and audiologists to whom children present with a profound hearing loss, to be aware of the potential for a concurrent ASD diagnosis3.
**Discussion**

In the past 20 years, there has been a rapid increase in autism diagnosis, as healthcare professionals gain more understanding of the condition. An autism diagnosis can benefit the child and the support system around them. Families can better understand the context of their child’s symptoms experiences and learn how to deal with challenging situations. More ASD diagnoses will, however, put more pressure on primary and secondary care services.

Hyperacusis and tinnitus can cause elevated levels of anxiety in autistic children, leading to maladaptations detrimental to their education. Through recognition of the impact of hyperacusis, autistic children can receive the support and consideration they need. With such support, autistic children are far more likely to acquire literacy\(^\text{13}\). As the recognition of auditory differences in autistic children improves, education systems will need to adapt accordingly.

The ENT and Audiology problems that autistic children face are multiple and complex, and their true prevalence is often unknown due to the diagnostic challenges which the intersection of the two conditions presents. In non-verbal autistic children, unmanaged DST can cause emotional and sometimes aggressive outbursts. If their autism is undiagnosed and their DST is not suspected, this can present a conundrum, and diagnostic delays can have a profound impact on a child’s wellbeing and education. When these conditions are recognised and acknowledged, simple interventions can improve the child’s quality of life. These might include cognitive behavioural therapy, use of noise-reducing headphones or simply removing the auditory stimulus\(^\text{10}\).
Hearing loss in ASD remains poorly understood, with contradicting published evidence regarding its prevalence. Hearing loss is currently not a recognised symptom of ASD. However if an autistic child has a concurrent hearing loss, this can lead to a delayed diagnosis of both conditions. There is therefore a clear need for Audiologists and Otolaryngologists to be aware of this diagnostic conundrum.

The ICD-11\textsuperscript{42} diagnostic criteria were published in January 2022, to align with the American DSM-5 diagnostic criteria. Within this, there is no mention of auditory processing disorders, hearing loss or other Audiological or Otolaryngological problems. There is brief reference to hyper/hyposensitivity to sensory stimuli including sound, light, taste, smell, pain and touch, making this a vague reference and underplaying the importance of auditory and hearing symptoms. An updated, more specific, diagnostic criteria which recognises the prevalence of auditory processing disorders and hearing loss, could help the affected children. This would promote earlier diagnosis and management strategies.

There is an urgent need for further research, to enhance understanding of ASD in children, and particularly concerning the link between hearing, auditory symptoms, and ASD. This scoping review has identified different key areas that are important in the interface between ASD and paediatric ENT, highlighting potential areas of research and education which could aid future recognition, diagnosis and treatment. We recommend the following:

A) Robust studies to ascertain the true prevalence of hearing loss, auditory symptoms and auditory processing disorders, in autistic children.
B) Ethical trials to identify the incidence of undiagnosed ASD presenting to Otolaryngology and Audiology professionals, in the form of sensory differences and auditory symptoms.

C) Studies to clarify the effect that auditory symptoms, DST and APDs can have on autistic children, including psychosocial as well as educational metrics.

D) Clarification of the aetiology of auditory symptoms in autistic populations.

E) Training Audiology and Otolaryngology professionals to understand and recognise ASD, and to adjust their practices appropriately, especially when assessing children with unexplained hearing loss, auditory symptoms, APDs and DST disorders.

F) Engagement with autistic advocates and populations, for enhanced insight to the complex interface of challenges and symptoms.

G) More research into effective treatments for autistic children with auditory symptoms and hearing differences, with a multidisciplinary approach.

While Fahy et al\(^2\) recognise that healthcare professionals should consider parents of autistic children to be the experts of their condition, there is a growing body of evidence that Otolaryngologists also need to be experts. Autistic children’s sensory abnormalities frequently present to ENT and audiology clinics and would best be managed in the context of a neurodevelopmental disorder than a primary ear disorder. A multi-disciplinary team approach to these cases is essential to provide the best all-round care.
Conclusion

This scoping review identifies key themes within the interface of autism spectrum disorder and paediatric ENT, highlighting areas for future research. Improved recognition of autism concurrent with auditory symptoms, auditory processing disorders, hearing loss and decreased sound tolerance, would lower the average age of diagnosis of both conditions, leading to better social and educational outcomes for these children.

A consistent approach or mandatory guide for checking auditory symptoms and hearing following a diagnosis of ASD could help with the recognition of concurrent issues. In the same manner, Paediatric Otolaryngologist and Audiologist awareness of autistic behaviours, traits and presentations, and the potential prevalence of the overlap, would also be beneficial.

The diagnostic criteria for autism in the UK are evolving, but the most recent change does not acknowledge the importance and impact of auditory symptoms and hearing differences in ASD. Improved diagnostic criteria could be greatly beneficial for awareness and recognition, leading to better recognition and more efficient diagnostic and management strategies.

