

# Single-sided deafness and unilateral auditory deprivation in children: current challenge of improving sound localization ability

Journal of International Medical Research  
48(1) 1–13

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DOI: 10.1177/0300060519896912

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## Abstract

Interaural time difference and interaural level difference signals are insufficient in patients with single-sided deafness (SSD). This insufficiency leads to an absence of sound localization abilities and a decrease in speech intelligibility in noisy environments. SSD occurring in children further affects their language learning and cognitive abilities and their academic performance because they lack spatial abilities and binaural hearing. The early stages of central auditory system development are critical for auditory function development and morphological refinement. SSD occurring in the critical period can cause significant lateralization in the bilateral auditory pathway. This may increase the risk that affected individuals cannot re-establish binaural benefits after rehabilitation of hearing loss in the post-sensitive period. For otorhinolaryngologists, there is the concern that children with congenital SSD cannot benefit from cochlear implantation. Only a few studies have investigated auditory rehabilitation in children with congenital SSD with cochlear implantation and their results were inconsistent. The present review aims to clarify the main problems and challenges of clinical rehabilitation of congenital SSD, particularly focusing on the effect of CI on sound localization ability in children with congenital SSD.

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## Keywords

Congenital single-sided deafness, unilateral auditory deprivation, sound localization, cochlear implantation, hearing loss, pure tone average, children, speech intelligibility

Date received: 9 September 2019; accepted: 3 December 2019

## Introduction

Hearing loss (HL) can be classified according to various perspectives, including the type, degree, and configuration of the hearing loss. Basing on the effect of HL on the central auditory system, HL can be divided into bilateral hearing loss, unilateral hearing loss (UHL), and asymmetric hearing loss (AHL) on the basis of its degree of severity and lateral differentiation.<sup>1</sup> UHL is defined as HL only in one ear and disregards the types and degrees of HL. If UHL is severe-to-profound, it is called single-sided deafness (SSD).<sup>1</sup> However, the exact definition of SSD is still controversial. One reason for this controversy is that there are different standards for classifying SSD in adults. Van de Heyning et al.<sup>2</sup> suggested that SSD in adults should meet all of the following criteria: (1) in the poorer ear, the pure tone average (PTA) (mean hearing threshold levels at pure tone frequencies of 0.5, 1, 2, and 4 kHz) is  $\geq 70$  dB HL; (2) in the better ear, the PTA is  $\leq 30$  dB HL; and (3) the interaural threshold gap is  $\geq 40$  dB HL. In contrast, Ramos Macías et al.<sup>3,4</sup> considered that the main factor for defining SSD is that the poorer ear or the “bad” ear does not receive a benefit when traditional acoustic amplification is applied, and the “good” ear must have a PTA that is 20 dB HL or better across the set of pure tones (0.5, 1, 2, and 4 kHz). Therefore, the criteria for SSD in adults still need to be further established. However, the principle of SSD in adults should be that the unilateral ear does not have effective auditory

function, while the contralateral ear has normal auditory function.<sup>4,5</sup> Furthermore, the PTA in young children may not be easily acquired, and children’s standards need to be incorporated into auditory steady-state evoked potentials and/or behavioural observational audiometry. The degree of HL in tone audiometry in the definition of SSD for young children may also be inconsistent with that in adults. Severe HL is suggested as a PTA between 61 and 80 dB of hearing loss at frequencies of 0.5, 1, 2, and 4 kHz, and a PTA of worse than 80 dB HL signifies deafness in children.<sup>6</sup>

AHL is considered as having HL in both ears, but with a difference in PTA between the two ears.<sup>1,2</sup> However, there are also differences in the definition of AHL in the literature,<sup>1,2,7</sup> and some authors consider that AHL includes SSD.<sup>7</sup> These differences are mainly due to the two aspects of PTA in the better-hearing ear and the interaural threshold gap. Vincent et al.<sup>7</sup> suggested that the PTA (0.5, 1, 2, and 4 kHz) in the better-hearing ear should be better than 60 dB HL, including normal hearing thresholds for cases of SSD. In contrast, Van de Heyning et al.<sup>2</sup> considered that the PTA (0.5, 1, 2, and 4 kHz) should be higher than 30 and  $\leq 55$  dB HL in the better-hearing ear. The interaural threshold gap was defined as a difference of 30 dB HL or higher (PTA = 0.5, 1, 2, and 4 kHz) by Vincent et al.<sup>1</sup> Additionally, interaural asymmetry in the PTA was also considered as being  $>15$  dB HL (PTA = 0.5, 1, and

