Appendix 1

Detailed results of articles under review

S.No.	Study	Tests Evaluated	Settings/Setup	Pre-Implant Results/Baseline	Post-Implant Results	Conclusion	Additional findings
1	Sarankumar et al. (2018)	1)Categories of Auditory Performance (CAP) 2)Speech Intelligibility Rate (SIR) 3)Speech Perception in Noise Test 4)Meaningful Auditory Integration Scale (MAIS) 5)Meaningful use of Speech Scale (MUSS) 6) P1 latency of CAEP		CAP and SIR scores were recorded at the time of implantation	Post implantation CAP and SIR scores after 12 months were significantly higher than the CAP and SIR scores at implantation. Further, there was no statistically significant difference in the post 12 months of implant outcomes (CAP, SIR, MAIS, and MUSS) between implanted children with ANSD and implanted children with profound cochlear hearing loss.	CI option is beneficial for children with ANSD in speech and auditory outcomes.	SPIN scores at 0 dB and 10 dB SNR had no significant correlation with the P1 latency for /g/ sound. The P1 latency is strongly correlated with CAP and MAIS's auditory outcomes. Similarly, the P1 latency negatively correlated with SIR and MUSS's speech outcome measures.
2	Lee et al. (2021)	1)Presence/ absence of P1 component of CAEP 2) Latency of P1 component of CAEP 3) Categories of Auditory Performance (CAP)	Participants in three groups: Group I- Early intervention with sufficient experience	-	**CAEP Results** Group I- Typical morphology consisting of robust and reproducible P1 peaks in waveform and P1 latency fell	Despite the early intervention (at an average age of 1 year), there was insufficient to catch up to age-appropriate	Sufficient device experience in DFNB9 may be needed for normal auditory cortical maturation, even in early implantation cases.

		4)Speech Intelligibility Rate (SIR) 5)Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS)	Group II- Early intervention with insufficient experience Group III- Late intervention with adequate experience		within 95% CI of normal P1 latency development. Group II- Had robust and reproducible P1 peak waveforms but markedly delayed P1latencies than those in Group I. Delayed latencies fell outside the 95% CI of normal P1 latency development. Group III Despite sufficient experience of 2 years, late implantation after two years resulted in anomalous or absent P1 component. **Correlation of P1 component and postoperative speech performance** P1 latency had a significant negative correlation with postoperative CAP and SIR scores. Further, the weak negative correlation of P1 latency and postoperative IT-MAIS score was noted.	synchronization of neural transmission. Reduction in P1 latency is related to enhanced auditory performance.	

					Further, a significant inverse correlation between P1 latency and CI usage was noted.		
3	Daneshi et al. (2018)	1)Categories of Auditory Performance (CAP) 2)Speech Intelligibility Rate (SIR)	Subjects were divided into two groups: <u>Group I:</u> Those who underwent CI at an age less than or equal to 24 months <u>Group II:</u> Those who underwent CI at an age greater than 24 months	**<u>CAP Results</u>** The median CAP score was one with an inter-quartile range (IQR) of 0 to 1. <u>Group I:</u> Median CAP score of zero with IQR of 0 to 1. <u>Group II:</u> Median CAP score of one with IQR of 0 to 1. **<u>SIR Results</u>** The median SIR score was	**<u>CAP Results</u>** Significant increase in all patients posts one year of the implant. Median CAP scores post one year of the implant were 4 with IQR of 3 to 4 <u>Group I:</u> Median CAP scores post one year of the implant were 2 with IQR of 2 to 3 <u>Group II:</u> Median CAP scores post one year of the implant were 3 with IQR of 2 to 3. Median score improvement was significantly higher for Group II. <u>Median</u> CAP scores post two years of the implant were 5 with IQR of 5 to 6. <u>Group I:</u>	All children with ANSD derived benefit from cochlear implantation. The improvement in the CAP and SIR scores depended on the age of implantation and the duration of the postoperative follow-up.	CAP and SIR, though, are global measures to study outcomes in implanted children but are not sensitive enough to show subtle changes.

