Volume 14 • Number 1 • March 2024 ISSN 2083-389X

Journal of Hearing Science®

INDEX COPERNICUS INTERNATIONAL

INSTITUTE OF SENSORY ORGANS www.journalofhearingscience.com

Journal of Hearing Science®

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Journal of Hearing Science® is published by the **Institute of Sensory Organs** (Kajetany, Poland) in cooperation with the **Institute of Physiology and Pathology of Hearing** (Warsaw/Kajetany, Poland) – the leading Polish scientific institute in otolaryngology, audiology, phoniatrics, and related fields. The journal is affiliated with the **Society of Polish Otorhinolaryngologists, Phoniatrists, and Audiologists**.

Volume 14 • Number 1 • March 2024 ISSN 2083-389X

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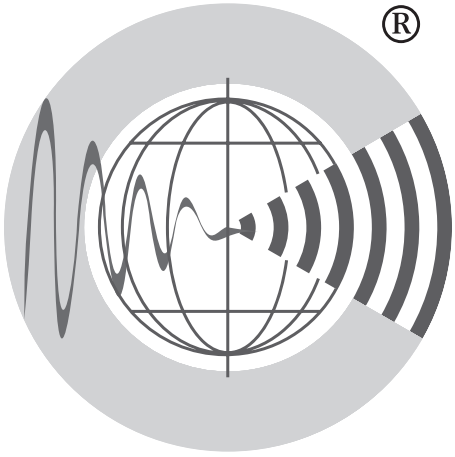

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Dear Colleagues,

Welcome to the latest issue of the *Journal of Hearing Science*. This issue presents a collection of papers that exemplify the breadth and depth of contemporary hearing science, from innovative hypotheses to comprehensive reviews and insightful original research.

We begin with a compelling hypothesis paper that proposes a novel method for integrating speech testing into hearing screening programs for young children. As early detection of hearing impairments and delays in speech development is crucial, this method could significantly enhance the accuracy and efficacy of screening programs, ensuring that children receive the support they need at the earliest possible stage.

Our review paper on the effects of cochlear implantation on the development of speech in children with Usher syndrome provides a thorough examination of existing research on the topic. Usher syndrome, a complex genetic condition that affects both hearing and vision, poses unique challenges. This review highlights the critical importance of early intervention with bilateral cochlear implantation in children with profound to severe hearing loss in promoting optimal speech development. The text offers valuable insights for clinicians and researchers alike.

This issue also features two original research articles that delve into diverse aspects of auditory science. The first explores how musicians' self-assessment of their musical abilities correlates with their auditory processing skills. The second original article presents a review of a substantial number of cases involving major salivary gland tumors from one hospital.

We hope this issue stimulates further research and discussion among our readers, fostering a deeper understanding of the complexities of hearing science and its practical applications. As always, we welcome your feedback and look forward to your contributions in future editions.

With kind regards and greetings,

Prof. Henryk Skarzynski, M.D., Ph.D., Dr. h.c. multi

Hypothesis papers

ISSN: 2083-389X
eISSN: 2084-3127

J Hear Sci, 2024; 14(1): 9–17
DOI: 10.17430/jhs/182820

A PROPOSED METHOD FOR SPEECH AND LANGUAGE TESTING WITHIN A HEARING SCREENING PROGRAM FOR CHILDREN STARTING SCHOOL

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Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

Abstract

This article sets out a proposal for how one might expand hearing screening in school children so as to include testing of speech mastery. From 2008 to 2019, the Institute of Physiology and Pathology of Hearing (IPPH) implemented a program to screen the hearing of Polish children starting school, covering over 1 million children. Its success prompted the development of a screening model to cover the entire process of verbal communication. One result was the "Equal Start in Education", a program implemented from 2019 to 2022 in the Lublin province, otherwise known as the Lublin Program for Early Detection and Therapy of Communication Disorders in Children Starting School. It was conducted by the University Children's Clinical Hospital in Lublin and the Maria Curie-Sklodowska University, in cooperation with IPPH, and was designed to assess hearing, auditory processing, voice, and speech-language. The program was successful in addressing various issues related to the screening of all communication skills. Here we present findings from the program and propose a testing procedure used to evaluate speech. The screening procedure might be applied more widely in the future, and could diagnose those children requiring audiological and speech-language therapy.

Keywords: hearing screening • speech screening • speech therapy

PROPONOWANA METODA BADANIA MOWY I JĘZYKA W RAMACH PROGRAMU PRZESIEWOWEGO BADANIA SŁUCHU U DZIECI ROZPOCZYNAJĄCYCH EDUKACJĘ SZKOLNĄ

Streszczenie

Artykuł przedstawia proponowany sposób poszerzenia badań przesiewowych słuchu u dzieci w wieku szkolnym i włączenie do nich testów opanowania mowy. W latach 2008–2019 Instytut Fizjologii i Patologii Słuchu (IFPS) wdrożył program badań przesiewowych słuchu u polskich dzieci rozpoczynających edukację szkolną, który objął ponad 1 milion dzieci. Powodzenie tego programu stanowiło impuls do opracowania modeli badań przesiewowych obejmujących cały proces komunikacji werbalnej. Jednym z efektów był program „Równy start w edukacji" wdrożony w latach 2019–2022 w województwie lubelskim, znany też pod nazwą „Lubelski program wczesnego wykrywania i leczenia zaburzeń komunikacyjnych u dzieci rozpoczynających edukację szkolną". Program był prowadzony przez Uniwersytecki Szpital Dziecięcy w Lublinie i Uniwersytet Marii Curie-Skłodowskiej we współpracy z IFPS. Jego celem była ocena słuchu, przetwarzania słuchowego, głosu i mowy-języka. Program z powodzeniem rozwiązał szereg problemów związanych z przesiewowym badaniem wszystkich kompetencji komunikacyjnych. W tym artykule prezentujemy wnioski z programu i proponujemy procedurę testową do oceny mowy. Procedurę badania przesiewowego można zastosować na szerszą skalę w przyszłości, może ona wykrywać dzieci potrzebujące terapii audiologicznej i logopedycznej.

Słowa kluczowe: badanie przesiewowe słuchu • badanie przesiewowe mowy • terapia mowy

Introduction

In modern terms, speech and language is "a set of activities that, with the help of language, a person performs in learning about reality and communicating its interpretations to other participants in social life" [1]. This means there are three basic functions that language performs: interactional, cognitive, and social. Verbal communication also requires linguistic and communicative competence, as well as perceptual and implementation skills [2]. If we could recognise speech disorders in children at an early age, this would allow early therapeutic intervention. In turn, this could prevent potential communication, educational, and emotional problems, increasing the well-being of individuals and society in general. This is only possible through a large-scale screening program.

The significance of speech screening can be appreciated by looking at similar tests for vision and hearing. Wide-ranging research has shown that when vision and hearing defects are detected early, many unwanted consequences can be prevented. Thus, 35 European countries have implemented national programs for vision screening in children, while 33 of them screen the hearing of children [3]. Poland's program of universal hearing screening in newborns is the largest preventive health program in the country. It has been fully implemented since 2002, although hearing screening began in 1995–1998 in various centers in Poland using otoacoustic emissions and auditory evoked potentials methods. The program was begun by a team from the Institute of Physiology and Pathology of Hearing (IPPH) following an order from the Minister of Health.

The result has been that, over the last 16 years, Poland has implemented both national and regional programs for hearing screening in school-aged children [4–7]. More than 1 million children have been covered. In 2008–2016, IPPH, in cooperation with the Agricultural Social Insurance Fund, conducted a study of children from rural areas. It found that 1 in every 5–6 children had temporary or permanent hearing disorders which hindered learning and communication. In about 65% of detected cases of childhood hearing disorders, their parents or guardians were unaware of the problem. The children underwent permanent or periodic care from an audiologist, phoniatrist, speech-language therapist, psychologist, or educator [4–6,8]. In more detail, the study found that, in children aged 6 to 9 from rural areas, 19.5% had a positive hearing screening result (defined as a hearing threshold worse than 20 dB HL at one or more frequencies). In children aged 10 to 13, positive results were found in 10.5% [6]. A Canadian study found an even higher prevalence of hearing problems in young children [9]. No doubt the higher prevalence of hearing disorders in younger children is due to a higher incidence of middle ear disease and respiratory infections, combined with limited access to pediatric care in rural areas. Among the conclusions of the IPPH study was the need for systematic monitoring of the hearing status of children, especially younger ones, as well as the need to provide hearing health education for children, parents, and teachers.

In 2017–2019, IPPH implemented a program of hearing screening in children who were starting school in the Mazovia region [10]. The program consisted of four modules: an information campaign, educational meetings for parents/guardians and teachers, training for medical personnel, and hearing tests in children (performed by video-otoscopy and pure tone audiometry). A unique feature of this program was its coverage of the entire population of first-grade students attending elementary schools in the Mazovia Province. In this study, 19.2% of the children were found to have a positive hearing screening result, and they were referred for specialised audiological diagnostics. More commonly, unilateral rather than bilateral hearing disorders were found, i.e. those disorders that are more difficult for parents and teachers to recognise. The IPPH researchers concluded that hearing screening should become standard for children starting school.

Of particular interest, during the Mazovian screening program an assessment was also made of the frequency and nature of voice disorders of the first-grade children [11]. In this case, the assessment was carried out by parents in response to a questionnaire. Data from 7631 questionnaires showed that voice disorders were present in 12.8% of children, with dysphonia more common in boys than in girls.

Extending this idea, in 2019–2022 the Lublin "Program for Early Detection and Therapy of Communication Disorders in Children Starting School" was implemented. Called "Equal Start in Education," it was conducted throughout the Lublin Province by the University Children's Clinical Hospital in Lublin and the Maria Curie-Sklodowska University in cooperation with IPPH.

Novel aspects of the program were that, in addition to looking for peripheral hearing disorders, screening was also performed to detect central auditory processing disorders and voice and speech-language disorders. The program reflected the importance of diagnosing children's overall communication skills rather than focusing just on hearing. The program also involved providing therapy for children with hearing disorders of central origin [12]. The introduction of this test, covering all aspects of speech, increased the figure for diagnosed speech problems appreciably. Whereas previous hearing screening of children of a similar age found problems in about 20% of the children tested [13], in the new expanded program a total of 35% of children were referred for further specialised diagnosis due to suspected speech-language disorders.

To complete the picture, hearing screening looked for both peripheral and central hearing disorders. Pure tone audiometry was used to measure thresholds for air conduction at 0.5, 1, 2, 4, and 8 kHz. For central auditory processing, tests involved the frequency pattern test (FPT), a test assessing the degree of speech understanding in noise, and the dichotic digit test (DDT). All tests were performed using a standardised device called the Sensory Testing Platform. This device is used in hearing screening, both in Poland and in other countries, and can be used for telemedicine [14].

From December 2019 to May 2022, hearing tests were performed on 28,580 children as part of the Lublin Program for Early Detection and Therapy of Communication Disorders in Children Starting School ("Equal Start in Education"). The percentage of children diagnosed with peripheral hearing disorders (temporary or permanent) was 8%, while the percentage of children with suspected auditory processing disorders was 34.8%. These figures are largely consistent with those from other Polish and foreign hearing screening programs. Similar data were presented by Feder et al. [9], who found peripheral hearing disorders in 7.7% of children from a representative sample of the Canadian pediatric population aged 6 to 19. In a U.S. study, one conducted on children aged 3 to 10, peripheral hearing impairment was found in about 11% of participants [15]. Some researchers report higher figures [16], but it must be noted that study protocols, research methods, and adopted criteria may vary and lead to slightly different estimates. A study conducted by IPPH and published in 2015 showed that central auditory processing disorders occur in about 11% of children aged 7 to 12 years [17]. It should be emphasised, however, that at that time only

the DDT test was used and a rather restrictive criterion was adopted (fifth centile as a cutoff point), which significantly reduced the number of positive results (i.e. those indicating the probable presence of CAPD). Current normative values for tests assessing central auditory processing can be found in Czajka et al. [18].

Review of speech and language screening in children

The review presented below deals only with screening tests, i.e. those that do not set out to provide a firm diagnosis of a disorder, but are sensitive enough to signal a problem that requires further investigation. From our working definition of speech and language adopted earlier, it follows that screening should include all components of verbal communication, that is, comprehension and speech expression, the latter involving pronunciation, vocabulary, correct grammar, verbal fluency, and narrative skills. This is a difficult but not impossible task. The problem as we see it is to identify the most important elements of speech and determine a suitable rating system. First we discuss the solutions adopted so far internationally and in Poland.

Worldwide, there are dozens of screening tests for assessing speech competency. The most common in the literature, and available on the Internet, are English-language tests. This reflects the ubiquity of English, well-developed systems of speech therapy in English-speaking countries, good theoretical development, and high levels of practical performance. Many tests developed in English are available in other languages (Spanish, Portuguese). However, the following deals only with English-language and Polish tests.

Screening tests are often designed for children of preschool and early school age. The skills assessed involve speech perception (comprehension), speech construction (articulatory, systemic, and pragmatic skills), and prosody (voice and fluency of speech). The age range usually covers 0 to 21 years of age, although in making a speech evaluation, a speech therapist will typically use a variety of tests depending on the age of the person being tested. Typically, speech is assessed in terms of the level of development of perceptual, motor, and cognitive skills. Tools can be completely standardised, partially standardised, or have no standards (in which case the interpretation of the results requires specialist judgment). Most investigations involve questionnaires, in which subjects are presented with material requiring them to repeat, name, or observe, after which they must respond verbally. Tests mostly take about 20 minutes. Children who fall below a certain rating are referred for further diagnostics.

Most screening tests fall into the following four types, based on the mode and scope of the examination.

1. Parents provide information based on a questionnaire presented by a speech-language therapist or electronically;
2. Parents make a report, which is then interpreted by a speech-language therapist;
3. A speech-language therapist or other professional evaluates speech against a background of overall behavior;
4. A speech-language therapist or other specialist evaluates speech and language in isolation.

The first three categories can be used with preschool children. The fourth category can be applied to children up to the age of 21. Below is an overview of selected diagnostic tools in terms of these categories.

