OBJECTIVES

Bilateral Congenital Cholesteatoma (BCC) is an extremely rare disease; surgical treatment focuses on preservation of normal anatomy and physiologic integrity of the middle ear to reduce impact on hearing outcomes and on quality of life. A minimally invasive technique that allows a good surgical control in the hidden areas, preserving the mastoid when the cholesteatoma involves only the tympanic cavity is of utmost importance, particularly when both ears are involved.

METHODS

Description of 3 cases of children affected by BCC underwent surgery at the ENT Department of Verona University Hospital in the last 3 years.

RESULTS

Case 1: a 2-year-old male affected by BCC (stage I left side and stage III right side - Potsic’s staging system) who underwent simultaneous transcanal exclusive endoscopic procedure bilaterally in March 2016.

Case 2: 3-year-old female affected by a very large BCC (stage IV bilaterally) who underwent subsequent (2 months) canal wall up mastoidectomy bilaterally in 2015, revision surgery on the right ear for cholesteatoma recurrence in 2015 and surgical bilateral endoscopic check with ossiculoplasty on the right side in 2017.

Case 3: 6-year-old male with a BCC (stage III left side, stage IV right side): endoscopic timpanoplasty on left side in May 2017 and combined approach on the right side in January 2018.

Audiological results: in 3 operated ears we had the closure of the air-bone gap; in 2 ears stable hearing situation. About the last surgery the child had just a modest improvement. Due to a short follow up, at the moment we are not able to say the long term audiological outcome.

CONCLUSION

The introduction of totally transcanal endoscopic surgery has reduced post-operative morbidity which has several benefits in the setting of bilateral disease. The surgical management of BCC should be carefully planned as it is of major importance to preserve anatomy and physiologic integrity of the middle ear to minimize effects of treatment on hearing function and therefore on quality of life.