# Cochlear Implantation in Patients with Keratitis-Ichthyosis-Deafness Syndrome: A Systematic Review

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## Introduction

- Keratitis-Ichthyosis-Deafness (KID) syndrome is a rare congenital disorder identified by triad of vascularizing keratitis, hyperkeratosis, and profound sensorineural hearing loss
- Presence of skin debris and chronic otitis may render conventional hearing aids ineffective, and progressive visual impairment may preclude use of sign language

## Objective

This study aims to assess the unique challenges and outcomes associated with cochlear implantation (CI) in this patient population

## Methods

- Comprehensive search of PubMed, Web of Science, and Embase from database inception to present day (November 2024) was conducted in compliance with PRISMA* guidelines
- Search strategy developed using Medical Subject Headings and other keywords
- Eligible studies met the following inclusion criteria: 1) patients diagnosed with Keratitis-Ichthyosis-Deafness Syndrome; 2) patients with no prior diagnosis of ear-related skin conditions other than Keratitis-Ichthyosis-Deafness Syndrome; 3) case reports, studies, or series; and 4) published in English
- Selected studies examined for patient demographics, lateralization of first CI (and second when applicable), previous use of hearing aids, CI model utilized, and length of follow-up
- A qualitative synthesis of post-operative complications, long-term outcomes, and consistent use of CI was employed for all of the studies
- Studies with one- or both-sided pre-operative hearing threshold measurements were grouped to determine average pre-operative hearing threshold on either side
- When possible, hearing improvement was calculated by subtracting post-operative hearing perception from aided pre-operative hearing perception

***Preferred Reporting Items for Systematic Reviews and Meta-Analyses**

## Results

- 9 articles met the inclusion criteria, comprising a total of 13 patients and 17 ears
- **Table 1** details study characteristics and patient demographics; **Table 2** summarizes pre- and post-operative hearing thresholds
- There was one adult patient (50 years) and twelve pediatric patients; the average age of the pediatric patients at first implantation was 3.3 years, with a range of twelve months to eight years
- Bilateral hearing loss reported in all thirteen patients
- Twelve patients had trialed hearing aids at some point before CI, each with unsatisfactory hearing improvement per patient/caregiver report
- There were five pre-CI response thresholds for left ears, averaging at 109.6 dB (80 dB-123 dB)
- There were four pre-CI response thresholds for right ears, averaging at 107 dB (90 dB-120 dB)
- Four studies reported pre-operative aided hearing perception for a total of six patients, with an average hearing perception of 85.2 dB among these patients (73 dB-90 dB)
- Genetic testing consistent with mutations in GJB2 gene in eleven of the studies (85%)
- When family testing was done (seven studies), all genetic mutations found were de novo

| Identifying Number, First Author | Year | Country | n = | Age at time of first CI | Sex | Left CI? (year or age when available) | Right CI? (year or age) |
|---|---|---|---|---|---|---|---|
| 1, Arndt | 2010 | Germany | 2 | 50 years | Female | Yes (2003) | Yes (1998) |
| | | | | 14 mos. | Female | No | Yes (14 mos.) |
| 2, Choung | 2008 | South Korea | 1 | 4 years | Male | No | Yes (4 years) |
| 3, Smyth | 2013 | United Kingdom | 2 | 3 years | Male | Yes (3 years) | No |
| | | | | 8 years | Female | Yes (8 years) | No |
| 4, Hampton | 1997 | United Kingdom | 1** | See above | See above | See above | See above |
| 5, Gumus | 2017 | Turkey | 2 | 7 years | Male | No | Yes (7 years) |
| | | | | 5 years | Female | No | Yes (5 years) |
| 6, Cushing | 2008 | Canada | 1 | 1 year | Unspecified | No | Yes (1 year) |
| 7, Barker | 2009 | Australia | 3 | 14 mos. | Female | Yes (14 mos.) | Yes (47 mos.) |
| | | | | 28 mos. | Female | Yes (28 mos.) | No |
| | | | | 39 mos. | Female | Yes (62 mos.) | Yes (39 mos.) |
| 8, Markova | 2016 | Russia | 1 | 3 years 6 mos. | Female | Unspecified unilateral (42 mos.) | Unspecified unilateral (42 mos.) |
| 9, Dalamón | 2016 | Argentina | 1 | 18 mos. | Male | Yes (18 mos.) | Yes (18 mos.) |

**Indicates duplicate data from another included study
Mos. = Months

## Results Continued

- Notable complications included postoperative infection (30.8%, n=4) and superficial wound dehiscence (15.4%; n=2)
- One patient required revision surgery with local tissue rearrangement, and one other patient was offered revision due to complications but refused
- Ten ears (58.8%) had very limited or no complications after CI
- The average hearing improvement was 45 dB (28 dB-60 dB)
- Time to post-op testing was 9 months to 1 year

| Study # (see Table 1) | Pre-Op Response Threshold – Left (dB) | Pre-Op Response Threshold – Right (dB) | Pre-Op Aided Auditory Perception (dB) | Post-Op Auditory Perception (dB) | Hearing Improve-ment (dB) |
|---|---|---|---|---|---|
| 2 | NR | NR | 90 | 35 | 55 |
| 3 | 80 | 90 | 83 | 41 | 42 |
| | 123 | 118 | 73 | 45 | 28 |
| 4 | NR | NR | 80 | 50 | 30 |
| 5 | NR | NR | 90 | 35 | 55 |
| | NR | NR | 90 | 30 | 60 |
| 7 | 120 | 120 | NR | NR | NA |
| | 115 | NR | NR | NR | NA |
| | 110 | 100 | NR | NR | NA |
| 9 | NR | NR | NR | 25 | NA |
| Average | 110 | 107 | 84 | 37 | 45 |

**TABLE 1 (Left):** Study characteristics and patient demographics
**TABLE 2 (Above):** Pre- and Post-Operative Hearing **(**NA=Not Applicable, NR=Not Reported)

## Conclusion

- Our review shows CI is a reasonable approach to facilitating communication in KID patients
- In spite of the elevated risk for delayed wound healing and flap necrosis, the post-implantation improvement in hearing threshold is substantial in most patients (improved to an average of 45 dB), which may outweigh the increased risks
- Extensive pre-operative planning of flaps and close post-operative wound site monitoring are essential to CI success in patients with KID syndrome