Method 1. Parents provide information

A representative test in this category is the *Speech-Language and Learning Parent Questionnaire for Children 5 and Above* developed by the Foundations Developmental House of Arizona (USA) [19]. The questionnaire consists of 50 questions aimed at parents of children 5 or more years old. The questions are divided into sections that address: general observations about speech disorders, the presence of speech disorders in the family, the child's health and development, voice quality and speech fluency, hearing and learning skills, sensory and motor features (such as the presence of tactile or gustatory hypersensitivity), social behavior, and other relevant information about the child (e.g., disorders diagnosed by other specialists, the child's abilities, family situation, etc.). The test does not involve scoring but instead requires expert judgment of whether speech disorders are present.

A comprehensive and thorough overview of speech-related screening assessment tests for children younger than 5 years (and as young as 12 months) is provided by a 2015 report, *Screening for Speech and Language Delays and Disorders in Children Age 5 Years or Younger: A Systematic Review for the U.S. Preventive Services Task Force* [20]. The report evaluates dozens of diagnostic tools, assessing their reliability for diagnosis and intervention in early speech problems. The diagnostic sensitivity of some of them was rated to be as high as 100%.

Method 2. Reports made by parents

An example of a widespread test in which mothers self-report their child's development, including speech, is the *Minnesota Child Development Inventory* (MCDI) [21]. The MCDI measures the development of children between the ages of 36 and 60 months. The estimated sensitivity of this test is 75%, perhaps comparable to the possible 100% accuracy of assessment by professionals. Children are assessed in five categories of development: cognitive, language, motor, social, and adaptive skills. MCDI is an example of a tool in which speech is one of several areas of assessment.

Method 3. A speech therapist evaluates speech against behavior

The most popular screening test in this category is the *Denver Developmental Screening Test* (DDST), commonly known as the *Denver Scale*. Developed by Frankenburg and Dodds, it is a test for screening cognitive and behavioral problems in preschool children [22]. The test is marketed by Denver Developmental Materials, of Denver, Colorado (USA), hence the name. Tests, manuals, and other materials are available for free online at www.denverII.com. The current version is *Denver II* [23], a revision and update of DDST. The tool is designed for use by a physician, teacher, or speech-language therapist to monitor the development of infants and preschool children (from birth to age 6). The tests identify children whose development differs

significantly from others and provide a basis for further diagnosis to determine if there is a problem requiring therapy. The tests assess four basic functions: personal and social development (e.g., reciprocating a smile), fine motor skills (e.g., grasping and drawing), speech skills (e.g., sentence building), and gross motor skills (e.g., walking).

Another test in this category is the *Battelle Developmental Inventory Screening Test-2* (BDI-2) [24] designed to screen children from birth to 7 years 11 months. The test assesses the following areas: behavioral skills (self-care, showing responsibility), social skills (interactions with adults, peers, social role recognition), communication skills (speech comprehension and expression), motor skills (large and small), and cognitive skills (perception, memory, learning). BDI-2 is widely used and forms the basis of many studies described in the literature.

Method 4. A speech-language therapist evaluates speech in isolation

This category includes a number of diagnostic tools. It is also the most interesting from the point of view of constructing new tests for the screening of speech and language. In this paper we focus on the most popular, ranging from those designed for preschoolers to those that assess adolescents.

The first of the tests for preschool children is the *Bankson Language Screening Test* [25]. The test consists of 17 items, assessing speech in five categories: knowledge–semantics, morphological rules, syntactic rules, visual perception, and auditory perception. Another tool designed for preschoolers is the *Hackney Early Language Screening Test* [20]. The latter is a 20-item test divided into 7 sections: 1) comprehension of simple commands, where the child needs to follow instructions (e.g., "give the teddy bear a drink"); 2) speech expression, where the examiner manipulates a toy and the child answers questions about it; 3) comprehension of more complex commands, such as following instructions to place objects (e.g., "put the spoon in the box"); 4) comprehension, where the child needs to choose one picture from three options; 5) expression, where the child answers questions about the pictures presented; 6) expression, which is the ability to name a picture; and 7) comprehension, where the child chooses a picture from four options.

New Zealand's *Junior Oral Language Screening Tool* (JOST) [26] is used to assess preschool and early school-age children (ages 4–7, but mostly in the range 4.6 to 5.6). The test is intended to be administered by a teacher who knows the child well. It is intended as a possible indicator for deciding whether to place the child in an appropriate educational program or to refer them to a speech therapist. The test is divided into three sections: vocabulary, pragmatics (use of language for basic social communication), and grammar. The test does not assess pronunciation. Individual tasks consist of naming body parts and functions, use of verbs, adjectives, and prepositions, recognising and naming emotional states, forming plurals, forming sentences (use of tense and negation), creating a narrative statement, answering basic questions (what school do you go to?; where do you live?; how old are you?), and conversational skills. The test is non-standardised, and the final assessment consists of placing the child into one of three categories: 1) most answers correct, 2) a few correct answers in each section, and 3) very few correct answers. Placement in the third category is an indication for consultating a speech therapist.

A more extensive tool than JOST, but one where again the teacher administers the test, is the *Melbourne Speech Pathology Screening Package* [27], which is designed for preschool and school-aged children. The test covers the following aspects: 1) pronunciation, 2) speech reception and comprehension (following instructions, answering questions, acquiring new information, 3) pragmatic abilities (carrying on a conversation, understanding non-verbal cues), 4) speech expression (grammar, narrative and re-narrative skills), 5) fluency, and 6) voice. Each category is described and includes indicative developmental standards. The final evaluation also determines whether the child should be referred to a speech-language therapist.

For screening assessments of individuals 5 to 21 years of age, the most advanced tool is the 1995 *Clinical Evaluation of Language Fundamentals 4th edition* (CELF-4) [28]. Compared to earlier editions, there have been a number of changes. CELF-4 is used to test individuals suspected of having speech delays or disorders. It assesses four aspects of language: morphology and syntax, semantics, pragmatics, and phonological awareness. The test involves four steps: 1) determining whether a speech disorder is present, 2) describing the disorder, 3) assessing the clinical symptoms of the disorder (with reference to norms), and 4) evaluating speech and communication in a natural context (such as in a classroom). The questions are age-appropriate and therefore vary.

Compared to previous versions, CELF-4 has been expanded. In addition to the previous 10 subtests (following instructions, repeating sentences, constructing sentences, linking related words, understanding sentences, answering questions, composing sentences from scattered words, understanding of semantic relationships, naming colors and shapes, and linking words into categories), 5 more subtests have been added: 1) assessment of active vocabulary, a subtest for children aged 6–9 which involves naming pictures (nouns and verbs) and using names in spontaneous utterances; 2) ability to define words (for ages 10–21); 3) phonological awareness, which assesses how will the subject understands the sound structure of language, recognises phonemes, and can manipulate phonological units (such as in rhyming and segmenting sentences into syllables and phonemes); 4) pragmatic skills, which assesses routine conversational ability (verbal and non-verbal), asking and giving information, and using language in the classroom; and, finally, 5) an assessment of working memory, such as the ability to say the days of the week backwards) and accurately repeat several items on a list. In addition, the developers added an *Observational Rating Scale* to the subtests in CELF-4 to specifically evaluate a child's communication skills. It consists of 40 statements of possible difficulties a tested child may have with listening, speaking, and writing. The rating, on a 4-point scale, is made by the student themself or by a parent or teacher.

Turning to tools that are specifically designed to test Polish children, a number of questionnaires have been published, but they are still mostly at the experimental stage. They are designed to be carried out by a speech therapist or other specialist (teacher, pedagogue, psychologist, pediatrician). Examples include *Screening for the detection of speech disorders in two-, four- and six-year-old children* [29] and *Test for the examination of preschool children* [30], where the assessor gauges the competency of speech development. The former tool assesses 1) understanding of verbal commands, 2) ability to speak, 3) correct utterance of speech sounds, and 4) structural basis and efficiency of articulation. The test is done during play with the child and from an interview with parents or guardians. The latter test evaluates 1) understanding of speech, 2) pronunciation, 3) efficiency and structure of the articulatory organs. The problem with both these tests is the narrow scope of the assessment and the lack of age-related norms, or at least some indication of how to interpret the result.

Among Polish standardised tools for speech screening, there are two main tests: *The Speech and Language Screening Test for School-Aged Children* [2] and *The Speech and Language Screening Test* [30]. The first is the only test that, beside implementation and perceptual skills, can assess linguistic, communicative, and cultural competency. It is designed to test children between the ages of 6 and 14. It has been standardised on a sample of 1,800 children. It contains four subtests assessing: 1) pronunciation, in which a 20-picture questionnaire tests the subject's ability to repeat four phonetically difficult sentences; 2) narrative efficiency or story-telling ability, involving the character of a dwarf and narrating a five-element picture story; 3) motor skills of the speech organs, particularly movements of the tongue; and 4) perception of speech sounds, including the hearing of phonemes and distinguishing sounds within a word. The second test, again in Polish, aims to evaluate the speech of children between the ages of 4 and 8 and has been standardised on a group of 1,000 children. It contains four subtests: 1) sentence comprehension, assessed by understanding sentences based on dog and cat figurines; 2) vocabulary, including the names of colors, animals, and plants; 3) grammar, assessed by the ability to construct sentences from given words; and 4) pronunciation and speech fluency, gauged by repeating syllables and naming pictures.

The most extensive, standardised, and normalised Polish tool for assessing speech development is the *Test of Language Development* by Smoczynska [31], but it is more a way of accurately measuring language competency in children and is not suitable for screening purposes.

Speech testing with the *Lublin Screening Tool*

The *Lublin Screening Tool* which we have developed makes the assumptions that:

1. The examination will be conducted by a specialist.
2. The examination needs to cover basic linguistic competence, communicative competence, and implementation skills.
3. The examination is short, since it will be performed as part of a screening package covering hearing, auditory processing, speech, and voice. The initial plan was to screen 32,000 children entering school, and so far more than 28,580 have been tested. The test has no absolute time limit, but the average time to conduct a test is 5–7 minutes.
4. The goal was to identify children with suspected speech and language disorders and refer them for further specialised diagnosis. This means the assessment need not be elaborate or standardised, giving just a zero or one rating.

Development of the test stipulated that a computer would be used on which images and a picture stories are displayed. After taking the test, an evaluation is performed. Scoring for each trial is 0 or 1, with each trial having separate categories, as described in the test instructions. A finding of no disturbance (a correct answer) equates to a score of 0, while finding some disturbance, giving no answer, or an incorrect response means a score of 1.

The *Lublin Screening Tool* evaluates the following categories:

1. Pronunciation.
2. Lexical and semantic competence.
3. Narrative competence.
4. Vocabulary.
5. Speech fluency.
6. Grammatical correctness (syntax and inflection).

Receiving a score of "1" in any category is an indication that further testing and diagnosis is needed.

The test should be conducted individually, in a separate room. We do not give any feedback about the performance of individual trials. At the end, if the child asks about the results of the study, the prescribed answer is "it was pretty good". In some trials help is allowed, for example, when giving the name of a picture (pronunciation) or asking supporting questions (what does a dwarf look like? or what happened next? can you say more about it?).

Study instructions

1. Pronunciation test

The pronunciation test is based on a pictorial questionnaire containing 20 items. In addition, we assess speech fluency at this point. Fluency is also assessed during the lexical-grammatical competency test and the narration test (see item 5).

We assess the pronunciation of the name of each picture separately to its identification. If the child doesn't know the name of an item depicted in a picture, it is permissible to give them the name and ask them to repeat it. If the child fails to recognise the items in more than four pictures, the score in the vocabulary category should be given a zero.

For correct pronunciation, we consider the orthophonic standard where the school is located (regional variations are allowed).

If there is defective pronunciation, we consider whether the child has used substitutions, deformations, elisions, or a change in the ordering of phonemes. A final score of

Table 1. Pronunciation testing in the *Lublin Screening Tool* (own study)

No	Picture name	Pronunciation correct	Pronunciation incorrect	Nonfluent syllables
1.	szafa (cabinet)	0	1	– –
2.	żyrafa (giraffe)	0	1	– – –
3.	czapka (hat)	0	1	– –
4.	czekolada (chocolate)	0	1	– – – –
5.	dżem (jam)	0	1	–
6.	samolot (plane)	0	1	– – –
7.	zamek (castle)	0	1	– –
8.	język (tongue)	0	1	– –
9.	autobus (bus)	0	1	– – –
10.	cytryna (lemon)	0	1	– – –
11.	cukierek (candy)	0	1	– – –
12.	cebula (onion)	0	1	– – –
13.	widelec (fork)	0	1	– – –
14.	pędzel (brush)	0	1	– –
15.	ciastko (cookie)	0	1	– –
16.	misie (teddy bears)	0	1	– –
17.	rower (bike)	0	1	– –
18.	korale (beads)	0	1	– – –
19.	guziki (buttons)	0	1	– – –
20.	lody (ice cream)	0	1	– –
Total				/50

"1" in the pronunciation assessment is given only if the defect is repetitive (we do not fail the child based on just a single mistake).

Up to two non-fluent syllables are allowed. If the child utters more than two syllables nonfluently, then a "1" should be entered in the fluency assessment (item 5). As cases of non-fluency, we focus only on repetitions of a syllable or phoneme, or dragging out of sounds. We do not count pauses or interjections.

Command: Let me show you different pictures. Name what you see in the picture.

The names of the pictures used in the pronunciation and speech fluency screening are shown in **Table 1**. The pictures come from the Sensory Testing Platform developed by IPPH.

2. Lexical-semantic competency test

The test is a modified task from the *Logopedic Screening Test for School-Age Children* [2].

Command: There is someone who has never heard that dwarfs exist. Tell them everything you know about dwarfs.

We evaluate one by one:
1. Did the child include the dwarf character in some overarching category, e.g., human, fairy tale character, movie character, creature?
2. Did they point out the physical characteristics of the dwarf, e.g. height, appearance, clothing?

In addition, as correct answers we include:
1. Indicating the location of the dwarfs (e.g. forest, under a mushroom, cave, etc.).
2. Their mental characteristics (e.g. cheerful, hardworking, clever).
3. Modes of action (e.g., they work in a mine, mischief, play).

Passing this test requires correct answers in a minimum of two categories.

