

Surgical considerations and speech outcomes in infants who undergo cochlear implantation

Experience of the King Abdullah Ear Specialist Center

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ABSTRACT

الأهداف: هذه الدراسة تهدف لدراسة مدى جدوى عمليات زراعة القوقعة للأطفال الرضع.

المنهجية: تمت المراجعة بأثر رجعي للأطفال الرضع الذين خضعوا لعملية زراعة القوقعة في مركز إحالة متخصصة في المملكة العربية السعودية من تاريخ يناير 2011م إلى أكتوبر 2018م. البيانات الديموغرافية، العوامل المرتبطة بالكشف المبكر، وأي صعوبات أو مضاعفات جراحية تم استخراجها من السجلات الطبية. نتائج الزراعة تم تقييمها من قبل أخصائي أمراض النطق والتخاطب.

النتائج: خمسة عشر رضيعاً أجريت لهم عملية زراعة القوقعة خلال فترة الدراسة (9 حالات تلقوا الزراعة في كلتا الأذنين في ذات الوقت، و6 حالات تلقوا الزراعة في أذن واحدة). 5 حالات وجد لديها صعوبة في إيجاد النافذة المستديرة لوضع غرسة القوقعة، وتم توثيق حالة واحدة بإدراج غير كامل للغرسة نتيجة لوجود تحجر في القوقعة نتيجة للإصابة بالتهاب السحايا. لم يتم توثيق أي مضاعفات جراحية بعد العملية. كان متوسط درجة الأداء السمعي هو 7، وكان معدل وضوح الكلام 5.

الخلاصة: هذه أكبر دراسة أتراب وطنية للرضع الذين أجريت لهم عملية زراعة القوقعة، في هذه السلسلة كانت الجراحة آمنة وكانت نتائج الكلام جيدة، من خلال تنفيذ برنامج المسح السمعي لحديثي الولادة في المملكة العربية السعودية من المحتمل أن يزداد عدد الأطفال الرضع الذين تجرى لهم عملية زراعة القوقعة مما يمهد الطريق لمزيد من الأبحاث في هذا المجال.

Objectives: To evaluate the feasibility and outcomes of cochlear implantation (CI) in infancy.

Methods: All infants who underwent CI from January 2011 to October 2018 at a tertiary referral center in the Kingdom of Saudi Arabia were retrospectively reviewed. Demographic data, factors associated with early detection, and any surgical difficulties or postoperative complications were extracted from the

medical records. The outcome of CI was determined by a speech pathologist.

Results: Fifteen patients underwent CI during the study period (9 bilateral and performed simultaneously, 6 unilateral). The round window was difficult to identify in 5 cases. Incomplete electrode insertion because of cochlear ossification secondary to meningitis was documented in one patient. No major postoperative complications were encountered. The average auditory performance score was 7 and the speech intelligibility rating was 5.

Conclusions: This study represents the largest national cohort of pediatric patients undergoing CI in infancy. In this series, the surgery was safe and the speech outcome was good. With implementation of the neonatal screening program in the Kingdom of Saudi Arabia, the number of infants undergoing CI is likely to increase in the near future, paving the way for more research in infant CI.

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Permanent sensorineural hearing loss (SNHL) occurs in approximately 1-3 per 1000 live births.¹ In the Kingdom of Saudi Arabia (KSA), the prevalence of childhood SNHL ranges from 1-4 per 1000 live births.² A recently published study found that the median age of identification of SNHL in Saudi children was around 2.5 years; however, this age had a wide distribution, with 75% of the children being approximately 5 years, meaning that some children were not identified until preschool age.³

Early identification of hearing loss is effective because it can lead to appropriate intervention. When a hearing aid trial fails, cochlear implantation is the next management option. However, the long-term effectiveness of cochlear implantation (CI) depends on a variety of factors, of which age at placement is one of the most important since there is growing evidence to suggest that early CI has a good outcome in terms of language acquisition.⁴

The first year of life is an important period for language development and for establishing auditory input connections with the brain to promote neural development. A child with impaired hearing will have diminished neuroplasticity; therefore, reducing the age of CI would minimize the effect of auditory deprivation in these children.⁵

Surgical intervention during infancy can be associated with difficulties and complications. There are also some differences between infants and older children that need to be considered before surgery, namely, the size of the skull, the thickness of the skin flap, which has a considerable impact on the healing time and protection from device exposure, the facial nerve being more superficial than in older children, the amount of bleeding and the effect this can have on the circulation and stability of the infant under anesthesia and anesthesia-related complications, such as hypoxia and bradycardia.^{6,7} Although the US Food and Drug Administration has approved the use of CI for children as young as 12 months of age, it is important to consider infants with SNHL and their families on a case-by-case basis in order to provide the advantages of early implantation while avoiding any risks or complications associated with that decision.

