

In this issue:

- How Baha helps children with Down syndrome to hear better.
- Reviews of the latest Baha studies for Down syndrome.

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With best wishes, your BAS R&A team; Mark Flynn, Fredrik Breitholtz, Glenn Halvarsson, Agneta Rosengren and Andre Sadeghi.

## Baha® in children with Down syndrome

Hearing loss is common in Down syndrome. Baha may be the best treatment method.

### The genetics of Down syndrome

Down syndrome (also known as trisomy 21 or Down's syndrome), is the most common chromosome defect in humans<sup>(1)</sup>. The collection of congenital abnormalities associated with Down syndrome is caused by three copies of chromosome 21 instead of the usual two. Chromosome 21 is the smallest chromosome with the fewest number of genes and is one of the few trisomies tolerated during the development of the human embryo.

The incidence of Down syndrome is 1/600 births. While an increase in incidence is related to the mother's advanced birth age, not one of the known theories can explain why this chromosome abnormality occurs.

### Middle ear problems in Down syndrome

Glue ear has a particularly high incidence in children with Down syndrome. Glue ear is caused by a mucoid secretion accumulating in the middle ear that stops the ossicles from vibrating freely, thereby reducing hearing levels. Children with Down syndrome tend to have thicker "glue", which is less likely to drain away due to their narrow eustachian tubes, which makes those ears more likely to become infected.

Glue ear is often treated with the insertion of grommets (called pressure equalisation tubes or PETS in the US). Their insertion in the eardrum allows ventilation of the middle ear and prevents the condition from occurring.

In the child with Down syndrome insertion of grommets can be difficult as the ear canal is narrow. There is also a higher incidence of infection around the grommets<sup>(2)</sup> and they are more often extruded, which results in multiple insertions<sup>(3)</sup>. Middle ear infections (Otitis Media) are particularly common in children with Down syndrome. This is due to poor drainage of the middle ear as the eustachian tubes are

often narrower and the fact that children with Down syndrome are more susceptible to infections of all kinds. The treatment of middle ear infections usually involves antibiotics and, potentially, the insertion of grommets.



### Importance of early intervention

In a study of hearing loss in children with Down syndrome<sup>(3)</sup>, the authors stress the importance of frequent examinations with ear cleaning and aggressive treatment with grommets to minimise the risk of developing a hearing loss. In the study, only 2% of the subjects had developed a hearing loss after 5 years. This can be compared to figures ranging between 40-80% reported in the literature<sup>(3)</sup>. However, if these good results will remain in the longer perspective is unclear. As this patient group is already challenged in their development of speech and language understanding, it is of special importance that they are not further compromised if they develop a hearing loss. Several studies<sup>(4,5)</sup> have shown great benefits for children with Down syndrome of using amplification to overcome their hearing loss.

### Common types of hearing loss

**Conductive loss:** The cause of conductive loss is mainly associated with Eustachian tube dysfunction due to alterations in the skull base. However, excessive accumulation of wax can also cause obstruction of the outer ear canal as it is often narrowed in children with Down syndrome. Balkany<sup>(6)</sup> suggests that middle ear problems account for 83% of hearing loss in children with Down syndrome.

**Sensorineural loss:** There is evidence to suggest that this type of loss increases in later childhood<sup>(7)</sup>, with a resulting mixed hearing loss. Both regular and lifelong assessments should be performed to monitor sensorineural hearing loss.

### Treatment of hearing loss

The most common treatment of hearing loss uses the conventional hearing aid. However, Baha provides several advantages over conventional hearing aids:

- Baha bypasses the conductive hearing loss and provides better outcomes if the conductive element is  $>30$  dB<sup>(8)</sup>.
- Baha makes the ear mould unnecessary; Narrow ear canals are a common condition in Down syndrome and Baha entirely eliminates the need for an ear mould.
- Obstruction free ear canal: Because Baha never obstructs the ear canal, recurrent ear discharges dry properly, which further reduces the incidence of recurrent ear infections<sup>(9)</sup>.
- The Baha Softband solution: Many Down syndrome children miss out on the benefits of their conventional hearing aids<sup>(10)</sup> because those aids are removed or lost. The Baha Softband overcomes this problem to provide access to amplification.

Children with Down syndrome are at particular risk for some degree of hearing impairment due to physiological conditions. For a child who needs amplification, Baha is a good alternative if the conductive loss is  $>30$  dB. Several studies<sup>(4,5,10)</sup> have shown good results and high levels of patient satisfaction with Baha in this patient group.

In conclusion: Baha is an excellent treatment for many children with Down syndrome.



## After receiving a Baha, children with Down syndrome experience a significant improvement in their quality of life<sup>(4)</sup>.

### Articles

Articles supporting the use of Baha in Down syndrome are summarized below. An article describing the use of a protective cage during wound healing wraps up the section.

#### The role of bone anchored hearing aids in children with Down syndrome<sup>(4)</sup>.

This study followed 15 children with Down syndrome that were fitted with Baha. Implantation results, skin reactions, and other complications were examined. After receiving their Bahas, the subject's quality of life was assessed using the Glasgow Children's Benefit Inventory (GCBI).