3. Narrative competency test

The test consists of telling a picture story (**Figure 1**) consisting of four narrative images, representing a narrative scheme: introduction (orientation), complication, climax, resolution/completion. We evaluate the recognition and reference of each narrative image and the construction of a coherent, logical sequence of events.

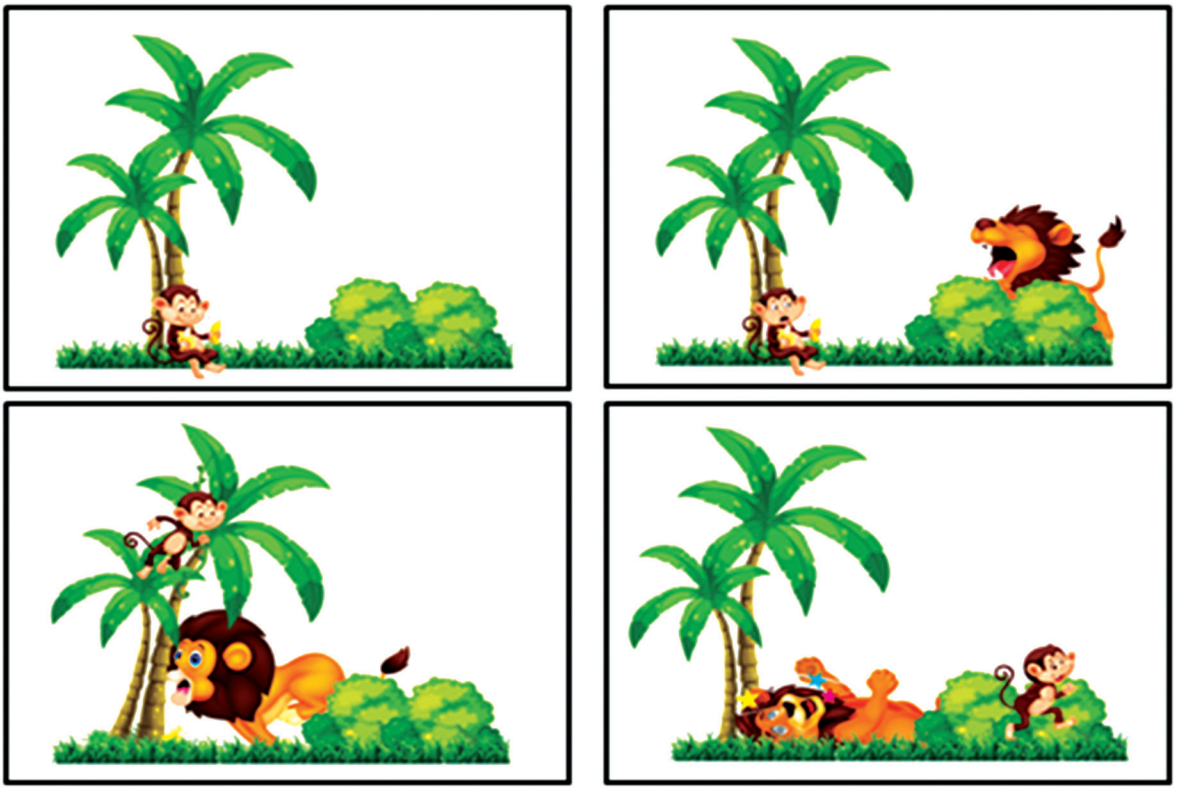

Figure 1. Picture story used in the *Lublin Screening Study* (source: own development)

Command: This is a picture story. Look at it carefully and tell someone who can't see the pictures what happened (we don't look at the monitor screen with the child).

Image 1: Shows a monkey sitting under a palm tree. The monkey is eating a banana. Next to it is a bush.

Image 2: A lion appears from the bush preparing to attack. The monkey notices it.

Image 3: The lion jumps out, but hits his head on the tree, because the monkey jumped up the palm tree just in time. The banana lies on the ground.

Image 4: The monkey has jumped down from the palm tree and is now running away. The unconscious lion is lying under the palm tree.

A positive score requires two conditions: description of all pictures and arranging them into a coherent, logical sequence of events.

4. Vocabulary (1 or 0)

Based on responses to commands 2 and 3, we also evaluate vocabulary, giving it a 1 or 0 rating. Does the child have enough vocabulary to follow instructions? We also take into account the result from the pronunciation test: if the child fails to recognise more than 4 pictures out of 20 the overall grade is "1".

5. Fluency of speech (1 or 0)

The evaluation here is analogous to that in the pronunciation test – that is, no more than two non-fluent syllables, regardless of the number of syllables spoken. Non-fluent vocalisations are regarded as repetitions of a syllable or phoneme, or dragging out of a sound. We do not count pauses and interjections. If there is a score of "1" on any of the tests then the final grade is "1".

6. Grammatical correctness of speech (1 or 0)

We consider as correct the construction of single sentences, with correct Polish syntax, including inflections. Minimally, the sentences should have correct subject, predicate, object, and adverbs, respecting syntactic relationships.

Final scoring

Obtaining a score of 1 in any of the assessed 6 categories: pronunciation, lexical-semantic competence, narration, vocabulary, fluency, and correct grammar is an indication that a more thorough diagnosis by a speech–language therapist is needed.

Conclusions

The use of the presented method has proven itself in practice, and has promoted effective hearing and voice diagnosis. Between December 2019 and May 2022, some 28,580 children have been screened despite the difficult

conditions associated with the COVID-19 epidemic. Extending screening to holistic language and communication development allows for more accurate diagnosis and presents the opportunity to prevent possible educational difficulties in the future. Speech and language screening increases the chances of early detection of developmental delays and speech disorders. However, it needs to be kept in mind that a positive test result does not always mean there is a finding of a speech disorder.

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Review papers

ISSN: 2083-389X
eISSN: 2084-3127

J Hear Sci, 2024; 14(1): 21–31
DOI: 10.17430/jhs/187260

THE BENEFITS OF EARLY COCHLEAR IMPLANTATION FOR SPEECH DEVELOPMENT IN CHILDREN WITH USHER SYNDROME: LITERATURE REVIEW

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A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

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Abstract

Introduction: Usher syndrome (USH) is a rare autosomal recessive genetic disorder characterized by sensorineural hearing loss (SNHL), vision loss (retinitis pigmentosa), and occasional balance impairment. Depending on the severity and onset of hearing loss and coexisting vestibular dysfunction, USH is divided into three clinical types – USH1, USH2, USH3 – as well as atypical USH which combines features of all these three. The purpose of this review is to present the impact of cochlear implantation on speech development in children diagnosed with all types of Usher syndrome.

Material and methods: All relevant publications published in from 2013 to 2023 were retrieved from PubMed based on the keywords *Usher syndrome*, *Usher syndrome diagnostics*, *Usher syndrome hearing loss*, *Usher syndrome cochlear*, and *Usher syndrome speech*. Exactly 67 papers were selected.

Results: Bilateral cochlear implantation in children with Usher syndrome is beneficial for audiological and verbal development provided that hearing loss is detected early and implantation done promptly. In USH1 the preferred age is before 3 in the case of severe to profound congenital SNHL; in USH2 and USH3 the optimal time for implantation is hampered by the difficulty of estimating when hearing loss occurred and its rate of progression. After bilateral cochlear implantation, studies showed improvements in the categories of auditory performance (CAP), speech intelligibility rate (SIR), meaningful auditory integration scale (MAIS), meaningful use of speech scale (MUSS), and speech reception score (SRS), together with good development of speech perception and verbal communication.

Conclusions: Early diagnosis (before the onset of vision loss) and early bilateral cochlear implantation in children who have suffered severe to profound SNHL due to Usher syndrome reduces disability and maximizes auditory–oral communication skills, significantly increasing their quality of life.

Keywords: Usher syndrome • diagnostics • hearing loss • cochlear • speech

KORZYŚCI Z WCZESNEJ IMPLANTACJI ŚLIMAKOWEJ W ROZWOJU MOWY U DZIECI ZE WSZYSTKIMI TYPAMI ZESPOŁU USHERA. PRZEGLĄD LITERATURY

Streszczenie

Wprowadzenie: Zespół Ushera (USH) jest rzadką chorobą genetyczną dziedziczoną w sposób autosomalny recesywny, charakteryzującą się odbiorczym ubytkiem słuchu (SNHL), utratą wzroku (zwyrodnienie barwnikowe siatkówki), czasami z zaburzeniami równowagi. W zależności od stopnia zaawansowania ubytku słuchu i jego momentu wystąpienia oraz współistniejącej dysfunkcji przedsionkowej zespół Ushera dzieli się na trzy typy kliniczne – USH1, USH2, USH3 oraz atypowy zespół Ushera, który łączy w sobie cechy wymienionych typów. Celem pracy jest przedstawienie wpływu wszczepienia implantu ślimakowego na rozwój mowy u dzieci ze zdiagnozowanymi wszystkimi postaciami zespołu Ushera.

Materiał i metody: Wszystkie istotne publikacje pobrano z bazy PubMed, z wykorzystaniem słów kluczowych takich jak *Usher syndrome*, *Usher syndrome diagnostics*, *Usher syndrome hearing loss*, *Usher syndrome cochlear*, *Usher syndrome speech*. Ostatecznie wybrano 67 prac. Wszystkie badania zostały opublikowane w ciągu ostatnich dziesięciu lat (2013–2023).

Wyniki: Badania wykazały, że obustronna implantacja ślimakowa u dzieci z zespołem Ushera korzystnie wpływa na rozwój słuchowy i werbalny. Jedynym warunkiem jest odpowiednio wczesne wykrycie niedosłuchu i wykonanie implantacji – najlepiej przed 3 rokiem życia w przypadku głębokiego lub ciężkiego wrodzonego SNHL w USH1. Optymalny czas implantacji w USH2 i USH3 jest utrudniony ze względu na niemożność

oszacowania momentu wystąpienia i tempa postępu ubytku słuchu. Badania wykazały poprawę w kategoriach sprawności słuchowej (CAP), wskaźnika zrozumiałości mowy (SIR), skali znaczącej integracji słuchowej (MAIS) i skali znaczącego wykorzystania mowy (MUSS) oraz wyniku odbioru mowy (SRS) po obustronnym wszczepieniu implantu ślimakowego oraz rozwojem prawidłowego postrzegania mowy i komunikacji werbalnej u dzieci z zespołem Ushera.

Wnioski: Wczesna diagnostyka i obustronna implantacja ślimakowa u dzieci z głębokim i ciężkim SNHL pozwala na redukcję niepełnosprawności i maksymalizację rozwoju umiejętności komunikacji słuchowo-ustnej u dzieci z zespołem Ushera (przed wystąpieniem utraty wzroku), co znacząco zwiększa komfort ich życia.

Słowa kluczowe: zespół Ushera • diagnostyka • niedosłuch • ślimak • mowa

Introduction

Usher syndrome (USH) is an autosomal recessive genetic disorder that causes hearing and vision dysfunction, sometimes with balance difficulties [1]. It occurs with an incidence of 1/10,000 and is the most common cause of combined deafness and blindness [2].

Depending on the severity and onset of hearing loss and coexisting vestibular dysfunction, Usher syndrome can be divided into three clinical types [3]. The most severe, Usher syndrome type I (USH1), involves inborn deafness (profound congenital bilateral and prelingual sensorineural hearing loss, SNHL) and serious balance problems. In many cases, vision loss due to retinitis pigmentosa (RP) appears within 10 years of birth and progressively constricts the field of vision (so-called tunnel vision) and reduces visual acuity, ultimately leading to complete blindness. Congenital hearing loss and vestibular dysfunction in USH1 cause severe developmental difficulties in children: there is delay in psychomotor development and, in the absence of intervention, halting verbal communication [3–6]. Type 2 (USH2) is characterised by congenital and bilateral SNHL of mild to moderate severity (affecting the low frequencies) or, more often, severe to profound SNHL affecting the higher frequencies, usually without vestibular dysfunction or with variable vestibular response. Visual problems usually start in adolescence and are progressive. Due to the clinical similarity of USH1 and USH2 – early hearing and vision loss – it is difficult to distinguish the types, but in USH2 vestibular function is often preserved [3,6–8]. Ramos et al. [9] also report possible olfactory dysfunction in patients with USH1 and USH2: they saw significantly lower olfactory threshold and shallower olfactory sulcus depth. In type 3 (USH3), the child is born with normal hearing, but in their teenage years, progressive SNHL with variable vestibular abnormalities begin. Vision loss often starts with night blindness [3]. Velde et al. [10] also distinguish Usher syndrome type 4 (USH4), where there is late-onset RP and SNHL, but no vestibular dysfunction. Finally, atypical Usher syndrome involves early and progressive SNHL without vestibular involvement and mild RP [11,12]. **Table 1** summarises the four types.

In Usher syndrome type 1 there are, depending on the mutation, six genetic subtypes: subtype 1B (mutation in myosin VIIa, *MYO7A*), subtype 1C (mutation in harmonin, *USH1C*), subtype 1D (mutation in cadherin 23, *CDH23*), subtype 1F (mutation in protocadherin 15, *PCDH15*), subtype 1G (mutation in scaffold protein containing ankyrin repeats and sam domain, SANS), and subtype 1J (mutation in calcium and integrin-binding family member 2, CIB2) [12–17]. The predominant subtype is 1B, which accounts for more than 50% of USH1 cases [18].

As for Usher type 2, Nisenbaum et al. [19,20] claim that its basis is a mutation in *CDH23* (similar to the mutation identified in USH1), as well as *USH2A* (usherin), *GPR98* (very large G protein–coupled receptor 1, also known as *VLGR1*), *WHRN* (whirlin, also known as *DFNB31*), and *ABHD12* (alpha/beta-hydrolase domain containing 12). Davies et al. [8] list four genetic subtypes of Usher syndrome type 2: 2A (mutation in usherin), 2B (mutation in *ADGRV1*), 2C (mutation in *VLGR1*), and 2D (mutation in whirlin). Stemerdink et al. [21] estimate that mutations in *USH2A* comprise 50% of the total number of cases of Usher syndrome type 2.

For USH3, the evidence is that mutations in *CLRN1* (clarin-1), *HARS* (histidyl-tRNA synthetase), and ABHD12 are typical.