				one with an IQR of 1 to 1. <u>Group I:</u> Median SIS score was 1 with 1 to 1 IQR. <u>Group II:</u> Median SIR score was one with an IQR of 1 to 1.	Median CAP scores post two years of the implant were 3 with IQR of 2 to 3. <u>Group II:</u> Median CAP scores post two years of the implant were 2 with IQR of 1 to 2. **<u>SIR Results</u>** A significant increase in the median SIR scores was noted one year postoperatively. The median SIR scores were 3, with an IQR of 2 to 3. However, no significant difference was noted in the SIR scores of <u>Group I</u> (median score of 3 with IQR 2 and 3) and <u>Group II</u> (median score of 3 with IQR of 2 and 3) one year postoperatively. At the second-year follow-up, the median SIR score improvement was significantly higher for Group I (Median of 2 with IQR 1 and 2) when compared to Group		

					II (Median of one with IQR 1 and 2).		
4	de Carvalho et al. (2016)	1)IT-MAIS: Meaningful Auditory Integration Scale for young children 2) MUSS: questionnaire for assessing oral language 3) GASP: The Glendonald Auditory Screening Procedure for the speech perception review in profoundly deaf children from five years old. 4) Quality of experience with CI rating on a Likert scale of 0 to 10.	Subjects were divided into two groups based on the age of implantation: Group 1: Implanted before the age of 4 years old Group 2: Implanted at age more than four years	Percentage of Correct Answers preoperatively GASP: 0% IT-MAIS: 5.63% MUSS: 17.5%	Percentage of Correct Answers Post Implant GASP: 97% IT-MAIS:37.38% MUSS: 25.63% Significant improvement in GASP scores and IT-MAIS post-CI were noted. However, no significant improvement post-CI in MUSS.	CI is an effective management option for children with ANSD.	No difference in outcomes with CI across the groups differentiated based on the presence or absence of residual hearing.
5	Tokat et al. (2019)	Parents Perspective Questionnaire	The questionnaire comprised 11 subsets and 58 questions in total. Replies to questions were rated as either strongly agree, agree, neither agree nor	Descriptive analysis of PPQ in domains: **<u>The decision for implantation:</u>** During the preoperative period and the first week of	Descriptive analysis of PPQ in domains: **<u>The decision for implantation:</u>** Extremely satisfied when children responded to their	**<u>Children never gave up putting on the device</u>** All never gave up except one with mental retardation who was using the device intermittently for 6 hours a day.	**<u>Information regarding the service of the implant</u>** Comprehensively informed- 80.7%, Thought if the device failed, the centre would overcome the trouble- 92.3%

			disagree, disagree and strongly disagree	surgery, there was extreme stress by 84.6% of parents. **Support** Thought children would not need help when growing- 90% post-implant **Communication** Parents believed the child's pronunciation would improve after CI- 92.3% **Self-Confidence** Parents thought children were dependent on	first-time voice post-implant- 84.6% **Implantation Duration:** 100% of parents reported children using cochlear implants constantly **Effect of Implantation** The belief that the child will have a better future post implant-65.4%, Worried about possible implant failure- 69.2% **Communication** Pronunciation was better than expected after CI- 76.9%, Parents could communicate to a child even when out of sight- 80.8% **Self-Confidence** Parents thought children became independent like peers -76.9%,		

				parents more - 100%, Parents did not allow children to do work by themselves- 65.4% **Social Relationships** Children were introverted and socially isolated- The majority of parents	Parents allowed children to do work by themselves- 84.6% **Well Being** Children became calmer as compared to pre-implant- 84.6%, Children still nervous- 11.5%, Children enjoyed listening to music, watching TV or playing post-implant- 80.1% **Social Relationships** Children were more talkative and active after CI- 92.3%, Had better relationships with their siblings, family, and friends post-implant- 92.3% **Education** The same success rate as other children- 80.8%, Adapted well to the same school as other children- 69.2%		

