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## Case Report

# Microtia and cholesteatoma: Implications for the reconstructive surgeon

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## ARTICLE INFO

### Article history:

Received 13 December 2020

Accepted 21 January 2021

Available online 9 February 2021

### Keywords:

Microtia reconstruction

Cholesteatoma

Ear atresia

## ABSTRACT

Infection after reconstructive surgery for microtia is a technical challenge. This can be a sign of cholesteatoma formation by entrapment of epithelium in the middle or outer ear, specifically when the patient does not respond to first choice antibiotic therapy and debridement.

Two patients with microtia presented themselves with severe infections after ear reconstruction. In both cases cholesteatoma was diagnosed as the cause of the infection. After cholesteatoma management an additional surgical procedure was necessary to improve the esthetic outcome. The plastic surgeon should identify possible signs of cholesteatoma after reconstruction of the auricle.

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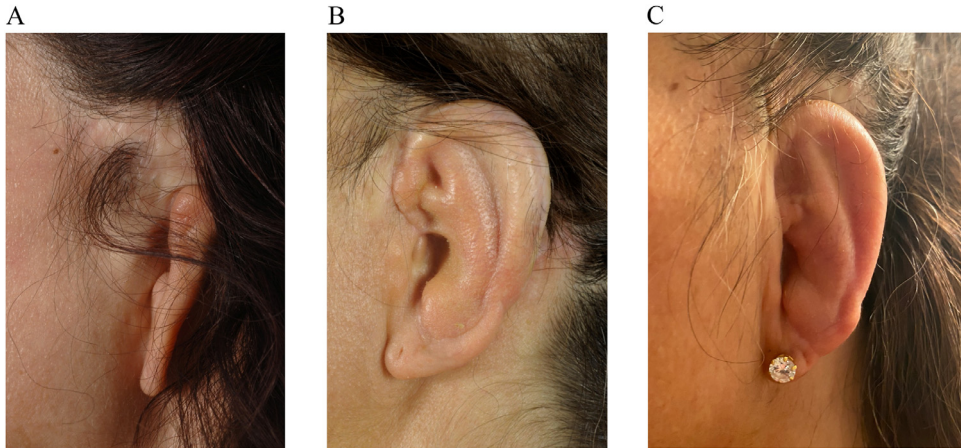
## Introduction

Microtia is a first and second branchial arch anomaly and has a wide range in presentation, ranging from anotia to minimal abnormalities of the auricula.<sup>1</sup>

In many cases, microtia is accompanied by congenital aural atresia (CAA), ranging from mild stenosis to complete absence of the external ear canal with varying degrees of middle and inner ear malformation and accompanying hearing loss.<sup>2</sup>

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**Figure 1.** Photographs of the patient described in Case 1. A: Microtia of the left ear, preoperative, before reconstruction. B: postoperative result after reconstruction of the left ear with rib cartilage. C: End result, 2 years after cholesteatoma surgery and additional elevation procedure of the ear.

Because the outer ear canal is covered by epithelial cells, an entrapment of these cells might result in cholesteatoma formation. This formation out of accumulated keratinizing squamous epithelium can result in destruction of the bone of the outer and middle ear and surroundings.<sup>3</sup>

Reconstructive surgery for the microtia ear is usually performed around 5–12 years depending on the type of reconstruction, wishes of the patient or family and experience of the physician. The most commonly used reconstruction is with autologous rib cartilage. Alternatively, allogenic materials such as Medpor (Stryker, Kamazoo, MI, USA) are being used.<sup>1,4</sup> To establish the best possible functional outcome in terms of hearing, it is imperative for the auricular reconstructive surgeon to work closely together with an audiologist and ENT surgeon<sup>4</sup> to consider the different options for hearing improvement. Thereby, taking into account that interventions to the (entrance of the) ear canal might result in secondary stenosis or formation of cholesteatoma.<sup>5</sup>

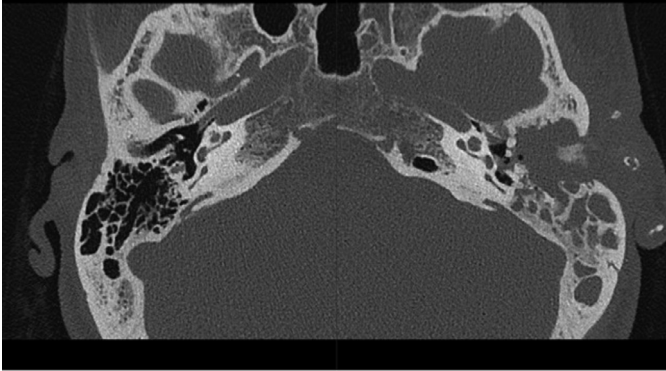
## Case presentation

We present two cases with a history of reconstruction of the microtia ear and presented with a late diagnosis of cholesteatoma, resulting in a salvage operation for the auricular reconstruction.