In atypical Usher syndrome, a number of mutations characteristic of the three previously mentioned types have been identified (*MYO7A*, *USH1G*, *USH2A*, *GPR98*, *HARS*, *ABHD12*) as well as other mutations – *CEP250* (C-Nap1),

Table 1. Severity of hearing loss and presence of vestibular dysfunction in types of Usher syndrome [3–9,11,12]

Type of Usher syndrome	Severity of hearing loss and vestibular dysfunction
USH1	Profound congenital sensorineural hearing loss or complete deafness; severe vestibular dysfunction
USH2	Congenital sensorineural hearing loss at low frequencies (from mild to moderate); at higher frequencies, from severe to profound; no vestibular dysfunction or variable vestibular responses
USH3	Progressive sensorineural hearing loss since adolescence (from normal to severe); variable vestibular responses
Atypical USH	Progressive sensorineural hearing loss since adolescence; no vestibular dysfunction

Table 2. Detected mutations in Usher syndrome [12–20]

Type of Usher syndrome	Detected mutations
USH1	*MYO7A, USH1C, CDH23, PCDH15, SANS, CIB2*
USH2	*CDH23, USH2A, GPR98, WHRN, ABHD12*
USH3	*CLRN1, HARS, ABHD12*
Atypical USH	*CEP250, CEP78, ARSG, MYO7A, USH1G, USH2A, GPR98, HARS, ABHD12*

CEP78 (centrosomal protein 78), and *ARSG* (arylsulfatase g) [8,19]. The PDZ domain-containing protein 7 (*PDZD7*) is considered a modifier for the retinal phenotype and the severity of Usher syndrome [8,19,22,23]. **Table 2** summarises detected mutations in all types of Usher syndrome.

It is estimated that hearing loss affects approximately 1.1–3.5 per 1000 newborns screened [24]. Auditory privation significantly impacts on a child's psychosocial development, and can include delays in speech and language development, depression, anxiety, low self-esteem, problems with self-acceptance, and reduced academic performance [24–26]. Depending on when hearing loss occurred, the loss can be classed as prelingual, which occurs before the development of speech (Usher syndromes types 1 and 2), or postlingual, which occurs after speech has been acquired (Usher syndrome type 3) [25]. To avoid the long-term consequences of hearing loss, early diagnosis (hearing screening) and treatment are crucial [24,25].

The degree of hearing impairment in Usher syndrome increases with age, but it is impossible to predict the rate of progression: in some people it progresses quickly to complete deafness, while in others the rate of progression is almost imperceptible. However, it has been noticed that the most rapid progression of hearing loss occurs within the first two decades of life [8].

Currently, the only treatments for hearing loss associated with Usher syndrome are hearing aids or cochlear implantation [27]. Hearing aids are often preferred in such patients, but in the case of USH1 (and some people with USH2 and USH3) they may prove ineffective (since good speech recognition is required), and then a cochlear implant (CI) is needed. CIs are frequently used in the treatment of profound hearing loss in children with Usher syndrome and allow children to achieve proper speech and language development [8,12,28,29]. According to Hoshino et al. [30], auditory stimulation in a child with congenital deafness restored before the age of 3 1/2 allows the child to acquire natural developmental abilities, and so early implantation in a case of Usher syndrome can enable proper speech and verbal communication skills.

According to Davies et al. [8], children with Usher syndrome type 1 are perfect candidates for a CI because they are usually born with prelingual deafness in which low-frequency hearing is preserved. Hence, early and bilateral implantation has the potential to confer significant audiological benefits – hearing and speech intelligibility can often be excellent. In USH2, however, hearing aids used from early childhood are usually the first choice, although if there are poor speech detection and communication problems in patients with severe and progressive hearing loss, then a CI is indicated. For patients with USH3, a CI may also be a suitable way to improve hearing, but only if the hearing loss is severe (otherwise there is a risk of damaging residual hearing) [8]. According to Koenekoop et al. [4], non-implanted children with USH1 often fail to develop speech.

In this review we present the benefits of receiving an early CI on the speech development of children diagnosed with Usher syndrome.

Material and methods

Aim

The purpose of this review is to summarise the impact of early cochlear implantation on speech development in children diagnosed with Usher syndrome.

Eligibility criteria

We analysed studies published within the last 10 years. The core focus was Usher syndrome, its impact on speech development, and the role of CIs in such children. We considered all types of observational studies.

Search strategy

The search was conducted in PubMed. Keywords were *Usher syndrome* (686 results), *Usher syndrome diagnostics* (338 results), *Usher syndrome hearing loss* (343 results), *Usher syndrome cochlear* (88 results), and *Usher syndrome speech* (17 results). The last time the source texts were reviewed was on 15/09/2023. The inclusion criteria used in the review were publication date (last 10 years), papers with full text available, English language, on-topic, approval of a bioethics committee, and high reliability. We excluded older studies, animal studies, pharmacological models, studies in languages other than English, studies with low reliability, and those without bioethics committee consent.

Data collection

First, papers were selected and then abstracts and full articles for chosen studies were read. The extracted data included the following information: clinical features, hearing disorders, speech development, and type of CI. There were 1472 papers from the PubMed database which were retrieved. Papers were searched using the above keywords and duplicates were removed. There were 243 articles whose titles were relevant to the topic. After reading their abstracts, 155 of them appeared to be highly reliable. We checked the papers in terms of quality of the results,

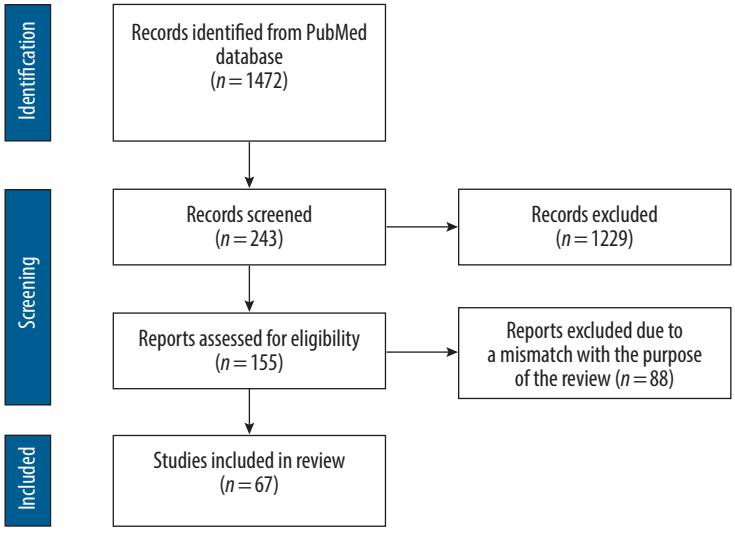

Figure 1. The search process PRISMA diagram

the type of technique used, the intervention and, finally, if they met our inclusion criteria, the full article was read. Finally, 67 articles were selected as being relevant to the topic. **Figure 1** illustrates the search process.

Results

Time and lateralisation of CIs in children with Usher syndrome

The literature recommends bilateral implantation in children with USH1 at the earliest opportunity. This will have the greatest developmental benefits for the child in terms of hearing, speech, and development. It has been found that in children with USH implanted before the age of 3, verbal communication is better developed than in children who received a CI at a later age [8,31]. In the case of USH2 and USH3, the optimum time for implantation is hampered by the inability to estimate when hearing loss began and its rate of progression [8]. Several authors document how early implantation in children allows them to develop speech, and the earlier the child receives a CI, the better will these skills be [8,12,28,32].

Bilateral implantation produces significantly better effects than unilateral implantation and better sound localisation (due to brain plasticity and bilateral noise blocking), and so children implanted early show faster development of speech and psychosocial skills than children with SNHL but no CI. This is especially important in people with disabilities, such as those with Usher syndrome, who develop RP in their teenage years [8,12]. Moreover, analysis by Davies et al. [8] showed that bilateral implantation at an early age (defined as less than 3 years) allowed children to develop better verbal communication skills than patients implanted later (defined as age above 13 years) who tended to have suboptimal speech reception scores. Among children with various subtypes of USH and implanted before the age of 9, the best effects in postoperative speech perception were achieved by children operated on before the age of 3. Nevertheless, findings show that receiving a CI within the first 20 years of life still allows for measurably better hearing results [8]. Of course, it is important for auditory rehabilitation to follow on from implantation, as it effectively improves communication skills [8].

Alsanosi [33] concludes that early, simultaneous bilateral implantation in patients with Usher syndrome with congenital profound bilateral deafness allows age-appropriate audiological results. For example, implantation in a 5-month-old boy with Usher syndrome, probably USH1, shows what is possible [33]. Hoshino et al. [30] find that late implantation in patients with USH1 allows speech recognition, but only in patients who have received previous hearing stimulation, because full development of the auditory pathway and central processes is necessary. Without prior central hearing skills, there is difficulty adapting to the CI and it often leads to failure [30].

Benefits of a CI in children with Usher syndrome

The benefits of a CI in children diagnosed with Usher syndrome were investigated by Nair et al. (2020) in a group of 27 patients aged 1–6 years with bilateral profound sensorineural hearing loss [34]. The control group were 30 implanted people of similar age, but without Usher syndrome. In both groups, the categories of auditory performance (CAP) and speech intelligibility rate (SIR) were examined after 3, 6, 9, and 12 months after the procedure. Both in the study and control groups, improvement in verbal communication was noted, but in the USH group it was less developed, which may have been affected by RP as part of Usher syndrome. The paper emphasises the importance of audio-verbal therapy after a CI, the significance of an individualised approach, and the need for early intervention, which will protect the child from severe impairment and enable appropriate development [34].

The results of the Nair et al. (2020) study [34] confirm similar research carried out by Remjasz-Jurek et al. (2023) in which auditory performance and speech intelligibility after a CI in children with Usher syndrome were also

Table 3. Summary of findings from research studies and case reports included in review

Authors	Type of USH	Number of patients (n) implanted with USH	Type of cochlear implantation	Conclusion
Henricson et al. (2018) [28]	USH1	*n* = 7	unilateral	Children with Usher syndrome implanted after the age of 2 achieve similar outcomes (memory capacity, phonological and lexical skills) to children implanted with congenital deafness
Hoshino et al. (2017) [30]	USH1	*n* = 10 average age at implantation = 18.9 years (5–49)	unilateral	Late cochlear implanttation in patients with USH1 allows speech recognition, but only in patients having previous hearing stimulation
Jatana et al. (2013) [32]	non-defined	*n* = 712 average age at implantation = 3.3 years	bilateral	Bilateral CI in children with USH permits development of open speech perception in 92% and verbal communication in 69%
Broomfield et al. (2013) [36]	non-defined	*n* = 9 Average age at implantation: early = 2.7 years late = 12.7 years	unilateral	Results achieved after a CI are usually satisfactory (higher reception scores and speech perception ability), but may differ in patients with the same genetic syndrome
Alsanosi (2015) [33]	non-defined	*n* = 1 (case report) age at implantation = 5 months	bilateral	Simultaneous bilateral implantation in children aged several months is recommended when performed by an experienced team, which allows for age-appropriate audiological results
Remjasz-Jurek et al. (2023) [35]	non-defined	*n* = 35 average age at implantation = 6.3 years (0.3–17.6 years)	unilateral	USH patients had marginally worse post-implant outcomes than asymptomatic implanted patients, CI significantly improved the hearing and speech intelligibility of children with Usher syndrome. Particular advancement was noticed in children who received a CI under the age of 3 years
Nair et al. (2020) [34]	non-defined	*n* = 27 average age at implantation = 2.9 years (11 months – 4.7 years)	unilateral	Both in the study and control groups, improvement in verbal communication was noted, but in the USH group it was less developed than in the control group

assessed [35]. The research group was 35 children aged 0.3–17.6 years (average age of implantation was 6.3) diagnosed with Usher syndrome (without specifying the type), whose results were compared to a control group of 46 implanted children without symptoms. Average PTA thresholds were 25.0 dB HL in the group of children with Usher syndrome, while in the control group it was 28.4 dB HL. Categories of auditory performance (CAP) was 5.3, compared to 5.1 in the study group. Speech intelligibility rate (SIR) in the study group was 4.1 and in the control group 3.9. The *Meaningful Auditory Integration Scale* (MAIS) in the Usher syndrome group was 82.3% and in the control group 80.5%, while the *Meaningful Use of Speech Scale* (MUSS) in the study group was 81.8% and in the control group 76.6%. The results showed that although USH patients had marginally worse post-implant outcomes than asymptomatic implanted patients, a CI significantly improved the hearing and speech intelligibility of children with Usher syndrome. Particular advancement was noticed in children who received a CI under 3 years of age. In general, better speech therapy results were achieved in children who were implanted early [35].