In this study, we examined the feasibility and outcomes of CI in a national cohort group of infants

in KSA with the aim of establishing an evidence base in terms of surgical difficulties and speech outcomes.

Methods. This retrospective study included all pre-lingual infants aged younger than one year who underwent unilateral or bilateral CI between January 2011 and October 2018 at King Abdullah Ear Specialist Center, Riyadh, which is one of the main tertiary care centers in KSA. All the study participants had Arabic-speaking parents with normal hearing who mainly used Arabic to communicate with their infants. The study protocol was approved by the Institutional Review Board at the College of Medicine Research Center, King Saud University (reference number 18/0831/IRB) and performed in accordance with the tenets of the Declaration of Helsinki.

The following data were collected: age at time of CI, gender, medical and family history, intraoperative findings, and the postoperative course (including any documented complications, either immediate or delayed). In order to be able to predict any surgical difficulties and complications, each CI surgery was divided into the following 3 steps: 1) cortical mastoidectomy and drilling of the bed; 2) identification of the facial nerve and drilling of the facial recess; and 3) identification of the middle ear landmark and visibility of the round window and electrode insertion. The total operating time was also documented.

The results of an age-appropriate speech evaluation by a speech pathologist were also recorded. The speech outcomes were measured using the Meaningful Auditory Integration Scale (MAIS), Meaningful Use of Speech Scale (MUSS), Categories of Auditory Performance (CAP), and Speech Intelligibility Rating (SIR) questionnaires. The MAIS questionnaire includes 10 questions that parents answered regarding their child's spontaneous listening behaviors in everyday situations. The questions assess 3 different areas of auditory skills, namely, using the device and how it affects the child's behavior, alertness to sounds, and understanding of the meaning of sounds. The MUSS questionnaire assesses the child's ability to use speech and language meaningfully. It consists of 10 questions including the use of sound, communication skills, and use of oral language.

All data were recorded in spreadsheet format (Excel for Mac V16.19, 2018; Microsoft Corp., Redmond, WA, USA).

Results. Fifteen infants (10 male, 5 female; mean age, 10 months) underwent CI during the study period. CI was bilateral in 9 patients and unilateral in 6. The

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decision was made to implant on only one side in some of the patients for the following reasons: limited experience of performing CI in infants on the part of the operating surgeon in 2011 and 2012 for patients 1 and 2; family preference in patients 4, 5, and 7; and a history of meningitis with complete loss of the intracochlear fluid signal on one side and partial patency on the other side in patient 6. Patient 6 was the youngest infant in our series (5.5 months of age at the time of surgery) and was considered an urgent candidate for CI to avoid complete cochlear ossification on both sides.

This patient also has seizure disorder secondary to her meningitis. Another patient had Usher syndrome and 1 had an ectopic anus. The remaining 12 patients (80%) were otherwise healthy and well.

The presence of a positive family history was the factor most commonly associated with early intervention; 12 (80%) of the 15 infants had a deaf family member who had already undergone CI.

Review of all operation reports showed that the surgical team consisted of a consultant specialized in CI accompanied by a fellow or resident as an assistant or both. There were 3 consultants during the study period. The surgical approach in all cases was via a small post-auricular incision. The surgical details are summarized in Table 1. All the patients had an uneventful postoperative course and were discharged home on the day one after surgery.

The average duration of postoperative follow-up was about 3 years but ranged from 11 months (for patients

Table 1 - Intraoperative findings of all infants who underwent cochlear implantation.

