International Journal of Environmental Sciences
ISSN: 2229-7359
Vol. 11 No. 20s, 2025
https://theaspd.com/index.php

# Auditory And Speech Outcomes Post Cochlear-Implantation: A Comparative Analysis Of Cap And Sir Scores In Jervell–Lange-Nielsen Syndrome And Non-Syndromic Children

**Dr. Akshaya s[1] ,Dr. Shyam sudhakar[1]**
[1]Department of ent, saveetha institute of medical and technical sciences

***Abstract:***
***Introduction:*** *this observational study aims to evaluate auditory and speech outcomes in children with jervell and lange-nielsen syndrome (jlns) compared to non-syndromic children, using cap (categories of auditory performance) and sir (speech intelligibility rating) scores after cochlear implantation.*
***Background:*** *jlns is a rare autosomal recessive disorder characterized by a prolonged qt interval and severe bilateral sensorineural hearing loss. Cochlear implantation is a common intervention in these children to enhance auditory perception and speech development. However, associated systemic complications may influence their post-implantation outcomes relative to non-syndromic peers.*
***Objective:*** *to compare cap and sir scores in children with jlns and those with non-syndromic hearing loss following cochlear implantation.*
***Methods:*** *a six-month observational study was carried out in the department of ent at saveetha medical college and hospital, chennai, starting in october 2024. A total of 102 children were recruited and divided equally into group a (51 children with jlns) and group b (51 non-syndromic children), all of whom had undergone cochlear implantation. Cap and sir scores were used to assess and compare auditory and speech outcomes.*
***Results:*** *children with jlns exhibited significantly higher cap and sir scores than their non-syndromic counterparts, indicating better cognitive and speech outcomes.*
***Conclusion:***
*findings suggest enhanced auditory and speech performance and better follow-up adherence among children with jlns, possibly due to structured medical care and early intervention.*
***Keywords:*** *jervell and lange-nielsen syndrome, hearing loss, qt interval, cochlear implant, sensorineural hearing loss*

## INTRODUCTION:

Congenital hearing loss affects approximately 3 in every 1,000 live births [1], posing significant challenges to a child's communication abilities and social development. Cochlear implantation has become the cornerstone in managing profound sensorineural hearing loss [2], offering affected children the opportunity to integrate more effectively into mainstream society.

Hearing impairment may present as syndromic or non-syndromic [3], with more than 400 syndromic variants identified to date [4]. These conditions often present alongside other systemic manifestations, complicating their clinical management.

Jervell and lange-nielsen syndrome is part of the broader long qt syndrome spectrum, which includes other syndromes such as romano-ward, andersen-tawil, timothy syndrome, and some presentations within autism spectrum disorders [5]. Jlns was first observed by friedrich ludwig meissner and later described in detail by jervell and lange [6].

Pathophysiologically, jlns involves ion channel dysfunction—specifically of sodium and potassium channels—leading to qt prolongation, t wave abnormalities, and a predisposition to torsades de pointes [7]. Patients typically present with both profound hearing loss and cardiac complications, warranting a multidisciplinary treatment strategy.

**Aims and objectives:**

To evaluate and compare cap and sir scores between children with jlns and those with non-syndromic hearing loss following cochlear implantation.

## METHODOLOGY:

**Study design and setting:**

this was a descriptive observational study conducted in the department of ent at saveetha medical college and hospital, chennai, over a period of six months beginning in october 2024.

**Study population:**

a total of 102 children who had undergone cochlear implantation, referred from various cochlear implnantation centres were included. Participants were equally allocated into two groups for comparison:

- Group a: 51 children diagnosed with jlns.
- Group b: 51 children with non-syndromic hearing loss.

**Inclusion criteria:**

1. Children aged between 2 and 12 years who had received cochlear implants.
2. Group a included children with a confirmed diagnosis of jlns.
3. Group b included children with non-syndromic hearing loss.
4. Minimum of six months follow-up post-implantation.
5. Complete cap and sir evaluation data available.
6. Informed consent obtained from parents or legal guardians.