Jatana et al. (2013) evaluated the benefits in speech perception and movement after bilateral cochlear implantation in 712 children with USH (also without determination of USH subtype) [32]. Children were implanted at ages from 6 months to 11.6 years (average 3.3 years). Observations were carried out from 10 months to 15.6 years after implantation – an average of 7.8 years. The study showed that the vast majority of children (92%) developed open speech perception and more than half the children (69%) used verbal communication. The authors conclude that bilateral CIs in children with severe to profound SNHL with Usher syndrome is crucial for proper speech development [32]. Another study by Broomfield et al. (2013) demonstrated increased Bench–Kowal–Bamford (BKB) speech reception scores and speech perception ability using the

Table 4. Summary of findings from other papers included in review [1–27,29,31,37–67]

Authors	Conclusions in terms of diagnosis and treatment by a CI
Koenekoop et al. (2023) [4,7] Fuster-García et al. (2021) [5] Ramos et al. (2019) [9] Velde et al. (2022) [10] Gilmore et al. (2023) [13] Donaldson et al. (2018) [14] Miyasaka et al. (2013) [15] Chen et al. (2022) [16] Emptoz et al. (2017) [17] Toms et al. (2020) [18] Nisenbaum et al. (2022)[19] Stemerdink et al. (2022) [20] Blanco-Kelly et al. (2015) [21] Zou et al. (2014) [22] Bonnet et al. (2016) [23] de Joya et al. (2021) [27] Whatley et al. (2020) [66] Toualbi et al. (2020) [67]	Bilateral CI in patients with USH is, next to hearing aids, currently the best option for hearing rehabilitation, despite the detection of many mutations in USH and the dynamic development of gene therapies
Castiglione et al. (2022) [1] McKinney et al. (2017) [43] Miyamoto et al. (2018) [44] Karltorp et al. (2020) [45] Dettman et al. (2021) [46] Szyfter et al. (2019) [47]	In children with USH1, early CIs (age 6–12 months) is recommended to ensure normal development of hearing, speech, and social skills
Delmaghani et al. (2022) [2] Koenekoop et al. (2023) [4,7]	CIs and hearing aids may provide significant benefits in auditory-sensory orientation in most patients with USH
Davies et al. (2021) [8] Health Quality Ontario. Bilateral Cochlear Implantation: A Health Technology Assessment (2018) [48] Gifford et al. (2020) [50] Kumari et al. (2018) [51] Bae et al. (2019) [52]	When children affected by USH (severe to profound SNHL) are implanted bilaterally, they have better sound localisation, speech perception, language development, and greater vocalisation in preverbal communication compared to unilateral implantation. Additionally, children with bilateral CIs achieve better results at school and communicate more effectively with others
Tsang et al. (2023) [6] Cejas et al. (2015) [31] Virzob et al. (2023) [49]	Most patients with USH have better speech perception after CI, but the development of verbal communication depends on their age (the sooner the implantation, the better the speech perception). This is especially important in USH1 patients with prelingual deafness
Nolen et al. (2020) [3] Fowler et al. (2021) [11] Mathur et al. (2015) [12]	Most children with USH who received a CI early are able to develop verbal communication. SNHL occurring in atypical USH usually requires hearing aids only
Sommerfeldt et al. (2023) [24] Young et al. (2023) [25] Verstappen et al. (2023) [26] Korver et al. (2017) [29] Position Statement from the Joint Committee on Infant Hearing (2019) [37] Park et al. (2021) [40] Fitzpatrick et al. (2015) [41] Koffler et al. (2015) [53] Arias-Peso et al. (2023) [54] Stiff et al. (2020) [55]	Lack of early intervention (diagnosis and treatment, including CI) for hearing loss in people with USH can lead to serious developmental delays in children, including speech and language development. Thus early detection of hearing loss and then treatment remains crucial
Magliulo et al. (2015) [62] West et al. (2015) [63] Kletke et al. (2017) [64]	Newborn hearing screening (OAE, ABR) is a key to early detection of hearing loss. In the diagnosis of hearing loss, ECOG and assessment of speech may be helpful, while when assessing vestibular function key tests are VEMPs, vHIT, as well as Fitzgerald–Hallpike caloric test, rotary chair testing, ENG, posturography
Yoshimura et al. (2021) [56] Medina et al. (2021) [57] Ramzan et al. (2018) [58] Lenarduzzi et al. (2015) [59] Aparisi et al. (2014) [60] Magliulo et al. (2017) [61] Ambrosio et al. (2021) [65]	Comprehensive genetic tests are costly, but necessary for a definite diagnosis of USH (and differential diagnosis), and can help to detect the USH even before the appearance of ophthalmological symptoms

Table 4 continued. Summary of findings from other papers included in review [1–27,29,31,37–67)

Authors	Conclusions in terms of diagnosis and treatment by a CI
Sharma et al. (2020) [38] Warner-Czyz et al. (2022) [39]	To maximise the benefits of CIs in deaf children (USH1), support is essential. The greatest speech benefits from CIs are achieved by children with USH who have no other comorbidities and where intervention was begun quickly
Varadarajan et al. (2021) [42]	The benefits of CIs in children with USH depend on the degree of hearing loss, asymmetric or bilateral hearing loss, presence of residual hearing, inner ear malformation, and cochlear nerve deficiency

Speech Reception Score (SRS) in a group of 38 implanted children – 9 with Usher syndrome and 29 with other genetic syndromes with severe hearing impairment [36].

Table 3 and **Table 4** summarise the findings from the papers included in this review.

Discussion

Quick intervention helps proper speech development

The result of a CI depends on the age at which the hearing loss began and when it was diagnosed, whether the implant was done prelingually or postlingually, the age of implantation, the method of communication before and after intervention, rehabilitation, as well as motivation to learn and support from the family. The above studies agree that the earlier the implantation, the greater the chances for proper development of speech skills. However, for children with Usher syndrome, no clear guidelines exist from scientific societies specifying the appropriate age for implantation. Among the studies cited, most recommend an age of under 3 years for a child with USH1 to receive a CI. In other types of Usher syndrome, the best time depends on the level of hearing impairment.

The age of implantation is affected by when the hearing loss was detected, and here hearing screening programs play a major role. The current recommendations of the Joint Committee on Infant Hearing from 2019 include the need to perform a hearing screening by the age of 1 month, to identify hearing loss by 3 months, and to enroll for appropriate therapeutic intervention by 6 months. However, the committee encourages a 1–2–3 approach: a hearing screening by month 1, identifying hearing loss by month 2, and beginning therapy by month 3 [37,38]. Early use of hearing aids is also encouraged, and if progress is not achieved there is time for early referral to determine candidacy for a CI. This is particularly important in younger children so that they can develop verbal communication [38,39]. Delays in receiving a CI lead to poorer outcomes [40]. Significant problems hindering early implantation are diagnostic delays, the presence of residual hearing, comorbidities, family hesitancy and geographical location [38,39]. Fitzpatrick et al. (2015) point out that delays in implantation in children result from a failure to continually test audiological performance, and so it is important to constantly monitor children with hearing loss [41].

Although CIs are a proven method of treating sensorineural hearing loss in children and adults, better technology allows the indications for implantation to be expanded. This ensures that all children will have access to sounds and develop language and communication skills [39,42]. As set out in [42], the current FDA indications for receiving a CI in children depend on the degree of hearing loss. However, there is good evidence of successful implantation below these indications [33,43–46]. Furthermore, another study [45] has shown that in children implanted at 5–11 months, the level of speech recognition and vocabulary range was significantly better than in children implanted at 12–29 months (and there was no indication of an increase in surgical complications due to the lower age). Based on the Categories of Linguistic Performance (CLIP) analysis, another study found that children who received a CI before 9 months had better language development than children implanted later [46].

Bilateral cochlear implantation in children is still under discussion worldwide [47–51]. According to Szyfter et al. (2018), this solution should be used in children with visual impairment (Usher syndrome), with initial cochlear obstruction and insufficient audiological results from unilateral implantation [47]. In the case of bilateral congenital deafness, implantation should not be postponed for longer than 12 months. An assessment made by Health Quality Ontario demonstrated that in children with severe to profound SNHL, they had improved sound localisation, speech perception, language development, and greater vocalisation in preverbal communication when implanted bilaterally compared to unilaterally. The Canadian group concluded that bilateral implantation is effective and willingly used by patients [48]. Virzob et al. [49] reach similar conclusions, emphasising that the age at implantation, the level of language peformance before surgery, the duration of implant use, and auditory rehabilitation are key to achieving good results. Gifford et al. (2020) believe that residual hearing at low frequencies is not an obstacle to bilateral implantation after a trial period with bimodal stimulation [50]. Kumari et al. (2018) encourage the use of bilateral implantation as standard in severe prelingual bilateral SNHL in children; in their study mean CAP and SIR scores were significantly higher in children implanted bilaterally than in children implanted unilaterally [51].

A study by Bae et al. (2019) showed that children with bilateral prelingual deafness and a CI received between 1 and 3 years of age are more likely to attend mainstream schools than similar children without an implant [52]. They note that the rate of attending a tertiary institution of people with CIs is the same as in the general population.

Diagnosis of USH and importance of early detection

The differential diagnosis of Usher syndrome is a key. It is estimated that there are about 40 disorders in which vision and hearing are impaired (e.g. Alport syndrome, Stickler syndrome, Baraitser–Winter syndrome), but more than half the cases are Usher syndrome [53–55]. Typically, audiological symptoms precede vision loss in patients with USH [8]. To make a diagnosis, thorough ophthalmological and otorhinolaryngological examinations are needed, but genetic tests are crucial to confirm the diagnosis and make a prognosis [53–55].

Yoshimura et al. (2021) point out that although Usher syndrome is diagnosed based on clinical symptoms, comprehensive genetic tests can detect the disease before the appearance of ophthalmological symptoms [56]. Medina et al. (2021) emphasise that, when Usher syndrome is uncertain, it is important to do genetic testing for genes responsible for hearing and vision loss although a combination of genetic deafness and blindness does not always mean Usher syndrome [57]. According to these authors, the genes responsible are *ALMS1*, *TUBB4B*, *CEP78*, *ABHD12*, and *PRPS1*.

Precise genetic diagnosis is hampered by the genetic heterogeneity of Usher syndrome, its high cost, and the long time required to undertake multiple testing procedures [58,59]. So far, mutations in 11 genes responsible for USH have been described, but many patients have the condition without a specific mutation being identified [58]. There is hope that many previously unexplained genetic mutations will be detected by next-generation sequencing (NGS) using targeted panel sequencing and clinical exome sequencing (CES) and genome sequencing [58–60]. Ramzan et al. (2018) highlight the role of CES in identifying the genetic cause of hearing loss [58]. According to them, this method is accurate and allows rare genetic diseases such as Usher syndrome to be detected. Aparisi et al. (2014) designed a custom HaloPlex panel for Illumina platforms to capture exons of 10 Usher syndrome causative genes – *MYO7A*, *USH1C*, *CDH23*, *PCDH15*, *USH1G*, *CIB2*, *USH2A*, *GPR98*, *DFNB31*, and *CLRN1* – and the related genes *HARS* and *PDZD7* and candidate genes *VEZT* and *MYO15A* [60]. Among 44 patients with Usher syndrome participating in the study (11 in the control group with known mutations, and 33 in the study group without a detected mutation), the panel confirmed mutations in 40 of them (8 from the control group and 32 from the study group). Sequencing using the panel allowed 53 different mutations to be detected at the same time – both point mutations and large rearrangements, including the detection of mutations in previously genetically undiagnosed patients. According to the authors, genetic diagnosis of Usher syndrome using a panel allows for more genetic causes of USH to be detected and minimises the cost of testing [56]. According to Lenarduzzi et al. (2015), it is important to investigate all possible causative genes to detect mutations and direct treatment [59].

Newborn hearing screening remains crucial, especially in the diagnosis of Usher syndrome type 1. A child with an abnormal result of otoacoustic emissions (OAEs) or auditory brainstem response (ABR) can be subsequently tested by otoscopy, cytomegalovirus (CMV) testing, temporal imaging, and possible genetic testing [4,8]. Electrocochleography (ECOG) and, in older children, assessment of speech may also be added [4]. Vestibular function can be evaluated using caloric testing, cervical vestibular evoked myogenic potentials (cVEMPs), ocular vestibular evoked myogenic potentials (oVEMPs), video head impulse test (vHIT), rotary chair testing, electronystagmography (ENG), and posturography [8,60]. According to several authors, VEMP and vHIT remain the most sensitive tests for detecting hidden vestibular damage in USH2; both tests are recommended to assess vestibular nerve deficit in patients with USH, which also helps determine the type of USH [4,61,62].

Ophthalmological diagnosis of children with profound to severe preverbal SNHL is often essential in the diagnosis of Usher syndrome [63,64]. West et al. (2015) mention the necessity of performing an electroretinogram (ERG) in those patients with SNHL and a CI or with ophthalmological symptoms (retinal dystrophy) [63]. According to Kletke et al. (2017), in children with congenital SNHL and co-occurring vestibular disorders, the risk of USH is increased, and so performing an ophthalmological examination (including an electroretinogram) and genetic tests for USH are recommended, because they will speed up diagnosis and treatment [64]. When diagnosing Usher syndrome in children with SNHL, Ambrosio et al. (2021) recommend, as well as performing an electroretinogram, determination of the dark-adapted threshold [65]. However, in all cases, genetic testing is necessary for a definite diagnosis of Usher syndrome.

Identification of genes responsible for Usher syndrome and the development of gene therapies provide opportunities for cures and for improving the quality of life of patients with USH. However, even though hearing aids and CIs improve hearing and allow good speech development, there is currently still no treatment for retinitis pigmentosa [27,66,67].

Limitations

Many of the available publications do not determine the type or subtype of Usher syndrome, often because there may be blurring of symptoms between the different types. Additionally, due to the small database of records from the last 10 years, some of the chosen studies were carried out on small groups, and so there is a need for further research to confirm the results, particularly on the impact of a CI on improving children's speech.

Conclusions

Early cochlear implantation in children with severe to profound SNHL and Usher syndrome reduces their disability and maximises their auditory and oral communication skills, significantly increasing their quality of life. The earlier the implantation, the greater the chances that the child will develop good speech and be able to effectively communicate verbally, provided of course that there is adequate auditory–verbal rehabilitation. If Usher syndrome is detected early and treated appropriately (including cochlear implantation), children with USH can be rehabilitated even before the onset of vision loss.

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Original articles

ISSN: 2083-389X
eISSN: 2084-3127

J Hear Sci, 2024; 14(1): 35–39
DOI: 10.17430/jhs/185922

RELATIONSHIP BETWEEN SELF-RATED MUSICAL COMPETENCE AND AUDITORY PROCESSING IN INSTRUMENTALISTS

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Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

Abstract

Introduction: Although the benefits of musical training have been shown to affect a number of auditory processes, the question of how self-rated musical competence correlates with auditory perception remains largely unexplored. The present study aimed to investigate how self-perceived musical competence correlates with musicians' ratings of their own abilities in speech perception, spatial hearing, and sound qualities.

Material and methods: The *Edinburgh Lifetime Musical Experience Questionnaire* (ELMEQ) was administered on 57 instrumentalists aged 19 to 53 years (mean = 25.1 ± 4.9 years, 34 males and 23 females). All had normal hearing and had undergone formal training on musical instruments for 1–2 years. All of them also regularly practised music for at least 1–2 h/week. Based on their ELMEQ scores, they were divided into two groups: low (ELMEQ score ≤ 7; $n = 12$) and high (ELMEQ score ≥ 11; $n = 16$; self-perceived musical competence. Participants were sent a modified questionnaire from the *Speech, Spatial and Qualities of Hearing Scale* (SSQ) using Google forms, and the data statistically analysed.

