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Review Article

Summary of longitudinal outcomes of cochlear implantation in children with congenital single-sided deafness in Belgium

Zusammenfassung der Langzeitergebnisse von Cochlea-Implantaten bei Kindern mit angeborener einseitiger Taubheit in Belgien

Abstract

Children with single-sided deafness (SSD) experience difficulties with localizing sounds and understanding speech in noisy environments and may also be at risk for speech-language delays. Providing a cochlear implant (CI) at an early age may support performance across multiple domains. Between 2014 and 2024, three groups of children were followed up at regular intervals: children with SSD who had received a CI in the deaf ear at a very early age, children with SSD, and children with bilateral normal hearing. Longitudinal analyses show that the children with SSD who did not receive a CI were at risk for grammar skills, poorer speech perception in noise, sound localization, and verbal IQ. Children with SSD and CI were on par with normal hearing children regarding grammar and speech perception in noise. An overview of the main findings are presented here. These confirm that children with congenital SSD can benefit from a CI provided at an early age to support their development across multiple domains. The longitudinal data led to a federal policy change in health care in Belgium.

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Zusammenfassung

Kinder mit einseitiger Taubheit (Single-sided Deafness, SSD) haben Schwierigkeiten, Geräusche zu lokalisieren und Sprache in lauter Umgebung zu verstehen. Zudem besteht ein erhöhtes Risiko für Sprachentwicklungsverzögerungen. Die frühzeitige Versorgung mit einem Cochlea-Implantat (CI) kann die Entwicklung in verschiedenen Bereichen fördern. Zwischen 2014 und 2024 wurden drei Kindergruppen regelmäßig untersucht: Kinder mit SSD, die bereits im frühen Kindesalter ein CI im tauben Ohr erhalten hatten, Kinder mit SSD und Kinder mit beidseitig normalem Hörvermögen. Längsschnittanalysen zeigen, dass Kinder mit SSD ohne CI ein erhöhtes Risiko für Defizite in Grammatik, Sprachverstehen im Störgeräusch, Schalllokalisation und verbaler Intelligenz aufwiesen. Kinder mit SSD und CI erreichten hinsichtlich Grammatik und Sprachverstehen im Störgeräusch das Niveau normalhörender Kinder. Die wichtigsten Ergebnisse werden hier zusammengefasst. Sie bestätigen, dass Kinder mit angeborener SSD von der frühzeitigen CI-Versorgung profitieren, um ihre Entwicklung in verschiedenen Bereichen zu unterstützen. Die Längsschnittdaten führten zu einer Änderung der Gesundheitspolitik in Belgien.

Introduction

Listening with two ears supports sound localization, speech perception in noisy environments, spatial awareness, listening ease, and the development of spoken language (for an overview, see [1]). Sensorineural hearing loss (HL) is present at birth in approximately 1.86 per 1,000 newborns in developed countries, with 30–40% of cases being unilateral. In roughly one-third of these unilateral cases, hearing loss is so profound (>90 dB HL), that a hearing aid on the deaf side is not effective.

The absence of binaural input, combined with reduced audibility, affects preverbal vocalizations in infants with unilateral hearing loss [2]. These early deficits can cascade across multiple developmental domains, leading to persistent difficulties with behavior and academic performance (e.g., [3]). Importantly, for some children, these challenges do not resolve naturally with age and may require targeted intervention.

A cochlear implant (CI) is the only intervention that can facilitate binaural hearing, as it transmits sound to the brain via electrical stimulation of the auditory nerve on the impaired side. To ensure optimal performance, a CI should be provided when neural plasticity is high, i.e., during the first few years of life [4]. Improved binaural processing is expected to facilitate speech-in-noise perception, thereby cascading into effects on spoken language development.

Qualification of a cochlear implant

However, not all children with congenital single-sided deafness (cSSD) qualify for a CI. A mock-up of the etiology of 180 children at the University of Leuven Hospital (UZL) and the University of Antwerp Hospital (UZA) showed that the most common reason for exclusion was cochlear nerve deficiency [1]. These children do not qualify for a CI, as a functional auditory nerve is essential for the device to transmit sound effectively. Among children who did qualify and proceeded with implantation, the leading cause of cSSD was congenital cytomegalovirus (cCMV) infection, accounting for roughly 40% of implanted cases. Other qualifying etiologies included inner-ear malformations, trauma, and SSD of unknown origin, provided that imaging confirmed an intact cochlear nerve. Etiologies associated with progressive hearing loss, such as cCMV, were considered particularly compelling indications because of the potential risk to the better-hearing ear. This study emphasizes the importance of etiology-specific assessment, particularly cochlear nerve imaging, in determining which children with SSD are suitable candidates for CI.

