Original Study

International Pediatric Otolaryngology Group (IPOG) Consensus Recommendations: Congenital Cholesteatoma

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Objective: To provide recommendations to otolaryngologists and allied physicians for the comprehensive management of children who present with signs and symptoms of congenital cholesteatoma.

Methods: A two-iterative Delphi method questionnaire was used to establish expert recommendations by the members of the International Pediatric Otolaryngology Group, on the preoperative work-up, the perioperative considerations, and follow-up.

Results: Twenty-two members completed the survey, in 14 tertiary-care center departments representing 5 countries. The main consensual recommendations were: a precise otoscopic description of the quadrants involved, extensive audiological workup (bilateral tonal, vocal audiometry, and BERA), and a CT scan are required. Facial nerve monitoring and a combination of microscope and telescope are recommended for surgical removal. Clinical and audiological follow-up should be pursued yearly for at least 5 years. First MRI follow-up should be done at 18 months postoperatively if the removal violated the matrix. MRI follow-up duration depends on the initial extent of the cholesteatoma.

Conclusion: The goal of preoperative and follow-up consensus from International Pediatric Otolaryngology Group participants is to help manage infants and children with congenital cholesteatoma. The operative techniques may vary, and experienced surgeons must perform these procedures.

Key Words: Children—Congenital cholesteatoma—Delphi method—IPOG—Surgery.

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CONSENSUS OBJECTIVES

To provide recommendations for the comprehensive management of children who present with signs or symptoms of congenital cholesteatoma (CC). CC is a rare disease, thus it is difficult to gather personal experience of its management and the majority of studies have low levels of evidence. This survey of experts in pediatric otology aims to help surgeons by combining state-of-the-art scientific knowledge and invaluable surgical experience.

Congenital cholesteatoma is defined as a white retrotympanic mass associated with a normal tympanic

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Profs FD and NL were the lead authors, Dr Reza Rahbar provided primary consulting and guidance regarding the design and the wording of the consensus recommendations, Dr FS helped write the manuscript. All remaining authors are listed in alphabetical order. The authorship list follows the agreement of the members of the IPOG. All authors have contributed to the conception and the design of the work, drafting and revising the consensus recommendations for important intellectual content, final approval of the version to be published, and agreement to be accountable for all the aspects of the work.

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