2 kHz), >20 dB HL (PTA=3, 4, and 6 kHz),<sup>1</sup> or >15 dB HL (PTA=0.5, 1, 2, and 4 kHz).<sup>2</sup> Therefore, there is still a need for further consensus on the definition of AHL and its distinction from SSD.

Nevertheless, the prevalence of SSD is high and treatment of SSD is insufficient. SSD has been estimated to affect between 12 and 27 individuals in every 100,000 people in the general population, with the majority of HL being sudden and idiopathic.<sup>3</sup> The overall prevalence of UHL in adult Americans is 7.2%, with 1.5% (0.1% to 2.1%) experiencing moderate (or worse) UHL (SSD).<sup>8</sup> Additionally, nearly one third of those who experience at least moderate UHL report having trouble hearing. Hearing aid use is low, even when there is a perceived handicap.<sup>8</sup> The prevalence of SSD in children is also high. Despite the introduction of universal newborn hearing screening, UHL is sometimes recognized late.<sup>6</sup>

Another important type of congenital SSD is found in children with congenital bilateral deafness who only receive unilateral cochlear implantation (CI) in the early stage of development. In China, children with bilateral congenital deafness are detected early, diagnosed, and receive intervention because of successful and standardized universal newborn hearing screening.<sup>9</sup> CI has also helped more than 30,000 deaf children acquire hearing.<sup>10</sup> However, most children with bilateral deafness in China only receive CI in one side,<sup>10</sup> with the other ear without hearing, similar to SSD.

Having one normal-hearing ear used to be considered sufficient.<sup>1</sup> However, we have now recognized the importance of binaural hearing for development of normal auditory processing. Children with UHL require intervention to prevent impairment in speech and language development.<sup>1</sup> A delay in diagnosis and intervention in children with SSD has a negative effect on integration of binaural information. Interaural

time difference (ITD) and interaural level difference (ILD) signals are insufficient in patients with SSD. This insufficiency leads to the absence of sound localization ability and a decrease in speech intelligibility in noisy environments.<sup>11</sup> In children, when the sound environment is more complex, such as in classrooms, playgrounds and schools, sound localization is more challenging than that experienced in the “cocktail party effect”.<sup>12</sup> Therefore, language, cognitive, and academic performance could be affected in children with SSD because of impaired spatial abilities and binaural hearing. The main processes involved in spatial hearing and the binaural advantage are the head shadow effect, binaural unmasking, and binaural summation.<sup>13</sup>

In the early stages of development, a critical period of auditory function and morphological refinement is observed. At this stage, the auditory centre is vulnerable to the ambient environment and auditory inputs.<sup>14,15</sup> In children with congenital and early acquired SSD, monaural inputs can have an extensive effect on the development of brain networks related to higher-order cognitive function,<sup>4</sup> which may have a negative effect on spoken language processing.<sup>5</sup> Lieu<sup>16</sup> further showed that children with SSD also have worse language skills than their normal hearing peers, and they face the risk of learning problems at school.<sup>16</sup> At present, auditory rehabilitation in those with congenital SSD is still a great challenge. For otorhinolaryngologists, there is concern that children with congenital SSD cannot benefit from CI. The present review aims to clarify the main problems and challenges of clinical rehabilitation of congenital SSD, particularly focussing on the effect of CI on the sound localization ability in children with congenital SSD. We searched the literature in the PubMed database for the keywords “cochlear implantation” AND “single-sided deafness” AND

“congenital” or “cochlear implantation” AND “unilateral deafness” AND “congenital”. The publication date of the articles was up to May 2019. There were only six studies that investigated auditory rehabilitation in congenital SSD with CI.<sup>4,17-21</sup> A study by Távora-Vieira and Rajan<sup>21</sup> was not included because they had the same subjects as another study by Távora-Vieira and Rajan<sup>19</sup> The remaining five studies (Table 1)<sup>4,17-20</sup> were included and discussed in this review.

