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The use of bone-conduction implants in patients with rare genetic syndromes associated with ear malformations

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INTRODUCTION

Bone-conduction implants have emerged as a viable solution for patients with conductive or mixed hearing loss who are unable to use conventional hearing aids due to anatomical abnormalities. Individuals with rare genetic syndromes associated with ear malformations, often experience significant hearing impairments due to external and middle ear malformations.

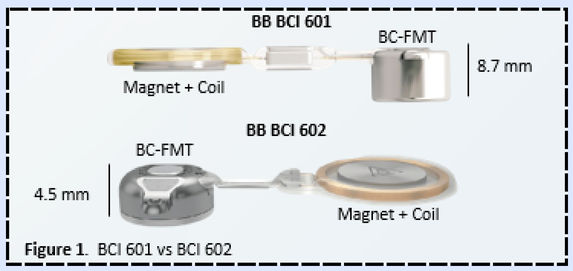

Figure 1. BCI 601 vs BCI 602

Comparison of BCI 602 and 601

- Improved stabilisation in the temporal bone
- Smaller size
- More manageable shape

AIM

To evaluate the effectiveness and safety of bone-conduction implants in patients with genetic syndromes, presenting with ear malformations and conductive or mixed hearing loss.

RESULTS

The results demonstrated significant hearing improvements following implantation. Audiometric assessments showed enhanced speech recognition (fig.2.) and hearing thresholds (fig.3.)

Word Recogniction Score:

- Preoperative: M = 2.2%;
- Activation appointment; M = 79.5%;
- Follow-up appointment: M = 86.3%.

APHAB assesment: Patientsalso reported high satisfaction levels, noting improved daily communication abilities.

The surgical procedures were performed safely, with a low incidence of minor complications.

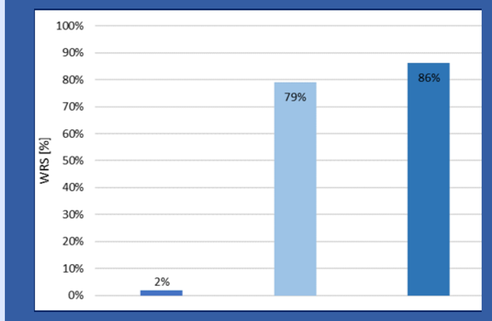

Figure 2. Mean percentage of speech discrimination measured at three time points: before implant insertion, during implant activation and at follow-up.

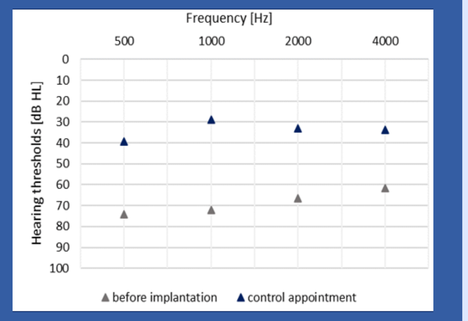

Figure 3. Mean free field hearing thresholds measuredbefore and at the follow-up appintment.

MATERIAL AND METHODS

Study group:

The studyincluded **9 patients** agedbetween 6 and 45 years (M = 17.7) diagnosed with genetic syndromes:

- Treacher Collins Syndrome (n = 5)
- Goldenhar Syndrome (n = 2)
- CHARGE Syndrome (n = 1)
- Klippel-Feil Syndrome (n = 1)

All patients had no benefits or possibility of using conventional hearing aids.

Surgery procedure:

Due to the need to place the implant near the sigmoid sinus and dura mater, precise planning was essential. Preoperative assessments was conducted using MRI or CT scans.

Hearing and questionnaire assesment before implantation and 3 - 6 months after activation:

- Free field tonalaudiometry
- Speech audiometry
- APHAB (Abbreviated Profile of Hearing Aid Benefit)

CONCLUSION

This research highlights the effectiveness of bone-CONDUCTION implants for patients with rare genetic syndromes and ear malformations, addressing an important gap in clinical practice.

Check it out – conference link inside!

Cywka, K., Ratuszniak, A., & Skarżyński, P. H. (2024). Audiological and Subjective Benefits in a Child with Microtia and Atresia After Sequential Bilateral Implantation with Active Bone Conduction Devices: A Case Study. Children, 11(11), 1285.
Cywka, K. B., Skarzynski, P. H., Krol, B., Hatzopoulos, S., & Skarzynski, H. (2022). Evaluation of the Bonebridge BCI 602 active bone conductive implant in adults: efficacy and stability of audiological, surgical, and functional outcomes. European Archives of Oto-Rhino-Laryngology, 279(7), 3525-3534.