CI in children with cSSD: the CICADE project

To draw evidence-based conclusions about cochlear implantation for children with cSSD, the research group of ExpORL initiated a longitudinal, multicenter study, CICADE, in the fall of 2014. CICADE is an acronym of Cochlear Implants in Children with one Deaf Ear. Over time, 16 children with congenital single-sided deafness (cSSD) received a cochlear implant in the University Hospital Leuven, the University Hospital Antwerp, the University Hospital Gent, or the European Institute for ORL-HNS. The average age of implantation was 13.6 months (SD 4.8 months), which falls within a developmental period characterized by high cortical plasticity. Every 6 months, linguistic, neurocognitive, and auditory processing skills were assessed in these children and in two control groups: children with cSSD without a CI and children with bilateral normal hearing (NH). The latter is necessary to disentangle the effects of maturation from those of intervention and to compensate for potential recruitment bias. This article summarizes the main findings from studies conducted in Belgium on the benefits of cochlear implantation in children with congenital single-sided deafness (cSSD).

Language development

We began evaluations during the first year after cochlear implantation, primarily using questionnaires, but these early outcomes did not reveal performance differences among the three groups. By the age of two, however, formal assessments became feasible. Preliminary data from 8 toddlers with a CI and 13 without a CI already showed that the children with a CI performed largely in line with their normal-hearing peers on language and cognitive measures [5]. Follow-up in a larger cohort two years later confirmed these findings: children with cSSD and a CI achieved significantly higher grammar scores than children with cSSD without a CI [6]. Overall, children with cSSD and a CI performed comparably to bilaterally normal-hearing children, while outcomes for children with cSSD but without a CI were more variable. Vocabulary and receptive language scores did not differ significantly between the three groups. Both studies suggest that early implantation may help mitigate or even prevent language delays associated with cSSD.

A follow-up study tested whether early CI supports storytelling and comprehension in children with SSD [7]. Implanted children performed on par with normal-hearing peers in narrative skills and verbal short-term memory, while non-implanted SSD children scored lower on both. Early grammar gains may further boost later narrative skills, reinforcing the case for early CI to support typical language development. Narrative skills depend on multiple component abilities: grammar, verbal short-term memory, vocabulary, etc.

Auditory development: speech understanding in noise and sound localization

Providing a CI to children with prelingual SSD also has a significant positive impact on the children's speech perception in noise performance [8], [9] and sound localization (for some children). On both tasks, they achieved scores that were overall poorer than those of the NH group, but much better than those of the cSSD group. Speech thresholds in noise are better for older children than for younger ones and better for children with cSSD and a CI than for children with cSSD without a CI when the speech is presented to the CI (or deaf) side and noise to the good ear. Nevertheless, children with NH outperform the children with cSSD and a CI on these tasks, suggesting that even when speech is presented frontally, children with SSD and a CI struggle more to understand it [9].

Regarding sound localization, the children were aged 3.9–8.3 years at the time of evaluation. In the most recent report [9]. Eleven out of the 18 SSD+CI children who participated in the test performed better-than-chance accuracy on at least one assessment. Many could not localize sounds at the first assessment, but showed gradual improvement with age. It is likely that the localization skills of some of them will further improve as they age [9].

Balance

Balance was assessed with the BOT balance scale. Children with SSD, regardless of CI use, scored lower than the NH group, with the poorest scores in those with CMV-related SSD. No difference was found between CI and non-CI children [9]. Notably, balance was evaluated only with the BOT scale, without a full vestibular assessment. We also examined how children with single-sided deafness (SSD) and a cochlear implant (CI) perceive two main acoustic voice cues: fundamental frequency (F0), which corresponds to the pitch of the voice, and vocal tract length (VTL), which shapes formants and provides information about speaker size and timbre. These data were cross-sectional (not part of the longitudinal study). Findings showed that early cochlear implantation offers clear benefits for perceiving voice cues, particularly F0, and supports tasks such as voice gender categorization [10]. However, challenges remain: discrimination of VTL is less accurate with the CI, and children gain only a limited advantage from voice-cue differences in more complex listening conditions, such as understanding speech in competing speech. Importantly, children with SSD and a CI actively use their CI ear. This highlights the importance of supporting consistent, full-time CI use to maximize benefits.

On average, the children wore their CI for about 8.3 hours per day [11]. More daily device use was significantly associated with better word perception in quiet. Thus, a longer duration of CI use – both in terms of years since implantation and daily wearing time – positively influenced speech perception outcomes, highlighting the importance of consistent CI use for optimal auditory development.

Summary

Although additional longitudinal follow-up is needed to draw firm conclusions for long-term use, our language and hearing studies indicate that early cochlear implantation enhances speech-in-noise perception, verbal IQ, and grammatical skills. These improvements may, in turn, help reduce barriers to cognitive, academic, and psychosocial development. The findings from this longitudinal research have already informed health policy in Belgium: since January 2024, children with cSSD up to 4 years of age and those with acquired SSD up to 18 years of age are eligible for CI reimbursement.

Several important research questions remain, including the optimal age for cochlear implantation and potential shifts in attitudes toward SSD and cochlear implants as children reach adolescence. It is important to note that deficits associated with (profound) unilateral hearing loss are typically more subtle than those observed in bilateral hearing loss. In the present study, all children received early implantation, which likely contributed to the favorable outcomes. Toddlers in the SSD_CI group wore their devices for many hours each day, a factor that likely enhanced their speech perception performance and underscores the importance of consistent CI use for optimal auditory development. Effective parental counseling and ongoing follow-up into adolescence, therefore, remain crucial.

Notes

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Competing interests

The author declares that she has no competing interests.

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