### **Importance of hearing rehabilitation of SSD in the early stages of development**

#### *Hearing rehabilitation for SSD in the sensitive period*

The auditory system has a remarkable ability to adjust to an ever-changing environment.<sup>15</sup> This ability is described as plasticity. In the early stages of development, the human central auditory system remains maximally plastic, and it is the critical period of auditory function development and morphological refinement, and relies on auditory experience.<sup>11,14,15</sup> In this period, the auditory centre is more susceptible to the ambient environment and auditory inputs. Because this period is also sensitive to central auditory adaptability after HL, SSD occurring at this time can cause significant lateralization in the bilateral auditory pathway.<sup>22-24</sup> However, this plasticity cannot last long and this critical period is just a small time window. Sharma et al.<sup>14,25,26</sup> considered that there is a sensitive period of approximately 3.5 years during which the human central auditory system remains maximally plastic. After the age of 7 years, plasticity is greatly reduced. Therefore, hearing rehabilitation in children with congenital SSD should be performed during this sensitive period.

#### *Delayed hearing rehabilitation of SSD leads to unilateral auditory deprivation*

Auditory deprivation (AD) mainly occurs when there is a lack of effective auditory stimulation in the auditory centre during the sensitive period. This lack of stimulation leads to weakening or loss of the ability to process auditory information in the central auditory system.<sup>11</sup> During the sensitive period, a decrease in unilateral auditory input due to SSD leads to a series of adaptive or compensatory morphological and functional changes in the auditory centre. This is called unilateral auditory deprivation (UAD).<sup>11</sup> Liu et al.<sup>11</sup> considered that UAD can distort tonotopic maps, disrupt binaural integration, reorganize neural networks, and change synaptic transmission in the primary auditory cortex or subcortex.<sup>11,27</sup> Younger individuals who suffer from SSD experience stronger plasticity of the auditory centre. This situation results in obvious degeneration of the affected side and adaptive enhancement of the contralateral side of the auditory pathway. Similarly, a longer duration of SSD can also lead to more obvious degeneration in the affected side and adaptive enhancement in the contralateral side.

Adaptive enhancement of the auditory pathway corresponding to healthy ears relies on monaural spatial cues that are available to the intact ear to improve sound localization in the horizontal plane.<sup>28</sup> In contrast, degeneration of the auditory pathway corresponding to SSD may be limited in re-reorganization from auditory deprivation, even after hearing rehabilitation in the post-sensitive period. This limitation may increase the risk that individuals cannot re-establish binaural benefits after hearing rehabilitation in the post-sensitive period. Therefore, patients with SSD should participate in hearing rehabilitation as soon as possible to

**Table 1.** Sound localization, behavioural audiometry, and the SSQ in children with congenital single-sided deafness with CI.