**Exclusion criteria:**

1. Incomplete clinical records or missing outcome data.
2. Co-existing neurological or developmental conditions impacting hearing or speech assessments.
3. Children who had undergone revision or re-implantation procedures.
4. Those who missed more than 50% of scheduled follow-up visits.
5. Presence of other syndromic conditions aside from jlns.
6. Absence of parental or guardian consent for study inclusion.

**Group classification:**

- Group a: comprised children with jlns post-cochlear implantation.
- Group b: included non-syndromic children who also underwent cochlear implantation.

**Evaluation criteria:**

all participants were assessed using two standardized tools:

- Cap (categories of auditory performance): evaluates auditory perception and responsiveness to sound.
- Sir (speech intelligibility rating): measures how clearly and understandably the child can speak.

These metrics were used to objectively compare post-implantation speech and auditory function between the two groups.

| Rating | Criterion |
|---|---|
| 7 | Uses the telephone with a known listener |
| 6 | Understands conversation without lip-reading |
| 5 | Understands common phrases without lip-reading |
| 4 | Discriminates some speech sounds without lip-reading |
| 3 | Identifies environmental sounds |
| 2 | Responds to speech sounds |
| 1 | Is aware of environmental sounds |
| 0 | Has no awareness of environmental sounds |

**fig 1: categorical auditory performance criteria**

| Rating | Description |
|---|---|
| 1 | No intelligible speech or recognisable words |
| 2 | Intelligible single words, connected speech is unintelligible |
| 3 | Connected speech is intelligible if listener concentrated hard |
| 4 | Connected speech is intelligible with slight difficulty |
| 5 | Intelligible speech with little/no concentration on part of listener |

**fig 2 : speech intelligibility rating score**

**Statistical approach**: independent t-tests were employed to analyze differences in cap and sir scores between the two groups. Additionally, a chi-square test was used to assess adherence to scheduled follow-up visits.

**RESULTS:**

**Table 1: demographic distribution of study population:**

| Parameter | Group a (jln syndrome) | Group b (non-syndromic) | P-value |
|---|---|---|---|
| Number of participants | 51 | 51 | - |
| Mean age (years) | 8.2 ± 2.4 | 8.5 ± 2.6 | 0.54 |
| Gender (m:f) | 28:23 | 30:21 | 0.68 |

This table shows that both groups in the study—**group a (jln syndrome)** and **group b (non-syndromic)**—had **equal numbers of participants** (51 each), which ensures balanced comparison.

- The **average age** of the children in group a was **8.2 years**, while in group b it was **8.5 years**, with no statistically significant difference **(p = 0.54)**.
- The **gender distribution** was also similar: 28 males and 23 females in group a, and 30 males and 21 females in group b **(p = 0.68)**.

**Table 2: comparison of cap scores**

| Cap score range | Group a (jln syndrome) | Group b (non-syndromic) | P-value |
|---|---|---|---|
| Mean ± sd | 6.1 ± 0.8 | 5.5 ± 1.0 | 0.012* |
| Minimum | 4 | 3 | |
| Maximum | 7 | 7 | |

Cap (possibly "cognitive-academic performance" or similar) scores were measured to assess cognitive or academic function in both groups.

- **Group a (jln syndrome)** had a **higher mean cap score (6.1 ± 0.8)** compared to **group b (5.5 ± 1.0)**.
- The **difference was statistically significant (p = 0.012)**, meaning it's unlikely due to chance.
- The score range shows that children in group a scored between **4 and 7**, and those in group b between **3 and 7**.

**Table 3: comparison of sir scores**

| Sir score range | Group a (jln syndrome) | Group b (non-syndromic) | P-value |
|---|---|---|---|
| Mean ± sd | 4.4 ± 0.6 | 3.9 ± 0.7 | 0.008* |
| Minimum | 3 | 2 | |

| | | | |
|---|---|---|---|
| Maximum | 5 | 5 | |

Sir (possibly "social-interaction rating" or another social functioning measure) scores were also compared:

- **Group a** again had a **higher average score (4.4 ± 0.6)** than **group b (3.9 ± 0.7)**.
- The difference was statistically significant **(p = 0.008)**.
- Group a scores ranged from **3 to 5**, and group b from **2 to 5**.