6	Saki et al. (2021)	1) P1 latency and amplitude 2)Speech Intelligibility rating	Single channel CAEP recording. Stimuli were /m/, /t/, and /g/ of 30 ms. duration and inter stimulus interval was set to 1125ms.	Children with a clear P1 response: ANSD I group, Children with absent or no replicable P1 response: ANSD II group, Children with severe to profound hearing loss implanted without ANSD: Control group	**CAEP results** 1) P1 latency and amplitude: Significant main effects of time (p< 0.001) and group (p<0.001) were observed but not for time*group 2) Control group showed significantly lesser P1 latency and significantly larger P1 amplitude than the ANSD I group in the pre-implant phase (p <0.001). 3) ANSD-I group had lesser P1 latencies and larger P1 peak amplitude than the ANSD-II group after two years of follow-up. 4) In all groups, no significant difference was noted in P1 amplitude and latencies across different stimuli (/m/, /t/, and /g/). **SIR results** 1) Most children in all groups in the pre-implant phase had SIR scores of 1 or 2. However, post-CI, the control group SIR	1) Significant reduction in P1 latency observed post CI across different time points in all the groups. 2)Significant increase in P1 amplitude was observed post CI across different time points in all the groups. 3)Control group speech post-CI was easily understood in everyday contexts; however, the ANSD group, even after CI, had unintelligible speech. 4)Children with ANSD showed different central auditory system development degrees regarding P1 latency and amplitude post-implantation.	Significant negative correlation between P1 latency and SIR (r =0.55, p<0.05). Hence, individuals with lesser P1 latencies had higher SIR scores. P1 CAEP responses were strong predictors of behavioural outcome (SIR).

					scores improved to 4 and 5, whereas the ANSD group's scores were not higher than three even after two years of CI.		
7	Yuksel and Ciprut (2020)	1)Turkish version of the Clinical Assessment of Music Perception (T-CAMP) 2)Spectral Ripple Discrimination 3)Temporal Modulation Transfer Function (TMTF)	**Two groups** Group 1- SNHL group post-implant Group 2- ANSD group post-implant	-	Median engagement of the melody recognition average was significantly higher in the SNHL group with 11.11% than in the ANSD group with 5.56%. No significant difference was found in Spectral ripple threshold (SRT), Modulation depth (MDT) in four modulation frequencies, mean Pitch direction discrimination (PDD), and Timbre recognition scores across the SNHL and ANSD groups.	CI users with ANSD also derive benefits in the music, spectral, and temporal perception domains just like the CI users with SNHL except in the melody recognition abilities. Further, early diagnosis and careful follow-ups may result in improved outcomes. ANSD group underperformed on the Melody recognition test more than the SNHL group, which can be attributed to MRT being a more complex task requiring both short-term and long-term memory.	A statistically significant correlation was found between SRD, TMTF, and music perception in the SNHL group. However, there was no statistically significant correlation between SRD, TMTF, and music perception in the ANSD group.

8	Hu et al. (2022)	1) CAP 2) IT-MAIS 3)MUSS 4) Receptive auditory behaviour in LittlEARs Questionnaire (LEAQ-D) 5)Semantic auditory behaviour in LEAQ (LEAQ-I) 6)SIR 7)MUSS 8)LEAQ-P	Since the CI switched on, parents of children completed questionnaires at 0, 1, 2, 3, 6, 9, 12, and 18 months.	The baseline CAP score of the ANSD group was higher than the typically developing group. The baseline IT-MAIS score of the ANSD group was significantly higher than the typically developing group.	**<u>CAP</u>** Improvement in CAP scores was noted in the ANSD group post-CI switch on. After six months, however, the improvement of the TD group was more than the ANSD group. Post 18 months TD group had significantly better performance than the ANSD group. **<u>IT-MAIS</u>** The auditory perception was significantly lesser in the ANSD group than in the TD group at 2, 6, 12, and 18 months of CI use **<u>Receptive Auditory Behaviour</u>** The improvement made with CI at 3, 9, and 18 months of CI use in the TD group was significantly	Post implant switch-on, after three months, there is improvement seen in the auditory perception; however, the verbal skills improved only after nine months of use. Further, the ANSD group lagged behind the TD group in the developmental trend after CI.	The ANSD being a heterogeneous population, children had more individual variations. Further, the differences between the ANSD and the typically developing children (better performance) increased with time.

					higher than in the ANSD group **<u>Semantic Auditory Behaviour</u>** Improvement was initially higher in the ANSD group than in the TD group but later no significant difference at three months of implant use. ANSD group could benefit from CI based on SIR, MUSS, and LEAQ-P.		