Author	Subject	Age at CI	Sound localization	Behavioural audiometry and the SSQ score
Arndt et al., 2015 <sup>18</sup>	Child A	4.3 y	A substantial increase in the localization error	Poor performance in audiological testing The SSQ score showed almost no improvement after CI compared with the preoperative condition
	Child B	13.8 y	A substantial decrease in the localization error 12 months after CI compared with that in the unaided condition	Poor performance in audiological testing SSQ scores were lower for spatial hearing and slightly higher for hearing quality with CI compared with scores in the unaided situation
	Child C	3.2 y	–	–
	Child D	1.8 y	–	–
Távora-Vieira and Rajan, 2015 <sup>19</sup>	S1	17 mo	–	S1 was too young to be assessed with formal measures
	S2	4.5 y	–	–
	S3	6.8 y	S3 did not achieve any improvement in localization ability	Speech perception in noise scores with CI on did not differ from those with CI off
Beck et al., 2017 <sup>20</sup>	I0	21–80 mo	–	(1) Children implanted at a younger age ( $\leq 38$ mo) showed measurable speech discrimination benefits, but older children ( $\geq 48$ mo) had lower speech discrimination scores (2) SSQ: An increase in the SSQ score was found in evaluation by parents; self-assessment of children failed to reach significance
Thomas et al., 2017 <sup>17</sup>	I4/21	5 y and 6 mo; 3 y and 0 mo	Lateralization ability ( $n = 14$ ) in the CI-aided condition was significantly better than that in the unaided condition when stimuli were presented on the deaf side and on the normal hearing side, but not when presented from the front ( $0^\circ$ )	Speech comprehension in noise was significantly better with CI than without CI SSQ: Scores on the parental questionnaire were significantly higher after CI than preoperatively before CI for all three subscales of speech, spatial, and quality

(continued)

Table 1. Continued

Author	Subject	Age at CI	Sound localization	Behavioural audiometry and the SSQ score
Ramos Macías et al., 2019 <sup>4</sup>	4/11	29 mo	Positive results were obtained in the Auditory Lateralization Test for the following modalities: 0°, 45°, and 90°	No results related to improvement in speech discrimination SSQ: The SSQ questionnaire results showed a significant improvement

Abbreviations: SSQ: Speech, Spatial, and Qualities of Hearing Scale; CI: cochlear implantation; y: years; mo: months.

obtain good binaural benefits, especially for sound localization.<sup>11</sup>

## How to assess UAD remains a challenge

Compensation in the auditory centre after SSD is a double-edged sword. Lateralization in the auditory pathway caused by UAD is persistent and difficult to reverse, even in long-term bilateral hearing.<sup>11</sup> This persistence may cause difficulty for children in obtaining binaural auditory advantages after CI. Therefore, judging the effect of hearing rehabilitation by accurately evaluating the degree of remodelling (severity of UAD) in children's auditory centre before auditory rehabilitation is helpful. However, there have been few quantitative studies on the degree of UAD. Based on the results of existing research, the following evaluations may reflect the severity of UAD.

### Behavioural audiometry

Behavioural audiometry includes test sound localization ability in the horizontal azimuth and/or speech intelligibility in noise. Horizontal azimuth localization is primarily based on encoding of the ITD and ILD. Spectral shape cues provided by the pinna are only used for localization in the vertical plane and barely provide horizontal azimuth information in the normal hearing population. Horizontal azimuth localization in patients with SSD is severely impaired because ITD and ILD cues are insufficient. However, Agterberg et al.<sup>28</sup> showed that SSD listeners can rely on monaural spectral shape cues from their normal ear for localization in the horizontal azimuth by adaptive compensation of the auditory centre.<sup>28,29</sup> Therefore, for children with SSD, improvement in sound localization abilities in the horizontal azimuth also reflects that the degree of compensation



(reorganization or remodelling) in the auditory centre has deepened. This suggests the severity of UAD to a certain extent. In noisy environments, the listener needs to selectively locate the target sound source and pay attention to it, which requires sound localization abilities. The lack of sound localization ability is parallel to a decrease in speech intelligibility in noise. Therefore, a test on speech intelligibility in noise may have a similar function as evaluation of sound localization ability. However, for small children, evaluating sound localization ability and speech intelligibility in noise objectively is difficult.<sup>19</sup>