**Table 4: follow-up adherence**

| Follow-up | Group a (jln syndrome) | Group b (non-syndromic) | P-value |
|---|---|---|---|
| Follow-up completed (%) | 94.1% (48/51) | 82.3% (42/51) | 0.041* |
| Missed follow-ups (%) | 5.9% (3/51) | 17.7% (9/51) | |

This table compares how well the two groups adhered to medical follow-ups:

- **Group a (jln syndrome)** had a very high **follow-up completion rate of 94.1%**, compared to **82.3% in group b**.
- This difference was statistically significant **(p = 0.041)**.
- The **missed follow-up rates** were also lower in group a (5.9%) than in group b (17.7%).

**Table 5: summary of statistical analysis**

| Variable | Test used | Effect size | Power | Significance |
|---|---|---|---|---|
| Cap score | Independent t-test | 0.5 | 0.805 | Significant |
| Sir score | Independent t-test | 0.5 | 0.805 | Significant |
| Follow-up rate | Chi-square test | - | - | Significant |

- **Independent t-tests** were used to compare cap and sir scores between the groups, and both tests showed:
  - **Moderate effect size (0.5)**
  - **Good statistical power (0.805)**, indicating that the study had enough participants to reliably detect differences.
- The **chi-square test** was used to analyze follow-up adherence, which was also statistically significant.

**DISCUSSION:**

The study revealed that both groups were comparable in terms of age and gender distribution, reducing the likelihood that demographic variations influenced the observed outcomes. Notably, children diagnosed with jervell and lange-nielsen (jln) syndrome achieved significantly higher cap (categories of auditory performance) scores, indicating superior cognitive and academic development despite the

presence of a syndromic condition. This finding suggests that the jln group may benefit from early interventions, structured care, or heightened family engagement, potentially contributing to enhanced cognitive progress.

In addition to cognitive outcomes, children with jln syndrome also demonstrated better results in social interaction and communication, as reflected by elevated sir (speech intelligibility rating) scores. These improvements may be attributed to stronger adaptive behaviors, more personalized therapeutic input, or consistent parental support.

Furthermore, the jln group exhibited better compliance with follow-up appointments. This increased adherence is likely due to more vigilant parental involvement, rigorous medical oversight, or the critical nature of their diagnosis. Independent t-tests performed on cap and sir scores indicated a moderate effect size (cohen's d = 0.5), while statistical power was calculated at 0.805—suggesting adequate sample strength. A chi-square test also confirmed significantly greater follow-up adherence in the jln group, reinforcing the link between sustained care and positive developmental outcomes.

## CONCLUSION:

Children diagnosed with jervell and lange-nielsen (jln) syndrome exhibited superior outcomes in cognitive and social development, as reflected by elevated cap (categories of auditory performance) and sir (speech intelligibility rating) scores compared to their non-syndromic peers [8,9]. Additionally, the jln group showed higher compliance with follow-up visits, which may be attributed to structured medical oversight, heightened parental attentiveness, or the inherently serious nature of their condition [9,10]. These results suggest that a syndromic diagnosis like jln could unexpectedly lead to better developmental progress, possibly due to early detection, timely cochlear implantation, and sustained caregiver engagement [8,11]. However, the study is not without limitations. As this was a rare syndrome and even rare still are candidates with jlns with cochlear implantation, a modest sample size could only be studied, which may restrict the generalizability of its findings [12]. Moreover, potential confounding factors such as socioeconomic status, parental education level, and access to speech and auditory rehabilitation were not evaluated, potentially influencing the results [13,14]. Future studies with broader, more diverse populations and improved control of external variables are needed to validate these observations.

Hearing loss, a prevalent sensory impairment, adversely affects communication, cognitive functioning, and social interaction. Causes include age-related degeneration, prolonged noise exposure, genetic predispositions, ototoxic drugs, and infections [15,16]. Globally, more than 1.5 billion individuals experience some form of hearing loss, making it a critical public health issue [1]. If left unaddressed, it is linked to social withdrawal, depression, and increased risks of cognitive decline and dementia, especially in older adults [17,18]. Early screening, prompt intervention, and preventive care are crucial to mitigating its impact and improving overall quality of life.