Results: A Mann–Whitney *U*-test showed that participants with high self-rated musical competence had significantly higher ratings on the spatial hearing and sound qualities sub-domains of SSQ compared to their counterparts who had lower ELMEQ scores. However, both groups scored about the same on the speech perception domain of SSQ. A Spearman test revealed a moderate to strong positive correlation between self-rated musical competence and SSQ ratings in each domain.

Conclusions: The findings show that instrumentalists with high self-rated musical competence exhibited high proficiency in the spatial hearing and sound quality domains of auditory processing.

Keywords: instrumentalists • musical competence • speech perception • sound qualities • spatial hearing

ZWIĄZEK POMIĘDZY WŁASNĄ OCENĄ KOMPETENCJI MUZYCZNYCH A POZIOMEM PRZETWARZANIA SŁUCHOWEGO WŚRÓD INSTRUMENTALISTÓW

Streszczenie

Wprowadzenie: Chociaż wykazano, że trening muzyczny wywiera korzystny wpływ na wiele procesów słuchowych, to pytanie, jak własna ocena poziomu kompetencji muzycznych koreluje z percepcją słuchową pozostaje w dużej mierze niezbadane. Celem obecnego badania była ocena korelacji pomiędzy własną oceną kompetencji muzycznych a poziomem kompetencji w zakresie percepcji mowy, lokalizacji dźwięku i jakości dźwięku we własnej ocenie muzyków.

Materiał i metoda: Kwestionariusz *Edinburgh Lifetime Musical Experience Questionnaire* (ELMEQ) wypełniło 57 instrumentalistów w wieku 19–53 lat (średnia = 25,1 ± 4,9 lat, 34 mężczyzn i 23 kobiety). Wszyscy mieli słuch w normie i przeszli formalne szkolenie w zakresie gry na instrumencie muzycznym przez okres 1–2 lat. Wszyscy regularnie ćwiczyli granie przez co najmniej 1–2 godz. tygodniowo. Uczestnicy zostali podzieleni na dwie grupy na podstawie wyników kwestionariusza ELMEQ dotyczących kompetencji muzycznych w ocenie własnej: niskie (wynik ELMEQ ≤ 7; $n = 12$) i wysokie (wynik ELMEQ ≥ 11; $n = 16$). Uczestnicy otrzymali zmodyfikowany kwestionariusz *Speech, Spatial and Qualities of Hearing Scale* (SSQ 5,6) za pośrednictwem formularzy Google, a otrzymane dane zostały poddane analizie statystycznej.

Wyniki: Test U Manna–Whitneya pokazał, że uczestnicy, którzy wysoko ocenili swoje kompetencje muzyczne, uzyskali istotnie wyższe wyniki kwestionariusza SSQ w obszarach lokalizacja dźwięku i jakość dźwięku w porównaniu do uczestników, którzy uzyskali niskie wyniki ELMEQ. Obie grupy uzyskały podobny wyniki w obszarze percepcji mowy kwestionariusza SSQ. Test Spearmana wykazał istnienie średniej do silnej pozytywnej korelacji między własna oceną kompetencji muzycznych a wynikami SSQ w każdym obszarze.

Wnioski: Wyniki pokazują, ze instrumentaliści, którzy wysoko oceniali swoje kompetencje muzyczne, wykazywali wysoką sprawność przetwarzania słuchowego w zakresie lokalizacji dźwięku i jakości dźwięku.

Słowa kluczowe: instrumentaliści • kompetencje muzyczne • percepcja mowy • jakość dźwięku • lokalizacja dźwięku

Introduction

Enjoying music is universal, yet musical abilities vary from person to person. This variation also depends on training and musical competence. Musical competence refers to the ability to of a listener to perceive, remember, and discriminate musical melodies and rhythms [1]. It can consist of rehearsal, formal or informal training, and performances such as playing an instrument or singing. Research has shown that musical competence and exposure results in better cognitive abilities and slows down decline in aging-related auditory processes [1–3].

Musical background is linked to several other benefits as well, such as psychoacoustical abilities [4,5]. Studies suggest that, depending on the amount of musical experience, an individual's listening history can affect their cochlear frequency selectivity [6]. Pitch discrimination is also better in musicians than in non-musicians [7,8]. Musicians show better auditory attention [9,10], temporal processing skills [11,12], and speech perception in the presence of noise [12] compared to non-musicians. The literature shows that cortical auditory evoked potentials (CAEPs) are significantly enhanced in musicians [13].

The psychoacoustic correlates of musical competence can be found in tasks such as detecting the difference between two sequences of tones, intensity differences, and temporal differences. Self-perceived measures of musical competence involve the participant rating their ability to listen to music; perceiving features like rhythm, pitch, dynamics, melody, harmony, tone color, and texture; singing; and playing an instrument.

By assessing the musical competence and exposure of a person in detail, it appears theoretically possible to establish a link between musical experience and auditory domain-specific advantages. Several scales have been developed to measure musical competence: the *Self-assessment of Musical Skills and Experience* [14], *Ollen Musical Sophistication Index Questionnaire* [15], the *Music USE Questionnaire* [16], *Music Use and Background Questionnaire* [17], and the *Edinburgh Lifetime Musical Experience Questionnaire* (ELMEQ) [18]. They can also provide insight into other areas such as musical training, receptive sensitivity to music, how much time they invest in listening to music, and how much importance they give to music.

Research has established the impact of formal musical training on many auditory processes, including speech perception, sound qualities, and spatial hearing as measured by the SSQ scale [2,3]. However, there is scant evidence on the effect of self-perceived musical competence in musicians and its association with auditory performance in daily listening. The purpose of this study was to determine the relationship between self-perceived musical competence, as assessed using ELMEQ, and auditory performance (in terms of speech perception, spatial hearing, and sound qualities). In particular, the study aimed to determine correlations between self-rated musical competence and SSQ ratings.

Material and methods

Participants

A total of 57 musicians in the age range of 19 to 53 years (mean = 25.1 ± 4.9 years, 34 males and 23 females) participated in the study. All the participants had undergone formal training for musical instruments such as strings, piano, or percussion for 1–2 years, and they currently practised music for at least 1–2 h per week. The participants were divided into two groups based on their self-perceived musical competence abilities using ratings obtained from the *Edinburgh Lifetime Musical Experience Questionnaire* (ELMEQ) rating scale [18]. ELMEQ is a 30-item musical questionnaire with four sections which focus on musical instruments, singing, reading music notation, and listening to music. It provides information about the quantity and characteristics of musical training and expertise. It also includes questions about singing experience, music notation reading, self-rated musical ability, and music listening regardless of genre (classical, folk, pop, rock, or jazz). Here, a 5-point rating scale was used to assess the competence of the participants based on questions 6, 7, and 8 of the musical instruments section; these three questions are recommended by Okely et al. [18] for making assessments of self-perceived musical ability. Depending on the level of experience, participants can achieve a maximum possible score of 15. The questionnaire was administered using Google forms. The self-perceived ratings in ELMEQ were cross-checked against the questions on the *Music Performance Self-Efficacy Scale* [19].

Based on a pilot study, instrumentalists who scored above 11 were considered to have high self-perceived musical competence, while those who scored less than or equal to 7 were considered to have a lower musical competency. Participants who scored between 8 and 11 were not included in the data analysis because that was considered a grey area. Based on these criteria, two groups were formed: Group 1 comprised of 12 participants who had low competence (mean age = 24.8 ± 4.9 SD years, 8 males and 4 females) while Group 2 consisted of 16 participants (mean age = 24.1 ± 4.9 SD years, 11 males and 5 females) with high self-rated musical competence.

Procedure

Data collection for the study was done through Google forms. One Google form was designed to obtain demographic details and questions related to musical experience (**Table 1**). These questions were followed by administration of a simplified version of the Speech, Spatial and Qualities of Hearing Scale (SSQ v.5.6) [14].

Table 1. Google form used for data collection

Name: **Age:** **Gender:**				
Question	Options			
Have you ever learned to play a musical instrument?	○ Yes		○ No	
Which musical instrument do you play?	String instrument (guitar, violin, sitar, etc.)	Keyboard/piano	Percussion (drum, dholak, table, etc.)	Other
How long have you been playing the instrument?	0–2 years	2–5 years	More than 5 years	
Approximately how many hours do you currently play per week, on average?				
Have you ever played with a band, ensemble, or orchestra?	○ Yes		○ No	
If yes, how many years of your life did you play with a musical group?				

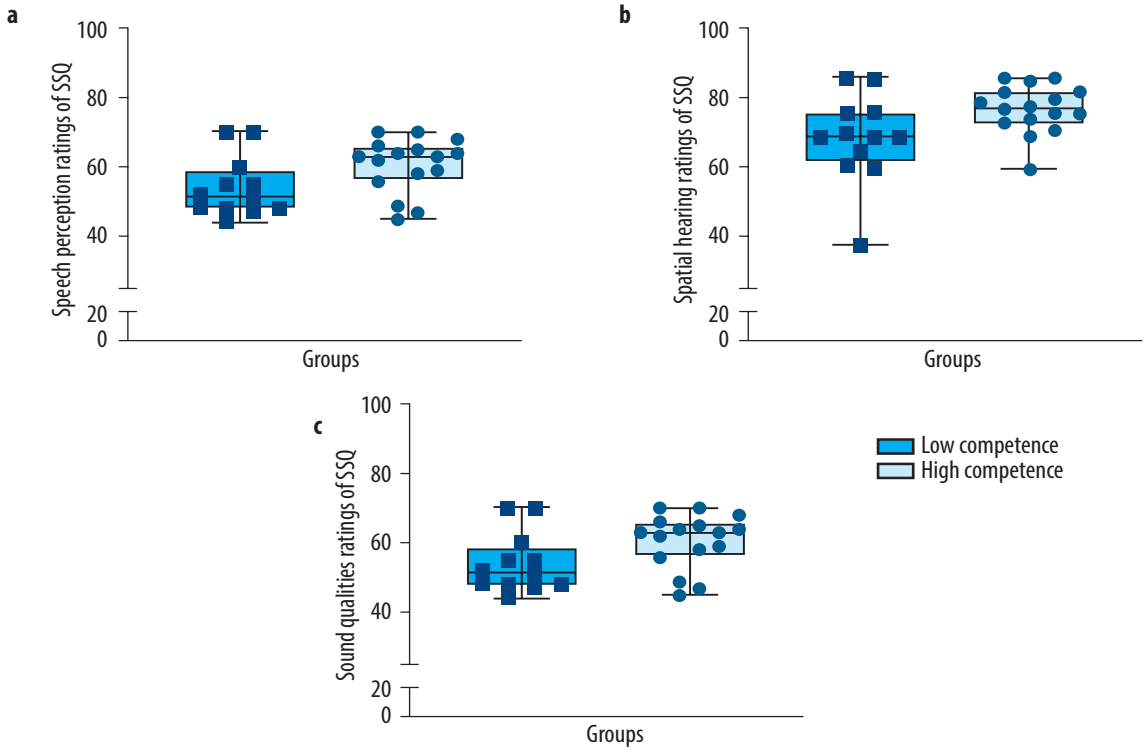

Figure 1. Box plots representing SSQ ratings for domains of speech perception **a**), spatial hearing **b**), and sound qualities **c**), divided according to musical competence. As indicated in Table 2, only **b**) and **c**) are statistically different. The plots show individual data points (symbols), medians (center lines), and inter-quartile ranges (Q1 and Q3)

SSQ was devised to self-assess hearing ability in different domains. It consists of a set of situations covering speech in noise, the spatial aspect of hearing, and qualitative judgments of hearing which are rated by the percentage of disability in the respective sections. The SSQ [14] was modified in the Google form so that the rating scale was simplified from a 10-point rating scale to a 5-point rating scale, where 1 denoted 100% difficulty, 2 denoted 75% difficulty, 3 denoted 50% difficulty, 4 denoted 25% difficulty, and 5 denoted 0% difficulty. There were 3 subsections: speech perception, having 14 questions; spatial hearing, with 17 questions; and sound qualities, 19 questions [14]. A participant's total achievable score was 70 for the speech section, 85 for spatial hearing, and 95 for sound qualities. Informed consent was obtained from the participants before beginning the survey. The study followed institutional

Table 2. Results of Mann–Whitney U-tests comparing SSQ ratings between the two groups (low and high self-rated musical competence). Bold text indicates observations with significant group differences; effect size is given for significant pairs

SSQ sub-domain	|Z|	*p*-value	Effect size, r_e
Speech perception	1.88	0.06	–
Spatial hearing	**2.09**	**0.03**	**0.39**
Sound qualities	**2.39**	**0.02**	**0.44**

Table 3. Results of Spearman correlation analysis (Sr) for the relationship between musical competence and three domains of SSQ

Association of musical competence with	S_r	*p*
Speech perception	0.43	0.02
Spatial hearing	0.50	0.006
Sound qualities	0.50	0.007

guidelines for biobehavioral research and was approved by the ethics committee. Participation in the study was voluntary and data confidentiality was ensured.

Statistical analysis

SPSS 25.0 software (IBM, Armonk, NY, USA) was used for statistical analysis. Speech perception, spatial hearing, sound qualities, and overall scores were the dependent variables in the study whereas musical competence of the participants was the independent variable. A Shapiro–Wilks test of normality was conducted to check if the data was normally distributed. A Mann–Whitney test was performed to check for group differences. A Spearman correlation test was administered to check the correlation between competence ratings on ELMEQ and self-perceived ratings of speech perception, spatial hearing, and sound qualities.

Results

A Shapiro–Wilk test revealed non-normal distribution ($p > 0.05$) of the data. The median scores along with the individual scores for each sub-section of SSQ are shown in **Figure 1**, revealing that participants with high self-perceived musical competence gave higher median ratings on all sub-sections of SSQ. These observations were statistically confirmed with a Mann–Whitney *U*-test. Results of the *U*-test (**Table 2**) revealed significant differences between groups categorized by self-perceived musical competence. Participants with higher self-perceived musical competence consistently demonstrated significantly higher ratings in spatial hearing ($p = 0.04$) and sound qualities ($p = 0.01$) compared to their counterparts with lower musical competence. However, such group differences were absent for speech perception ratings ($p = 0.06$), where both musical groups had similar ratings.

