Facts about Osteogenesis Imperfecta

Hearing

Approximately 50% of adults with OI have a measurable hearing loss, which typically arises in the second or third decade of life. Bone quality and structural abnormalities of the ear bones -- including visible deformities in the ossicles and inner ear -- contribute to the loss. Environmental factors affecting hearing may cause a loss sooner than in the unaffected population. Types of hearing loss are conductive, sensorineural and mixed; severity ranges from mild to profound. In addition, some patients report tinnitus and vertigo. Treatments include hearing aids and/or surgery such as stapedectomy or cochlear implant. There is a slightly elevated risk for facial nerve injury from cochlear implant surgery. Physicians should be alert to dangers of ototoxins in this population.

Children with OI may experience multiple transient middle ear abnormalities caused by a predisposition to Eustachian tube dysfunction. Language or speech delay is generally not related to hearing impairment, but rather to inadequate control of the muscles in the oral cavity and should be evaluated separately.

Recommendations:

- Children with OI should have a formal evaluation by a specialist with pediatric experience before starting school.
- Re-evaluate every 3-5 years or sooner if a change is noted by a parent or a teacher.
- Recurrent audiometric screening is justified given the high frequency of hearing loss.
- Adults with borderline hearing should have regular testing and follow up.
- Counseling regarding dangers of ototoxic medications is essential.
- Children, parents and adults benefit from counseling related to protecting their hearing from loud noises, and inappropriate use of headphones/ear-buds and instruction in the correct use of ear plugs and protective devices.

References:


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