Patient	Side	Mastoid cavity	Facial recess	Middle ear + visibility of RW	Total operating time
1	Unilateral (right)	Unremarkable	Good access	Normal middle ear, insertion through RW	100 min
2	Unilateral (left) + insertion of a VT in the right ear	Unremarkable	Good access	Left ear: Normal middle ear + insertion through RW.	120 min
3	Bilateral		Good access on right Narrow facial recess on left, combined technique (facial recess + trans-adtus approach+)	Effusion in right ear (a VT was inserted) Normal middle ear, insertion through RW	183 min
4	Unilateral (right)	Unremarkable	Good access	Normal middle ear, insertion through RW	96 min
5	Unilateral (right)	Unremarkable	Good access	Normal middle ear, insertion through RW	77 min
6	Unilateral (right)	Unremarkable	Good access	Difficult to identify the RW and the scala tympani was ossified; limited partial insertion was achieved	150 min
7	Unilateral (right)	Contracted mastoid (low lying dura, anterior displacement of sigmoid sinus)	Good access	Normal middle ear, insertion through RW	180 min
8	Bilateral	Unremarkable	Limited (facial nerve displaced anteriorly)	Normal middle ear, insertion through RW (thick niche)	173 min
9	Bilateral	Unremarkable	Good access	Normal middle ear, insertion through RW	190 min
10	Bilateral	Unremarkable	Good access	Normal middle ear, Deep round window on left side, insertion through RW	205 min
11	Bilateral	Left side filled with granulation tissue	Good access	Left side edematous mucosa with granulation, insertion through RW	240 min
12	Bilateral	Filled with granulation tissue bilaterally	Good access	Edematous mucosa with effusion, insertion through RW	260 min
13	Bilateral	Unremarkable	Good access	Normal middle ear, insertion through RW	175 min
14	Bilateral	Filled with granulation tissue bilaterally	Good access	Edematous mucosa with effusion, insertion through RW	195 min
15	Bilateral	Unremarkable	Good access	Normal middle ear, insertion through RW	180 min

RW - round window, VT - ventilation tube

14 and 15) to approximately 7 years (for patient 1). No patient had any documented complication requiring medical or surgical intervention at any follow-up visit. The performance of each patient on the different speech assessments is summarized in Table 2.

MAIS questionnaire scores. Dependence on the device and its importance. Eleven of the 15 patients were keen to use the device all the time and were able to identify and report any device-related problem. Three of the patients were compliant with wearing the device during waking hours on 92% of occasions. However, given the young age of the patients, no questions were asked about whether or not the device was working.

Ability to discriminate sounds. Fourteen patients responded to their names automatically in a quiet environment, relying on hearing alone, and were able to discriminate different environmental sounds. They could also recognize any new sound in an unfamiliar place on 100% of occasions, as would a child with normal hearing. However, 5 patients had a problem responding to their names in noisy environments, with a response rate of 55%.

Comprehension and understanding. Ten patients had a high comprehension ability and were able to discriminate between speech and environmental sounds on 100% of occasions. Moreover, they were able to discriminate different individual's voices and emotional states by voice alone. Two of the remaining patients could distinguish the voices of family members on

75% of occasions and 2 were able to recognize different meanings on the basis of tone of voice on 50% of occasions.

MUSS questionnaire scores. Use of sounds. Twelve patients used their voice to attract the attention of others and to produce sounds similar to the syllables of the intended word on 100% of occasions.

Communication skills. Ten patients showed appropriate use of pitch, intonation, and loudness on 100% of occasions. The rate of automatic use of expressive language in both familiar and unfamiliar situations was 100%, indicating no difference in speech between these patients and children of the same age with normal hearing. Two of the remaining patients used expressive language at least 50% of the time, and 2 did not show any development in the use of verbal language, which could be attributed to environmental reasons because one had 2 brothers who used sign language and the other did not receive any training from parents at home.

Use of verbal language. Twelve patients used speech to communicate with strangers and their speech was understood easily on 100% of occasions. In the remaining 3 patients, strangers understood half of their speech, and correction and clarification was used 50% of the time.

Discussion. King Abdullah Ear Specialist Center is a large referral facility performing more than 300 CIs per

Table 2 - Age at time of cochlear implantation and performance on speech tests.

Patient	Gender	Factor prompting early detection	Chronologic age, years	HA, years	CAP	SIR	MAIS	MUSS
1	F	+FH	7.11	7	7	4	40	37
2	M	+FH	7	6	7	5	38	40
3	M	+FH	6.4	6	7	5	38	40
4	M	+FH	5.8	4.10	8	5	38	40
5	M	+FH	4	3.4	5	4	38	28
6	F	Attack of meningitis	4	3	0	0	2	0
7	M	+FH	3.5	2.5	6	3	40	34
8	F	+FH	3.2	2.2	7	4	40	37
9	M	Preterm baby: screened for hearing	2.9	1.11	6	3	40	35
10	M	+FH	2.7	1.9	7	5	40	40
11	F	*Maternal suspicion	2.5	1.8	5	3	37	34
12	F	Positive FH	2.5	1.7	7	5	40	32
13	M	+FH	2.1	1.7	4	0	37.3	7
14	M	+FH	1.8	0.916 (11 months)	5	2	38	26
15	M	+FH	1.10	0.916 (11 months)	2	0	30	8