Spearman correlational analysis showed a moderate to strong correlation between self-perceived musical competence (measured by ELMEQ scores) and ratings on different SSQ domains (speech perception, spatial hearing, and sound quality) as shown in **Table 3**. While a moderate positive correlation was found between self-perceived musical competence and speech perception ratings ($S_r = 0.43$, $p = 0.02$) a strong positive correlation was observed between self-perceived musical competence score and both spatial perception rating ($S_r = 0.50$, $p = 0.006$) and sound qualities rating ($S_r = 0.50$, $p = 0.007$). In all the SSQ domains, the ratings tended to increase with an increase in self-perceived musical competence.

Discussion

The results of the study showed that there was a statistically significant effect of musical competence on spatial hearing and sound qualities perception in musicians. The low musical competence group produced lower ratings on both spatial hearing and sound qualities measures while the high musical competence group gave significantly higher ratings. However, these differences were not apparent for speech perception. The absence of group differences in speech perception lends support to the conclusion that both groups of musicians, irrespective of their self-perceived musical competency, enjoy equal advantage in the speech perception task.

The current study only pertains to self-perceived competence effects in instrumental musicians. However, the literature shows evidence, using a meta-analysis, that all musicians have an advantage on tasks related to speech in the presence of noise compared to non-musicians [12,20]. Inclusion of a non-musician group, and measuring self-perceived musical competence in them, would provide more insights.

In addition, in the present study the speech domain of SSQ comprised questions related to real-life listening, not those in noise. This discrepancy might explain the similar ratings between the two musical groups in speech perception, as musicians did not demonstrate a perceived advantage in everyday environments. Perhaps the incorporation of questions related to understanding speech perception in noisy conditions might reveal a concealed advantage.

Spatial hearing and the perception of sound qualities correlated better with high self-perceived musical competence. The higher the musical competence, the better were the scores in the spatial hearing and sound qualities domains. These findings support experimental studies that claim that musical experience has an effect on psychophysical abilities such as pitch perception and spatial hearing [1,4,8]. The better scores in the sound qualities domain in the high competence musician group can be explained by the evidence that musical training facilitates pitch discrimination abilities [6,7]. Positive correlation of musical competence with spatial hearing ability is documented in speech-in-noise studies [1,12]. Musicianship can also improve the

ability to segregate concurrently occurring sounds [21]. Better auditory stream segregation could be another possible reason for better spatial hearing abilities in the group with higher musical competence [10,22].

Overall, the impact of self-perceived musical competence on spatial hearing and sound qualities measures of SSQ suggest it would be useful to measure self-perceived musical competency in musicians prior to their inclusion in musicology studies. The presence of musicians with poor self-perceived competence might be a possible reason for seeing a lack of significant advantage, or reduced effect size, in findings from auditory processing tests. The findings of this study also emphasize the need for understanding domain-specific effects of musical competence using a variety of both psychoacoustical and electrophysiological tests. Future research should explore the impact of self-perceived musical competence across diverse populations (vocalists, different musical genres), and consider additional factors such as formal music education and early exposure to music.

This study has focused on musicians and their self-perceived expertise. Future research could encompass non-musical groups as well. Variables such as formal music education and childhood exposure to music could also be included. A screening questionnaire could be useful to assess these factors.

Conclusions

We found that self-rated musical competence was correlated with improved perceptions of spatial hearing and sound qualities. The group with high self-rated musical competence generally had better spatial hearing and more acute perception of sound qualities compared to the group who rated their musical competence as low.

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ISSN: 2083-389X
eISSN: 2084-3127

J Hear Sci, 2024; 14(1): 40–46
DOI: 10.17430/jhs/188537

MAJOR SALIVARY GLAND TUMORS: SINGLE INSTITUTION EXPERIENCE

Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

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Abstract

Introduction: The study analyses the occurrence and treatment outcomes of tumors of the major salivary glands in our patients.

Material and methods: Between 2019 and March 2023, a total of 182 salivary gland surgeries were conducted at our clinic. Of these, 111 were on females aged 18 to 76 years, and 71 on males aged 18 to 82 years. Diagnosis comprised a medical history, otolaryngological physical examination, ultrasonography (USG), fine-needle aspiration (FNA) biopsy of tumor, laboratory tests (CBC, CRP), and contrast-enhanced computed tomography (CECT) or magnetic resonance imaging (MRI).

Results: Predominantly (176 patients), benign tumors and neoplasms were found in the parotid salivary gland. Histopathology most commonly revealed pleomorphic adenoma (PA, 74 cases) and Warthin's tumor (WT, 70 cases). Other benign tumors and neoplasms made up only 18% of cases. Malignant neoplasms were only identified in the parotid salivary gland in 3.3% of cases; these were diagnosed as epithelial-myoepithelial carcinoma, adenoid cystic carcinoma, acinic cell carcinoma, mucoepidermoid carcinoma, and ductal carcinoma.

Conclusions: In our dataset, most patients with tumors of the major salivary glands were operated on at ages above 60 years. A low incidence of malignant tumors and a high incidence of WT of the large salivary glands were observed. The main postoperative complication was facial nerve paresis in 7.6% of cases.

Keywords: tumor occurrence • treatment outcomes • major salivary glands

GŁÓWNE NOWOTWORY GRUCZOŁÓW ŚLINOWYCH: DOŚWIADCZENIA JEDNEJ INSTYTUCJI

Streszczenie

Wprowadzenie: Celem pracy była analiza występowania i wyników leczenia guzów dużych gruczołów ślinowych oparta na materiale własnym.

Materiał i metody: W okresie od 2019 do marca 2023 w Klinice przeprowadzono 182 operacje gruczołów ślinowych, w tym 111 u kobiet, w wieku od 18 do 76 lat, a 71 u mężczyzn, w wieku od 18 do 82 lat. Diagnostyka dużych gruczołów ślinowych obejmowała: wywiad, badanie przedmiotowe otolaryngologiczne, badanie ultrasonograficzne (USG) oraz biopsję aspiracyjną cienkoigłową (BACC) (guza), badania laboratoryjne (morfologia, CRP) oraz badanie tomografii komputerowej z kontrastem (TK) lub rezonansu magnetycznego (MRI) w celu dokładnej oceny zmian w gruczołach ślinowych.

Wyniki: Wśród operowanych zmian nowotwory łagodne i guzy występowały jedynie w śliniance przyusznej u 176 chorych, z czego w badaniu histopatologicznym najczęściej stwierdzono: gruczolaka wielopostaciowego (PA) – w 74 przypadkach i guza Warthina (WT) – w 70 przypadkach. Pozostałe guzy i nowotwory łagodne stanowiły zaledwie 18,18% i zaobserwowano je w pojedynczych przypadkach, w tym: gruczolaka kwasochłonnego, torbiel limfocytowo-nabłonkową, gruczolaka mioepitelialnego, torbiel zastoinową, gruczolaka kanalikowego, torbiel z cechami metaplazji płaskonabłonkowej, zmianę limfoepitelialną, chłoniaka, gruczolaka limfatycznego i naczyniaka limfatycznego. Nowotwory złośliwe stwierdzono jedynie w śliniance przyusznej w 3,29% i histopatologicznie rozpoznano następujące nowotwory złośliwe: rak nabłonkowo-mioepitelialnokomórkowy, rak gruczołowo-torbielowaty, rak zrazikowo-komórkowy, rak śluzowo-naskórkowy, rak przewodowy.

Wnioski: W analizowanym materiale chorzy z guzami dużych gruczołów ślinowych najczęściej operowani byli w wieku 61–70 lat oraz powyżej 70 lat. Obserwowano niską częstość występowania nowotworów złośliwych oraz wysoką częstość występowania WT dużych gruczolów ślinowych. Głównym powikłaniem pooperacyjnym był niedowład nerwu twarzowego (7,60%).

Słowa kluczowe: występowanie nowotworów • wyniki leczenia • duże gruczoły ślinowe

Introduction

Salivary gland tumors are divided into benign and malignant, and according to the WHO classification from 2022 [1] can be further divided into non-cancerous epithelial lesions, benign epithelial tumors, malignant epithelial tumors, and mesenchymal tumors specific to the salivary glands. According to the WHO, the most common salivary gland cancer is mucoepidermoid carcinoma, followed by adenoid cystic carcinoma.

Pathologies of the salivary glands include non-neoplastic lesions, which include inflammations of various etiologies, cysts, developmental abnormalities, and salivary parenchymal lesions in the course of systemic diseases. The other group are neoplastic lesions, among which a distinction is made between benign and malignant tumors. The overall incidence of salivary gland tumors varies worldwide from approximately 0.4 to 13.5 cases per 100,000 individuals. Tumors originating from the salivary glands are relatively rare and account for approximately 3–4% of all head and neck cancers [2].

According to the Polish National Cancer Registry, in recent years head and neck cancers have accounted for 5.5–6.2% of all malignant tumors, which translates into about 5,500 to 6,000 new cases a year [3]. In 2015, a total of 347 new cases of malignancies of the major salivary glands were registered, and 181 men and 52 women died from salivary gland cancer [3].

Almost half of minor salivary glands tumors are benign. This discrepancy in the literature results from a variation in the center where the research was conducted. In oncological surgery centers, malignant tumors of the small salivary glands predominate, while in pathology centers benign tumors of the small salivary glands are most common.

In 2015, the crude incidence of malignant tumors of the major salivary glands in Poland was 0.3/100,000 (1.0 for parotid gland) for men and 0.2/100,000 for women (0.8 for parotid gland). In men and women respectively, there were 58 vs 46 new cases of malignant neoplasms of other and unspecified major salivary glands (and 181 vs 166 cases of malignant neoplasms of the parotid gland).

Salivary gland tumors are a heterogeneous group of tumors due to the complex embryogenesis of the salivary glands. The most common benign tumors are adenomas, i.e. pleomorphic adenoma (PA) and Warthin's tumor (WT); less common are cystic lymphadenoma, lymphangioma, and hemangioma (cystic hygroma) [4].

Malignant tumors account for approximately 25–30% of salivary gland tumors and include adenocarcinoma, acinic cell carcinoma, adenoid cystic carcinoma, carcinoma ex PA, and malignant lymphomas (MALT type, B-cell tumor, and also metastases of other malignancies) [4].

The incidence of malignancy depends on the type and location of the salivary gland. In the parotid gland, malignant neoplasms account for approximately 30%, in the submandibular gland, 50%, and in the sublingual gland, 90%. In the minor salivary glands, however, malignant neoplasms most commonly affect the tongue, floor of the mouth, retromolar area, and lower lip. In contrast, benign tumors are more common in the upper lip and buccal mucosa. Tumors in the area of the palate are 50% malignant [5].

The main risk factors for salivary gland cancer are exposure to radiation and dust, as well as addiction to nicotine (specifically associated with WT) [4]. Early diagnosis and introduction of appropriate therapy are of paramount importance and, in the case of malignant lesions, in the long-time prognosis [2].

In the case of neoplastic lesions, the first symptom is usually a tumor in the salivary gland area. It is usually non-painful and solid, with varying degrees of mobility in relation to the surrounding substrate. Symptoms suggestive of a malignant tumor are facial nerve palsy, skin infiltration, soreness, or concomitant enlargement of lymph nodes in the neck [2,6,7].

This study analyses the incidence and treatment outcomes of major salivary gland tumors in our clinic.

Material and methods

Between 2019 and March 2023, 182 salivary gland surgeries were performed in the Clinical Department of Otolaryngology, Head and Neck Surgery at the 4th Military Teaching Hospital and Polyclinic in Wrocław, comprising 111 women (61%) aged 18 to 76 years (mean age 67.0 years), and 71 men (39%) aged 18 to 82 years (mean age 63.0 years).

The diagnoses were based on a medical history, otolaryngological physical examination, ultrasonography (USG), fine-needle aspiration (FNA) biopsy, laboratory tests (CBC, CRP), and contrast-enhanced computed tomography (CECT) or magnetic resonance imaging (MRI) from which a detailed evaluation of the salivary glands, and location of lesions and lymph nodes was made.

The primary treatment was surgical resection of the salivary gland tumor under general endotracheal anesthesia. For benign, encapsulated tumors (e.g. WT), treatment involved removal of the tumor itself (so-called enucleation, local excision of the tumor, extracapsular dissection of the tumor) or removal of the tumor with part of the gland (so-called tissue margin). In some cases, partial parotidectomy, i.e. removal of the superficial lobe while sparing facial

Table 1. Number of patients, by gender and year, who underwent surgery

Year of surgery	Women		Men		Total	
	n	%	*n*	%	*n*	%
2019	34	18.7	20	11.0	54	29.7
2020	25	13.7	16	8.8	41	22.5
2021	22	12.1	13	7.1	35	19.2
2022	25	13.7	18	9.9	43	23.6
2023	5	2.8	4	2.2	9	4.9
Total	111	61.0	71	39.0	182	100.0

nerve function, was performed. There was no monitoring of the facial nerve during surgery.

Since the study was descriptive and not experimental, it did not require Bioethics Committee consent. Permission was obtained from the Commandant of the 4th Military Teaching Hospital in Wrocław to use the medical records for research purposes.

All statistical calculations were performed using Statistica version 14 (Tibco Software Inc., Palo Alto, CA, USA). The results of the study were statistically analysed and a Shapiro–Wilk test was used to assess the normality of age distribution, a Mann–Whitney *U*-test to compare the age difference between men and women, and contingency tables and chi-square tests for homogeneity and independence of distributions. Logistic regression was used to assess risk factors for postoperative complications. Results were considered statistically significant when p was < 0.05.

Results

The average age of the patients who underwent surgery was 61.0 ± 14.5 years (median = 65). Women were older than men by an average of four years (67 vs 63 years) but this difference was not statistically significant ($p = 0.153$).

Patients with tumors of the major salivary glands were most often above 60 years: 37 women (20.3%) and 18 men (9.9%) were aged 61–70 years, and 34 women (18.7%) and 19 men (10.4%) were aged over 70 years. There were few cases in the youngest age group: 2 women and 5 men aged 18–30 years.

The distribution of the ages of the women and men who underwent surgery differed significantly from a normal distribution (Shapiro–Wilk test). A chi-square homogeneity test revealed that there was no significant difference between the age distributions of women and men ($p = 0.422$).