### Case 1

A 44-year-old female patient with congenital aural atresia and microtia of the left ear had a history of multiple surgical procedures performed elsewhere to reconstruct the external auditory canal when she was 7 years old. During the next years, the external auditory canal was occluded whereby this canal was no longer accessible from the outside. Thereby, she had a fully conductive hearing loss on that side.

A two-stage ear reconstruction (Figure 1a, b) with autologous rib cartilage was performed. A third procedure involved minor adjustments of the concha and pre-auricular sulcus. During surgery, a rudimentary external auditory canal was seen, which was blocked with ear wax and postoperative antibiotics were started. Nine months later some minor adjustments were made on the caudal helix and earlobe. Eight months later the patient presented with an infection of the ear lobe and incision and drainage followed. Six months later the patient underwent a minor correction of the ear lobe. Eight months later a new infection with fluctuation of the concha and above the lobula was seen. Incision and drainage followed in combination with intravenous and subsequently oral antibiotics.



**Figure 2.** CT imaging of the patient described in Case 1. CT imaging of the petrous bone, axial plane. On the right side normal anatomy of the ear. On the left side opacification and destruction of cells of the mastoid and location of outer ear canal, with a partly aerated middle ear.

Several years later the patient presented with recurrence of minor infections of the left ear. CT scan of the petrosal bone showed a status after previous surgery with filling of the area of the external ear canal and mastoid with fluids, without signs of secondary bony destruction (Figure 2). MRI of the ear demonstrated scar tissue, and induration of tissue in the external acoustic canal but no signs of fluid or abscess. To rule out cholesteatoma formation, a dedicated MRI DWI sequence was performed, demonstrating signs of cholesteatoma in the remnant of the ear canal and mastoid. A subtotal petrosectomy was performed by the ENT surgeon to eradicate the cholesteatoma, and closing the ear by filling the petrosal bone with abdominal fat. A fistula through the auricula to the mastoid and ear canal was excised. Postoperative no complications where seen. The postauricular sulcus was lost and 4 months later a reconstruction was performed, with elevation of the ear with a piece of Medpor and a full thickness skin graft. No postoperative complications where seen after 2 years (Figure 1c).