+FH - positive family history of hearing loss requiring cochlear implantation (CI). *mother suspected a hearing issue at the age of 2 months when the baby was not responding to loud noises. CAP - categories of auditory performance, HA - hearing age (age of implantation represents the difference between CA and HA); MAIS - meaningful auditory integration scale, MUSS - meaningful use of speech scale, SIR - speech intelligibility rating

year. This report describes our experience of performing CI in infants, including surgical difficulties and speech outcomes. The first phase of the universal neonatal hearing screening (UNHS) program was started in KSA in 2016.⁸ However, at the time of our study, UNHS was implemented in only a few centers with incomplete coverage for all neonates. Therefore, most cases of congenital hearing loss were missed in infancy and presented at a later age.³ This could explain why our series contained such a limited number of children undergoing CI for profound to severe SNHL under the age of 12 years. In most cases, early identification was attributed to previous experience of SNHL requiring CI in the family; there was a positive family history in 80% of patients.

In the late 1990s, the US Food and Drug Administration decreased the lower age limit for CI from 24 months to 18 months and lowered it further to 12 months in 2000. These changes were the result of extensive research and evidence for the benefit and safety of early implantation when combined with early hearing rehabilitation.⁶ However, there are some reports in the literature of children as young as 4 months undergoing CI,⁹ the youngest patient in our series was approximately 5 months of age.

An important consideration before surgery is the many differences between infants and older children. Jöhr et al published a review on otologic procedures in infants that focused specifically on CI surgery, including all the risk factors to be taken into account during surgical planning, intervention, and perioperative anesthesia. One of the major considerations when performing CI in infants is intraoperative bleeding, which can lead to a significant decrease in the hemoglobin concentration; for example, blood loss of 100 mL in a 5-kg infant is reported to decrease the hemoglobin level from 90 g/L to 70 g/L.¹⁰ When the surgical team is familiar with these factors, CI can be performed safely in this young age group.

The complication rate associated with CI varies in the literature according to the duration of follow-up and the type of study participants. In general, the complications are classified as major or minor, with major complications including events requiring surgical intervention. An important study of the 30-day postoperative outcomes of CI published in *Laryngoscope* in 2017 reported an overall complication rate of 1.55%, with no significant difference in rates between infants and children aged 1-18 years ($p=0.96$).¹¹ Two infants in that study developed a superficial

surgical site infection. Another study that compared the outcomes of CI in infants with those in older children aged 12-18 months found: no difference in the rates of major or minor complications, no difference was observed in total operative time between the younger (127 ± 31.1 minutes) and older (136.9 ± 46.7 minutes) group and no statistically significant difference in the length of hospital stay.¹²

Although our study included a long follow-up period of 3 years on average, there were no major complications. The postoperative course was uneventful and there was no need for readmission for a surgery-related reason.

In general, CI can restore normal hearing in deaf children but cannot restore the hearing experience that has been lost in the period prior to the implant, and the gap in performance between hearing age and chronologic age in infants with CI is potentially an important area of research.¹³ In this study, we investigated the language and speech outcome using 4 different questionnaires, namely, the MAIS, MUSS, CAP, and SIR. In general, increasing the hearing age was associated with improvement in the speech score with some exception, namely, patient 6, whose performance was poor over time because of significant intra-cochlear ossification following meningitis and an inability to achieve good cochlear coverage as a result of limited insertion of the electrode. In a study of children who underwent CI at various ages, Dettman et al¹⁴ found that 151 children who underwent the procedure when aged younger than 12 months had higher cognitive ratings than their older counterparts and that their language outcomes were within the normal range for their hearing peers. Another prospective study that included 9 infants with bilateral profound hearing loss who underwent CI at the age of 12 months had a mean delay in vocabulary acquisition of 6 months at the age of approximately 24 months, indicating that CI at the age of 12 months decreased the expected delays by half.¹⁵

The main limitation of this study is that some of the patient records had missing data, namely the amount of blood loss, which could not be retrieved because of the retrospective nature of the research and the relatively limited number of infants that could be included. When UNHS coverage becomes nationwide, more patients in KSA would be expected to undergo CI in infancy, which will pave the way for further research on CI in this age group in the future.

In summary, this study represents the largest national cohort of pediatric patients undergoing CI in infancy. In our experience, this surgery appears to be safe with a good speech outcome.

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