Table 1 shows that between 2019 and 2023, a total of 182 patients underwent surgeries for tumors of major salivary glands. In each year there was no statistically significant difference between men and women in the percentage of surgeries ($p = 0.983$). Over the period 2019–2022, there was a negative trend in the number of operated patients, but the linear correlation coefficient ($r = -0.635$) was not significantly different from zero ($p = 0.365$).

Table 2 shows that, of the operated lesions, benign neoplasms and tumors were found only in the parotid gland in 176 patients (96.7%), with the most frequent histopathological findings of PA in 74 cases (42.0%), including 51 women (30.0%) and 23 men (13.1%), and WT in 70 cases (39.8%), including 41 women (23.3%) and 29 men (16.5%). Other benign tumors and neoplasms accounted for only 18.2% and were observed as cases of oncocytoma, lymphoepithelial cyst, myoepithelioma, retention cyst, tubular adenoma, cyst with features of squamous metaplasia, lymphoepithelial lesion, lymphoma, lymphoid adenoma, and lymphangioma.

Benign tumors and neoplasms occurred on the left side in 93 cases (52.8%), including women in 56 cases (31.8%) and men in 37 cases (21.0%), compared to 83 cases (47.2%) of the right, including 51 women (29.0%) and 32 men (18.2%).

The difference in the percentage of tumors located on the right and left side was not significant ($p = 0.337$). Among the 111 women who underwent surgery, tumors and benign neoplasms were diagnosed in 107 (96.4%), and among the 71 men who underwent surgery, tumors and benign neoplasms were diagnosed in 69 (97.2%). The difference in the percentage of benign tumors was not significant ($p = 0.768$).

Malignant tumors were only found in the parotid gland in 6 cases (3.29%). Histopathologically, the following malignancies were diagnosed: epithelial-myoepithelial carcinoma (pT3N0MO), adenoid cystic carcinoma (T2N0M0), acinic cell carcinoma (T2N0M0), mucoepidermoid carcinoma (T2N0M0), and ductal carcinoma (G2 pTxN2b). None of the genders or sides of the body were privileged in terms of diagnosis ($p > 0.05$).

Table 3 shows that, of the surgical methods used, the most frequent were: tumor enucleation in 102 cases (56.0%) and partial parotidectomy in 62 cases (34.1%); other methods were less frequent, including tumor removal with a margin of healthy tissue in 12 cases (6.6%) and total parotidectomy in 6 cases (3.3%). In the case of a malignant tumor, the lymph nodes of the neck were also removed (level I/II).

The frequency of surgery performed on patients with salivary gland tumors did not depend on the surgical method, side of the body, or gender ($p = 0.664$)

Table 2. Number of patients by tumor location, histopathological diagnosis, side of body, and gender

Histopathological diagnosis	Parotid gland				Total	
	Women		Men		*n*	%
	R	L	R	L		
Pleomorphic adenoma (PA)	24	27	11	12	74	42.0
Warthin's tumor (WT)	20	21	14	15	70	39.8
Oncocytoma	–	–	–	1	1	0.6
Lymphoepithelial cyst	1	1	1	1	4	2.3
Myoepithelioma	1	1	–	–	2	1.1
Retention cyst	1	3	0	3	7	4.0
Tubular adenoma	1	1	1	0	3	1.7
Cyst with signs of squamous metaplasia	–	–	–	1	1	0.6
Lymphoepithelial lesion	–	1	–	–	1	0.6
Lymphoma	–	1	1	1	3	1.7
Lymphadenoma	2	–	4	2	8	4.5
Lymphangioma	1	–	–	1	2	1.1
Total	51	56	32	37	176	100.0

Table 3. Number of salivary gland tumors by location, surgical method, side of body, and gender

Surgical method	Parotid gland				Total	
	Women		Men		*n*	%
	R	L	R	L		
Tumor enucleation	24	36	18	24	102	56.0
Tumor removal with a margin of healthy tissue	5	1	4	2	12	6.6
Total parotidectomy	3	1	–	2	6	3.3
Partial parotidectomy	22	19	10	11	62	34.1
Total	54	57	32	39	182	100.0

Table 4. Results of logistic regression of occurrence of complications

Risk factor for postoperative complications	Regression analysis				
	univariate		multivariate		
	b	*p*	beta	*p*	*OR* (95% CI)
Male gender	0.533	0.085	0.500	0.129	1.65 (0.86–3.14)
Days of hospital stay	0.314	0.018	0.301	**0.029**	1.35 (1.03–1.77)
Left side of body	0.846	0.006	0.784	**0.014**	2.19 (1.18–4.08)
Total parotidectomy	0.530	0.082	0.174	0.610	1.19 (0.61–2.33)
Tumor removal with a margin of healthy tissue	–1.216	0.077	–0.821	0.284	0.44 (0.10–1.99)
Parotid fistula	–1.895	0.084	–2.227	**0.048**	0.10 (0.01–0.98)

Legend: *b,* regression coefficient; *p,* significance; beta, multivariate regression coefficient; OR, odds ratio; 95% CI, odds ratio confidence interval. Risk factors significant at $p < 0.05$ are in bold. The logistic model for estimating the probability of a complication takes the form: Logit $P\,(complication = 1/X) = -1.67 + 0.30$ * days of hospital stay + 0.78 * left side – 2.23 * retention cyst.

The following postoperative complications were found: facial nerve paresis in 12 cases (7.6%), postoperative hematoma in 8 cases (4.4%), complete facial nerve palsy in 2 cases (1.1%), and tumor recurrence in 1 case (0.55%). The incidence of postoperative complications did not depend on gender ($p = 0.398$) or side of the body ($p = 0.294$). Logistic regression was used to assess the effect of the analyzed variables on the likelihood of surgical complications, and the results are shown in **Table 4**. In univariate analysis, factors contributing to surgical complications (stimulants) were the number of days of hospital stay and tumor location on the left side of the body. In the multivariate analysis, the number of days of hospital stay and the left side of the body were again the stimulants and, further, a retention cyst proved to be a destimulant. The odds of a postoperative complication in the group of patients with a tumor on the left side are more than twice as high compared to a tumor on the right side (OR = 2.19). The odds of a complication in patients staying one day longer in hospital are 1.35 times higher (OR = 1.35). The presence of a parotid fistula reduces the odds of a complication 10-fold (OR = 0.10; 1/OR = 10).

Patients with facial nerve paresis received galantamine injections (2.5–5 mg) for 14 days, vitamins B12, B6, and B1 (Milgamma N) 2 ml for 5 days; and physiotherapy. Patients were hospitalised for an average of about 4 days.

Discussion

An analysis of the gender structure in salivary gland pathologies in the available literature reveals some discrepancies. Most studies describe a prevalence of benign salivary gland neoplasms among women [8–10], although some authors report a male predominance [4,11]. The difference may be related to ethnic and geographical factors. Patients with malignant neoplasms are predominantly male in most publications. A similar relationship has also been observed in a Danish analysis based on 1,601 cases of malignant neoplasms: women accounted for 52% ($n = 832$) of surgically treated patients, while men accounted for 48% ($n = 769$) [6].

As far as histopathological diagnosis is concerned, the available literature records findings similar to those here. The predominant histopathological diagnosis among surgically treated salivary gland lesions was benign neoplasms, and among these, PA and WT [8,12–15]. In our work, PA was present in 42.5% and and WT in 39.8% of cases.

Analysis of the variation in the location of lesions in the major salivary glands showed that the majority of cases requiring surgical treatment involved the parotid gland, which was the most common location for both non-neoplastic lesions and neoplasms [4,8,9].

In one Mexican study ($n = 164$), there were different proportions of salivary gland pathology, but these were from an oral pathology center. Their study was dominated by indications for surgery due to pathology of the minor salivary glands, which accounted for 68.9% [8].

A study in northern Greece on 207 patients by Poutoglidis et al. [16] found that benign neoplasms accounted for 87.9% of cases. The most common neoplasm was WT, with a prevalence of 46.8%; the second most common was PA (31.9%). A higher incidence of parotid gland tumors was found in men ($p = 0.025$) and smokers ($p = 0.001$).

Jaremek-Ochniak et al. [17] reported 407 salivary gland neoplasms in their analysed dataset (over 11 years), of which malignant neoplasms accounted for 17.4%. The most common were adenoid cystic carcinoma (28.2%), mucoepidermoid carcinoma (12.7%), and serous cell carcinoma (9.9%). Lymphomas also represented a large group (1.5%). The predominant benign neoplasms were PA (54.1%) and WT (36%). Tumors of the salivary glands most commonly affected the parotid gland (92%).

In the literature, among the multiple histopathological types of salivary gland malignancies, one can observe that several diagnoses predominate. Among those investigated by Mengi et al. [11], the most common malignancies were mucoepidermoid carcinoma with 26 cases (24.3%), acinic cell carcinoma (9.3%), and adenoid cystic carcinoma (8.4%).

In the dataset analysed at the Medical University of Santa Catarina, Brazil, as well as among the Danish population, mucoepidermoid carcinoma predominated [6].

In Poland, the most common histopathological diagnoses were adenoid cystic carcinoma, adenocarcinoma, squamous cell carcinoma, and mucoepidermoid carcinoma [4,7].

Sowa et al. [18] assessed the effect of systemic oxidative stress in patients with selected benign and malignant parotid tumors. Patients with all parotid gland tumors included in the study had elevated plasma lipofuscin (LPS) levels. Furthermore, Cu/Zn-SOD activity in patients with WT was significantly lower than in the control group, the pleomorphic adenoma group, and the mucoepidermoid carcinoma group.

The surgical treatment of most conditions is determined by current surgical standards. There is also a widely accepted classification of salivary gland surgery developed by the European Salivary Gland Society (ESGS) [19]. The operative report of the ESGS indicates the level of removal of glandular parenchyma marked I to V and the non-glandular structures removed. Wong and Shetty [19] proposed an additional subdivision of levels I and II for the parotid glands and to divide them into levels Ia, Ib, IIa, and IIb based on facial nerve branches. The proposed sublevels make it possible to improve the description of key structures and thus increase the reliability of the operational protocol. Such meticulous reporting aims to optimise the management of complications and the planning of re-operations, emphasising the importance of an unambiguous classification system and a comprehensive surgical protocol.

In the present study, surgery involved tumor enucleation in 56.0% and partial parotidectomy in 34.1%, while other methods were used less frequently (tumor removal with a margin of healthy tissue in 6.6% and total parotidectomy in 3.3%).

In a study by Poutoglidis et al. [16], the majority of patients were treated using extracapsular dissection (60.4%) or partial superficial parotidectomy (22.6%). In 12 cases (5.7%), there was a recurrence of the lesion.

Combination therapy, i.e. surgery (total parotidectomy with facial nerve resection) and postoperative radiotherapy, is also used to treat malignant neoplasms.

Tumor recurrence depends on factors such as the size of the primary tumor, the presence of satellite tumors, incomplete tumor resection, or capsular rupture [19–21]. The decision to reoperate should take these factors into account, and be based on a physical examination, ultrasound, computed tomography, and/or magnetic resonance imaging. Data from the primary surgery should shed light on the likelihood of tumor spread, and patients to be identified who need to be followed up regularly [20,22]. Knowledge of all these factors reduces the risk of intra- and postoperative complications associated with reoperation. For example, if there is contact of the tumor with the trunk or branches of the facial nerve, or its close proximity to the trunk or branches, then before reoperation it is important to consider whether the nerve was previously dissected and exposed; if so, it may take a long time during the reoperation to localise the nerve within the fibrous tissue and preserve it [20].

If a tumor recurs, there are often difficulties in choosing the most appropriate treatment because there can be inconsistent expectations regarding observations in the surgical field, multifocal tumor spread, and scarring. Therefore, complete and accurate reporting of intraoperative observations should be performed during both primary and revision parotid gland surgery, following both the ESGS guidelines and those prepared in the operative report scheme proposed by Piwowarczyk et al. [23].

The standard treatment for pleomorphic adenoma (PA) of the parotid gland is radical surgical management. Radiotherapy (RT) as a primary treatment is controversial and not widely used. However, RT may be considered as an adjuvant therapy in some selected cases.

Piwowarczyk et al. [23] discussed the indications for RT in patients with parotid gland PA, based on the currently available published studies and their own experience. They recommended personalised treatment for each patient, based on the decision of a multidisciplinary panel of specialists. Adjuvant RT should be considered in cases of suboptimal resection of primary PA (close margins, intraoperative capsular disruption or tumor disintegration, risk of recurrence based on clinical factors and histological features) and in cases of recurrent PA. Recommended doses and techniques of radiation therapy were determined, depending on the clinical stage of the primary or recurrent tumor.

Although several reports in the literature document the surgical techniques and oncological outcomes obtained after parotidectomy, only a few describe the complications of parotid surgery and their management. Several complications have been reported after parotid gland surgery, which can be divided into intraoperative and postoperative (early and late). The most common complications after parotidectomy include temporary or permanent facial nerve palsy and Frey's syndrome [24]. The present study found the following postoperative complications: facial nerve paresis in 7.6%, postoperative hematoma in 4.4%, complete facial nerve palsy in 1.1%, and tumor recurrence in 0.6%.

In a study by Poutoglidis et al. [16], the most common complications were facial nerve damage, Frey's syndrome, and postoperative haematomas.

Benign salivary gland tumors have an excellent prognosis after complete surgical resection, and there is no need for adjuvant radiotherapy. In early-stage low-grade cancers, such as adenocarcinoma, mucoepidermoid carcinoma, or acinar cell carcinoma, postoperative radiotherapy (PORT) is not indicated if adequate margins are achieved [25]. However, for patients with high-risk factors, such as high-grade lesions at an advanced stage (T3 and more), positive surgical margins, perineural, vascular or lymphatic infiltration, lymph node involvement (especially extracapsular extension, ECE+), and skin and nerve infiltration, almost always PORT is beneficial for all adenomatous carcinomas [25].

Conclusions

In the analysed dataset, patients with tumors of major salivary glands were most frequently operated on beyond the age of 60 years. A low incidence of malignant tumors and a high incidence of WT of the large salivary glands were observed. The main postoperative complication was facial nerve paresis in 7.6% of cases.

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