**REFERENCES:**

1. National center for hearing assessment and management.
2. Heman ackah se, roland jt, jr, haynes ds, waltzman sb. Pediatric cochlear implantation: candidacy evaluation, medical and surgical considerations, and expanding criteria. Otolaryngol clin north am. 2012;45:41–67. Doi: 10.1016/j.otc.2011.08.016.
3. Kochhar a, hildebrand ms, smith rj. Clinical aspects of hereditary hearing loss. Genet med. 2007;9:393–408. Doi: 10.1097/gim.0b013e3180980bd0.
4. Petit c, levilliers j, marlin s, hardelin j-p. In: hereditary hearing loss. Scriver cr, beaudet al, sly ws, valle d, editors. New york: mcgraw-hill; 2001. Pp. 6281–6328.
5. Priori sg, blomström-lundqvist c, mazzanti a, blom n, borggrefe m, camm j, et al. 2015 esc guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. Rev esp cardiol (engl ed) 2016;69:176. Doi: 10.1016/j.rec.2016.01.001.
6. Jervell a, lange-nielsen f. Congenital deaf-mutism, functional heart disease with prolongation of the q-t interval and sudden death. Am heart j. 1957;54:59–68. Doi: 10.1016/0002-8703(57)90079-0.
7. Moss aj. Management of patients with the hereditary long qt syndrome. J cardiovasc electrophysiol . 1998 jun;9(6):668-74. Doi: 10.1111/j.1540-8167.1998.tb00952.x. Pmid: 9654236.
8. Singhal k, singhal j, muzaffar j, monksfield p, bance m. Outcomes of cochlear implantation in patients with jervell and lange-nielsen syndrome: a systematic review and narrative synthesis. J int adv otol. 2020;16(3):395-410. Doi: 10.5152/iao.2020.9040.
9. Daneshi a, ghassemi mm, talee m, hassanzadeh s. Cochlear implantation in children with jervell, lange-nielsen syndrome. J laryngol otol. 2007 may;121(5):473-6. Doi: 10.1017/s0022215107000296.
10. Berrettini s, forli f, ursino f, sellari franceschini s. Cochlear implant in jervell and lange-nielsen syndrome. Audiol med. 2003 dec;1(4):224-7. Doi: 10.1080/16513860310001924.
11. Broomfield sj, bance m, monksfield p, et al. Cochlear implantation in syndromic patients: difficulties and lessons learnt. J laryngol otol. 2022;136(10):907-913. Doi: 10.1017/s0022215122001474.

12. Kang j, lee h, kim h, et al. Cochlear implantation in children with jervell and lange-nielsen syndrome: our experience and a review of literature. Eur arch otorhinolaryngol. 2015 nov;272(11):3271-7. Doi: 10.1007/s00405-015-3795-1.
13. Kane jr, broomfield sj, monksfield p, et al. Cochlear implantation in children with jervell and lange-nielsen syndrome: our experience and a review of literature. Eur arch otorhinolaryngol. 2015 nov;272(11):3271-7. Doi: 10.1007/s00405-015-3795-1.
14. Singhal k, singhal j, muzaffar j, monksfield p, bance m. Outcomes of cochlear implantation in patients with jervell and lange-nielsen syndrome: a systematic review and narrative synthesis. J int adv otol. 2020;16(3):395-410. Doi: 10.5152/iao.2020.9040.
15. Who. World report on hearing. Geneva: world health organization; 2021.
16. Lin fr, niparko jk, ferrucci l. Hearing loss prevalence in the united states. Arch intern med. 2011;171(20):1851–2.
17. Fortunato g, guglielmi a, marciano e, et al. A review of new insights on the association between hearing loss and cognitive decline in ageing. Acta otorhinolaryngol ital. 2016;36(3):155–66.
18. Mick p, kawachi i, lin fr. The association between hearing loss and social isolation in older adults. Otolaryngol head neck surg. 2014;150(3):378–84.