### *Electrophysiological measures*

Electrophysiological tests can provide an objective and accurate evaluation index for central auditory information processing. By recording auditory evoked potentials, Gordon et al.<sup>23,24</sup> showed that over-strengthening of neural projections and the auditory cortex corresponding to unilateral stimulation (similar to SSD) resulted in abnormally strong contralateral lateralization of activity. Kral et al.<sup>30</sup> also showed unilateral aural preference caused by SSD within the early sensitive period, which demonstrated pronounced and rapid reorganization of the primary auditory cortex. Abnormal lateralization in the auditory cortex can reflect the degree of cortical remodelling and may also reflect the severity of UAD to a certain extent. These abnormal activities in the auditory cortex are associated with clinical findings of worse outcomes in the second implanted ear in bilaterally implanted children.<sup>30</sup> Sandmann et al.<sup>31</sup> used electroencephalography to examine cross-modal reorganization in the auditory cortex of post-lingually deafened CI users. These authors found visual take-over in the auditory cortex of CI users. Incomplete reversal of this deafness-induced cortical reorganization

might limit the clinical benefits of a cochlear implant and may help explain the high inter-subject variability in auditory speech comprehension. The above-mentioned results may be used as an index to predict the effects of hearing rehabilitation for SSD.

### *Other brain function assessment methods*

UAD leads to cross-modal reorganization of the auditory cortex. Similarly, the extent of cross-modal reorganization of the auditory cortex also reflects the degree of UAD. Task-related or resting-state functional magnetic resonance imaging (fMRI) is widely used to investigate cross-modal reorganization in the primary auditory cortex. Zhang et al.<sup>32</sup> found that the left primary auditory cortex (non-auditory-deprived cortex) in patients with left SSD was reorganized to accommodate visual and sensorimotor modalities through cross-modal plasticity. There are also other brain function assessment methods used in the study of auditory cortical activity, such as positron emission tomography and magnetoencephalography. Because these methods are objective, they are also used to investigate activation changes in the auditory cortex of patients with SSD or lateralization of cortical responses in normal hearing individuals.<sup>32</sup> However, there has not been any extensive research on the reliability, economy, and practicability of these methods clinically, and they are still not widely used at present. Currently, a new optical neural imaging technique called functional near-infrared spectroscopy (fNIRS) is used to determine whether pre-operative brain activity can explain the variability in CI results. Anderson et al.<sup>33</sup> showed that stronger preoperative cross-modal activation in auditory brain regions by visual speech was predictive of poorer auditory speech understanding after implantation. These authors suggested

that fNIRS imaging preoperatively may support more accurate predictions of individual CI outcomes. fNIRS has already been used in the same way as electroencephalography, fMRI, and the event-related potential technique, and it has become a powerful tool for exploring the human brain. With rapid progress in brain imaging technology, the value of fNIRS and fMRI is likely to be further exploited. A set of prognostic evaluation systems to be used before hearing rehabilitation may be formed on the basis of fNIRS, fMRI, sound localization ability, and auditory evoked potentials.

### **Improvement of sound localization ability in SSD**

The rehabilitation methods for UHL include the following: contralateral and ipsilateral routing of sound (CROS and IROS) hearing aids for air conduction (AC) and bone conduction (BC) devices; CI, which directly stimulates the spiral ganglion; and auditory brainstem implants, which directly stimulate the cochlear nucleus, according to the type and degree of hearing loss. For patients with SSD, the most widely used auditory rehabilitation measures are CROS hearing aids and CI.<sup>13,34,35</sup>

#### *Effect of CROS hearing aids on sound localization ability in children with SSD*

CROS hearing aids are designed for SSD, with both AC (AC-CROS) and BC (BC-CROS) types. These hearing aids use a microphone that picks up sounds on the deaf side and transmits the signal to the contralateral normal-hearing cochlea to obtain hearing information of the deaf side. Transmission of information from the CROS device is still based on a unilateral auditory pathway; therefore, achieving sound localization ability that represents an

advantage of binaural signal integration is difficult.<sup>35</sup> Ryu et al.<sup>36</sup> reported that wireless CROS (AC) provided increased satisfaction and overall improvement in localization and hearing. In contrast, more rigorous experiments have indicated that AC-CROS hearing aids cannot improve sound localization and also interfere with the monaural level and spectral cues that provide a basis for determining sound locations in the horizontal plane in patients with UHL.<sup>37</sup>