##### Key statements:

- After receiving a Baha, children with Down syndrome experience a significant improvement in their quality of life.
- No significant increase in complication rates was identified in children with Down syndrome.
- A high satisfaction level with Baha was noted by both patients and care givers.
- Bilateral Baha usage should receive additional consideration for this study group.

#### Bone-anchored hearing aid in patients with moderate mental retardation: impact and benefit assessment<sup>(5)</sup>.

This article assessed the impact and subjective benefits of Baha in patients with hearing impairment and a moderate mental retardation. 22 patients (12 with Down syndrome) were evaluated using the GCBI and the Listening inventory for education (LIFE).

##### Key statements:

- Patients with social communication developmental difficulties require proper hearing rehabilitation.
- All study participants responded positively to an increase in overall life quality after their Baha fitting.
- After receiving a Baha, five subjects reported an immediate improvement in daily activities, and three subjects reported a change in their daily working situation. The positive effect of Baha on social communication was the main reason for this improvement in their working situation.
- It's a valuable option to extend the indications for Baha implementation to this special group.

#### UK and Ireland experience of bone anchored hearing aids (BAHA) in individuals with Down syndrome<sup>(10)</sup>.

This study evaluates the incidence of complications for 43 patients with Down syndrome receiving a Baha implant. 24 of the patients were under the age of 16.

##### Key statements:

- Highlights a high level of patient satisfaction with the Baha system.
- Despite the learning disabilities

associated with children with Down syndrome, the post-operative management has not been difficult.

- Strongly supports the use of Baha for children with Down syndrome.

#### A protective cage for the postoperative care of the skin graft and wound of the Baha site in patients with Down syndrome or other learning disabilities<sup>(11)</sup>.

This article describes a protective cage covering the Baha surgical site. In patients with learning disabilities, this covering is beneficial in protecting the post-operative Baha site.

##### Key statements:

- The cage not only allows for aeration, but also enables the clinician to observe the healing process.
- Parents know that the operative site will remain undisturbed.

### Q & A

**Q** I tried measuring the transcranial attenuation in SSD patients according to the principles suggested in the August issue of the Clinical review, but I find that my measurements vary greatly between frequencies. A few Hertz in any direction can dramatically alter the result. Do you have any suggestions on how to measure this over a larger frequency spectrum?

**A** If you measure the transcranial attenuation using narrow band noise instead of pure tones this will eliminate some of the more dramatic differences between individual frequencies. However, the most relevant evaluation of Baha for an SSD patient will be to test it using a headband or Baha Softband for a longer period of time (1-3 weeks).

### REFERENCES

1. Giersch AB, Morton C. Genetic hearing loss associated with chromosome defects. Hereditary hearing loss and its syndromes 2nd edition 2004:462-465.
2. Itano Y, Imamura Y, Hariagi S, Tanak Y. Efficacy of tympanostomy tube insertion for otitis media with effusion in children with Down syndrome. International Journal of Pediatric Otorhinolaryngology 2001 61 (3) 199-205.
3. Shott, S.R. Hearing loss in children with Down syndrome. International Journal of Pediatric Otorhinolaryngology, 2001 6, 199-205.
4. McDermott AL, Williams J, Kuo MJ, Reid AP, Proops DW. The role of bone anchored hearing aids in children with Down syndrome. Int. J. Pediatr. Otorhinolaryngol. 2008 Jun;72(6):751-7.
5. Kunst SJ, Hol MK, Cremers CW, Mylanus EA. Bone-anchored hearing aid in patients with moderate mental retardation: impact and benefit assessment." Otol. Neurotol. 2007 Sept;28(6):793-7.
6. Balkany, T.J., Mischke, R.E., Downs, M.P. & Jafek, B.V. Ossicular abnormalities in Down's syndrome. Otolaryngology: Head and Neck Surgery, 1989 87, 372-384.
7. Davies, B. Auditory disorders. In B. Stratford & P. Gunn (Eds.) New Approaches to Down Syndrome. London: Cassell. 1996 (pp. 100-121).
8. Snik AF, Mylanus EA, Proops DW, Wolfardt J, Hodgetts WA, Somers T, Niparko JK, Wazen JJ, Sterkers O, Cremers CW, Tjellström A. Consensus statements on the Baha system: Where do we stand at present? Annals of Otolaryngology & Laryngology Dec 2005; 114, 12.
9. Hol MK, Snik AF, Mylanus EA, Cremers CW. Long-term results of bone-anchored hearing aid recipients who had previously used air-conduction hearing aids. Arch Otolaryngol Head Neck Surg 2005 Apr; 131(4):321-5.
10. Sheehan PZ, Hans PS. UK and Ireland experience of bone anchored hearing aids (Baha) in individuals with Down syndrome. Int J Pediatr. Otorhinolaryngol. 2006 Jun;70(6):981-6.
11. Watson GJ, Sheehan PZ. A protective cage for the postoperative care of the skin graft and wound of the Baha site in patients with Down syndrome or other learning disabilities. Clinical Otolaryngology 2008 Feb;33(1):73-4.