With an increasing body of literature strengthening the link between the conditions, improvements in the way we recognise, diagnose, and manage hearing differences and auditory symptoms in autistic children, could change the face of ASD.
ACKNOWLEDGEMENTS

Acknowledgments

This research was supported by the National Institute for Health Research Manchester Biomedical Research Centre.

Financial Support:

No funding was received for this work.

Competing Interests:

The authors declare none

Data sharing and data availability statement

Not applicable

Authorship statement

ES designed the work; SG and ES acquired and analysed data, drafted, revised, and approved the manuscript; SG and ES agree to be accountable for all aspects of the work.
REFERENCES


Disorders. Audiol Res 2021;11:547


Heterogeneity and Treatment Options. Neurosci Bull 2017;33:183–93


Summary

- A link between autistic spectrum disorder (ASD) and conditions which commonly present to Paediatric ENT (hearing loss, tinnitus, hyperacusis, auditory processing disorders) is well recognised in the literature.

- However, our understanding of the interface between these conditions is limited, leading to diagnostic delays for both ASD and ENT problems.

- This scoping review identifies key themes within the interface of autism spectrum disorder and paediatric ENT, highlighting areas for future research.

- Better recognition of ASD concurrent with auditory symptoms and disorders, would lower the average age of diagnosis of both conditions, leading to better social and educational outcomes for these children.

- With an increasing body of literature strengthening the link between the conditions, improvements in the way we recognise, diagnose, and manage hearing differences and auditory symptoms in autistic children, could change the face of ASD.

FIGURES AND TABLES

Table 1. Definitions of terms used within this manuscript

Figure 1. PRISMA flow diagram displaying the systematic search methodology

Figure 2. Chart showing literature types included within the analysis

Figure 3. Thematic analysis of the included literature
### Table 1. Definitions of terms used within this manuscript

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Autism spectrum disorder (ASD)</td>
<td>ASD is a neurodevelopmental disorder describing a collection of symptoms: communication, behaviour or sensory in origin. It affects how people with the condition communicate, socialise, learn, and behave. It can be diagnosed at any age, but symptoms must have been present since early life. While there is no cure, various treatments and services can relieve the burden.</td>
</tr>
<tr>
<td>Auditory processing disorder (APD)</td>
<td>The British Society of Audiology describe three APD categories; acquired APD, secondary APD or developmental APD. Acquired APD arises from a known neurological event, such as infection, trauma or a stroke. Secondary APD is a result of a genetic cause or peripheral hearing impairment. Thirdly, and more relevant to this scoping review, is developmental APD, where the APD is present from birth, exists with normal audiometry and has no identified aetiology.</td>
</tr>
<tr>
<td>Hyperacusis</td>
<td>Hyperacusis is a decreased sound tolerance (DST) disorder, where normal or non-threatening sounds appear louder and intolerable. Depending on the person and severity, this sensation can be painful or frightening or just unpleasant. Dislike or fear of specific sounds comes under the terms misophonia and phonophobia.</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>Tinnitus is another DST disorder, described as the auditory perception of a sound despite the absence of an external environmental stimulus. It can present unilaterally or bilaterally and to varying degrees, from only noticeable in quiet rooms to severely impacting on day to day life.</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>Hearing loss can be experienced unilaterally or bilaterally. It can either be sensorineural or conductive in nature. Its prevalence increases with age, but it can be present from childhood or even birth.</td>
</tr>
<tr>
<td>M100 latency</td>
<td>M100 latency is a measured magnetoencephalography response and is the expected delay in the auditory evoked cortical response, representing 100ms. This latency measurement can give an indication of white matter conduction and the speed of transmission between the synapses. It is a measurement that is often used when checking cortical responses to different stimuli, mainly auditory.</td>
</tr>
</tbody>
</table>
Figure 1. PRISMA flow diagram displaying the systematic search methodology

- Papers identified through database searching (n=104)
  - Additional record identified (n=0)
  - Papers excluded as case reports (n=2)
  - Papers excluded as irrelevant content (n=48)
  - Papers excluded as duplicate (n=1)
  - Full text papers excluded as content irrelevant (n=19)
- Full text articles assessed for eligibility (n=54)
- Studies included in scoping review (n=34)
Figure 2: Chart showing literature types included within the analysis

- Descriptive review
- Retrospective case series
- Prospective research paper
- Case control series
- Systematic reviews
- Population study
- Qualitative research paper
Figure 3. Thematic analysis of the included literature

- Pre-existing ASD in paediatric ENT
- ASD and auditory symptoms
- ASD and auditory processing
- ASD and hearing loss
- ASD and cochlear implantation