Because patients with SSD have only one functioning cochlea and treatment with a BC-CROS hearing aid does not restore binaural hearing, that sound localization abilities do not improve after applying a BC-CROS hearing aid is not surprising.<sup>38</sup> BC-CROS hearing aids are slightly different from AC-CROS hearing aids. Although the BC-CROS hearing aid is placed on the deaf side and picks up sound from the deaf side, signal transmission still uses the unilateral auditory pathway. However, the amplified signal is transmitted through the skull to the contralateral normal cochlea, and therefore, it may not interfere with the monaural localization signal. Therefore, BC-CROS hearing aids do not improve or deteriorate the localization abilities of patients with SSD.<sup>38</sup>

#### *Effect of CI on sound localization ability in children with acquired SSD*

CI is a superior treatment for auditory rehabilitation to AC and BC hearing aids for those with acquired SSD. CI can re-establish the benefits of binaural hearing in those with acquired SSD.<sup>39</sup> Liu et al.<sup>40</sup> showed that the ability of sound localization in most patients with acquired SSD (90%) with CI was significantly better than that of patients without CI. However, factors such as the age of onset, aetiology, and duration of SSD may interact with each other, affecting restoration of



binaural integration in acquired SSD.<sup>18</sup> As mentioned above, younger individuals with SSD have stronger plasticity in the auditory centre, leading to degeneration on the affected side and adaptive enhancement on the contralateral side of the auditory pathway. Similarly, a longer duration of SSD can also lead to more obvious degeneration on the affected side and adaptive enhancement on the contralateral side. Adaptive enhancement of the auditory pathway corresponding to the healthy ear may limit re-organization because of auditory deprivation, even after CI. This situation may increase the risk that a few individuals cannot re-establish binaural benefits after CI. Therefore, patients with acquired SSD should also accept CI as soon as possible to obtain good binaural benefits, especially for sound localization.<sup>40</sup>

### *Effect of CI on sound localization ability in children with congenital SSD*

Only five studies examined auditory rehabilitation in patients with congenital SSD with CI (Table 1).<sup>4,17–20</sup> Unfortunately, only a few of these cases have been reported in the literature, and even fewer children completed audiometric evaluation.<sup>4,18,19</sup> Furthermore, there is high heterogeneity in the children in different reports, leading to inconsistent results across the reports. The main results in the literature can be summarized as three points of view. The first point of view is that, in children with congenital SSD with CI, there are no improvements in the ability of sound localization and speech perception in noise. Távora-Vieira and Rajan<sup>19,21</sup> reported a 6.8-year-old child (S3) with congenital SSD and found that this child did not achieve any improvement in localization ability and speech perception in noise. Arndt et al.<sup>18</sup> reported results from a 4.3-year-old child (Child A) with congenital SSD who showed poor performance in

audiological testing. SSD showed almost no improvement in any of the three subcategories after CI surgery compared with measures in the preoperative condition. Ramos Macías et al.<sup>4</sup> reported results from four children with congenital SSD who showed no improvement in speech discrimination after CI. The second point of view is that sound localization and speech comprehension in noise significantly improve in children with congenital SSD with CI. Ramos Macías et al.<sup>4</sup> also reported results in four children with congenital SSD (average age: 29 months) and showed that lateralization test results improved after CI. Thomas et al.<sup>17</sup> reported significant promotion of true binaural hearing ( $n = 14$ ). The biggest improvement was in the combined head shadow effect of 2.11 dB. An improvement in lateralization ability was also found ( $n = 14$ ). The third point of view is that sound localization ability and speech audiometry scores of children with congenital SSD decrease after CI. Arndt et al.<sup>18</sup> reported results from a 4.3-year-old child (Child A) who showed a substantial increase in the localization error. They also showed that a 13.8-year-old child (Child B) with CI showed lower Speech, Spatial, and Qualities of Hearing Scale (SSQ) scores for spatial hearing and slightly higher scores for hearing quality compared with those in the unaided situation.<sup>18</sup> Therefore, the existing data are not in agreement regarding whether CI is beneficial or harmful to the sound localization ability of children with congenital SSD.

At present, there are many shortcomings in the literature, which reduces the value and reliability of study results as follows. First, as mentioned above, there are too few cases of auditory rehabilitation in congenital SSD with CI in the literature. Therefore, these cases cannot sufficiently reflect the natural characteristics of congenital SSD. Second, the heterogeneity of the samples may easily mask and interfere with

the characteristics of children with congenital SSD with CI. Heterogeneity of the cases includes the aetiology of congenital SSD,<sup>17</sup> the duration of congenital SSD, the implantation age of CI, the CI experience, the age at audiometric evaluation, the training experience after CI, and device use each day. Third, behavioural audiometry is a subjective evaluation that is limited by the developmental stage of children and the development level of the central nervous system, which could have affected the results of these reports. In particular, for small children, evaluating sound localization ability and speech intelligibility in noise objectively is difficult.<sup>20</sup> Finally, the subjective intentions of the authors may also have affected interpretation of their results.

## **Challenges and the main problems related to congenital SSD in children**

### *Age of CI in children with congenital SSD*

At present, there is concern that children with congenital SSD cannot benefit from CI. Based on previous results, the sound localization ability of children with congenital SSD after CI is not optimistic.<sup>18,19</sup> The ideal age of CI in children with congenital SSD is still a major problem related to improvement of sound localization ability. However, no specific age has been suggested for CI. Thomas et al.<sup>17</sup> considered that CI provided significant audiological and subjective benefits for children with congenital SSD, even in children who were implanted after the age of 3.5 years. In contrast, Arndt et al.<sup>18</sup> suggested that children with congenital SSD who are older than 4 years perform poorly in speech audiometry and sound localization on the basis of Távora-Vieira and Rajan's<sup>19</sup> results and their own data. Beck et al.<sup>20</sup> reported a case series study

that also appeared to suggest decreased performance when implantation occurs at 4 years or older. These authors found that speech audiometry results were lower at 4 years or older than those in children who were implanted at a younger age.

To date, the precise end of the possible CI window is unknown. Based on most previous results,<sup>4,18,19,21</sup> we consider that the results by Thomas et al.<sup>17</sup> may be too optimistic. These authors found that there was no difference in results between an implantation age of >6 years and <6 years,<sup>17</sup> which is debatable. Overall, early intervention for congenital SSD is particularly important. CI in children with congenital SSD provides some benefits of binaural hearing if implantation occurs within the critical period of auditory development.<sup>4,19,21</sup> The ideal implantation window in children with congenital SSD might be short, as expected, and may be at an age younger than 4 years.<sup>18,19,21</sup>

### *Rehabilitation training of patients with congenital SSD with CI*

UAD leads to asymmetrical changes in the sound localization pathway.<sup>11,23,24,30,41</sup> After CI in the deaf side of children with congenital SSD, lateralization of the sound localization pathway limits recovery of the corresponding pathway of the deaf ear. Therefore, the auditory rehabilitation training strategy for UAD needs to be further explored. Kral et al.<sup>30</sup> suggested that training focussing on the use of the previously deaf ear is required to counteract the consequences of unilateral experience in binaural implantations. Sequential bilateral CI results suggest active inhibition of the first ear that is implanted over that of the second ear that is implanted.<sup>42</sup> Smilsky et al.<sup>43</sup> suggested further discontinuing use of the first CI for a 4-week acclimatization period immediately following activation of the second CI. This was suggested to determine

if revoking use of the first CI affects adaptation to a sequential implant (second CI). These authors found that a period of revoking use of the first CI shortened the time to maximum speech perception in the second CI without long-term consequences on performance of the first CI.<sup>43</sup> For congenital SSD with CI, understanding how to assist in recovery of the deaf ear after CI by eliminating the restrictive effects of the healthy ear on the deaf ear is important. This knowledge may lead to a direction for rehabilitation training in patients with SSD with CI.


### Declaration of conflicting interest

The authors declare that there is no conflict of interest.

### Funding

This work was supported by the Beijing Natural Science Foundation (7162066 and 7162072) and the Natural Science Foundation of Beijing Chaoyang Hospital (CYYPY201917).

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