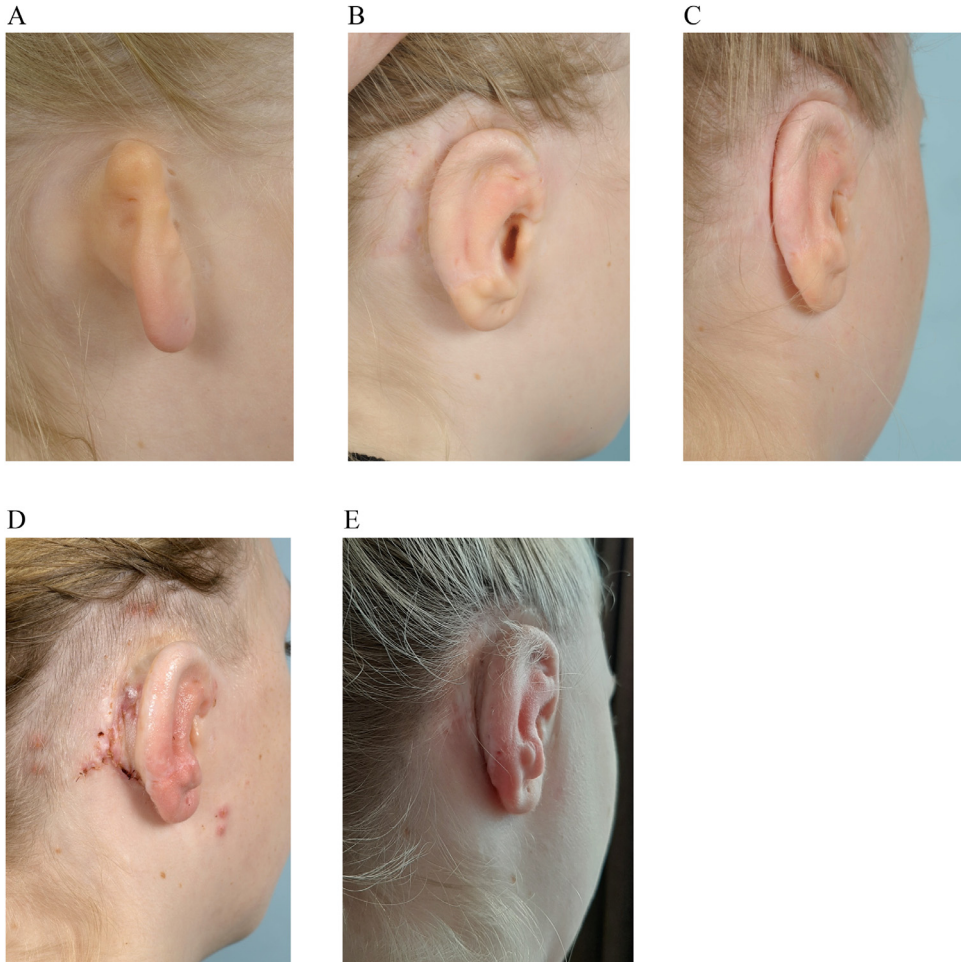
## Case 2

We performed a first stage ear reconstruction on a 9-year-old female with microtia using autologous rib cartilage (Figure 3a, b). The second stage procedure followed with rib cartilage for elevation of the ear. Six months later the patient presented with pain behind the right ear and swelling of the cheek. A CT scan was performed of the ear and mastoid whereby no abnormalities were found. We consulted the ENT surgeon who diagnosed a stenosis of the meatus of the external auditory canal with a very small lumen, and purulent discharge. She was treated with antibiotics. Because of persistent otorrhoea and intermittent fever a CT scan (Figure 4b) and MRI was performed of the ear which showed signs of a cholesteatoma of the ear canal, middle ear and mastoid. A subtotal petrosectomy was performed to eradicate the disease and close the external ear canal to avoid any further complications. Again, the postauricular sulcus was lost (Figure 3c). A third reconstructive procedure was performed to elevate the ear with a block of Medpor (Figure 3d). The end result 2 years after surgery was satisfactory (Figure 3e). No complications were seen.

## Discussion

Microtia is often accompanied with congenital aural atresia.<sup>1</sup> In children with CAA, especially in those cases with an existing, though narrow, outer ear canal, the literature shows that cholesteatoma formation can be a realistic scenario. In previous retrospective chart studies of CAA patients with a congenital aural stenosis (defined as a canal diameter of less than 4 mm), 19–48% of patients were found to develop cholesteatoma during follow-up.<sup>6,7</sup>

For reconstruction of the microtia ear it is of utmost importance to prevent exposure of the implant, advocating aural atresia repair when indicated before reconstruction of the auricula.<sup>4</sup> However,



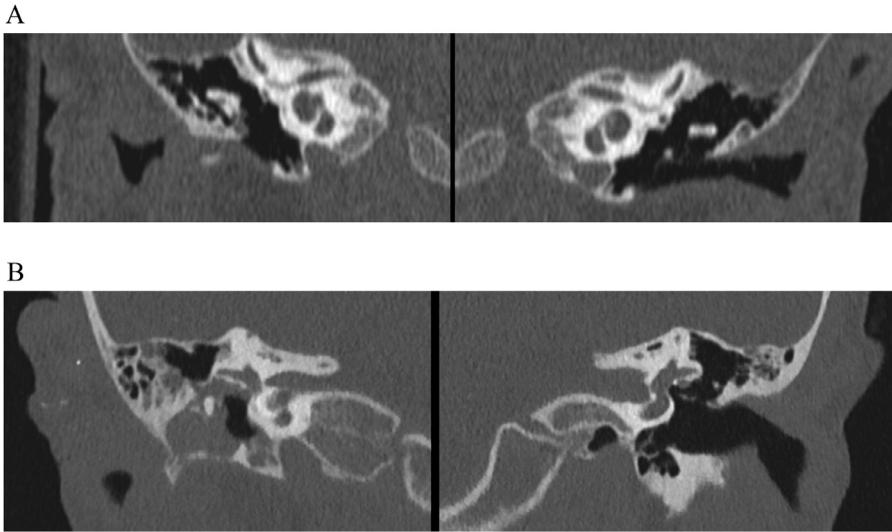
**Figure 3.** Photographs of the patient described in Case 2. A: Microtia of the right ear, concha type. B: Postoperative result after microtia reconstruction of the right ear using rib cartilage. C: Loss of projection of the right ear after cholesteatoma and salvage procedure done by the ENT surgeon. D: 2 weeks postoperative after additional elevation procedure with a piece of Medpor. E: Result 2 years after the last procedure.

scar formation by the atresia repair could limit the tissue needed for reconstruction<sup>4,8</sup> and should be part of the discussion about the timing of surgical interventions.

As described in the literature and from the above cases, it is important for plastic surgeons dealing with ear reconstruction to be aware of primary or secondary cholesteatoma formation by accumulation of epithelia from the ear canal<sup>9</sup> in the area of the auricular reconstruction. Therefore, it is imperative for the plastic surgeon and ENT surgeon to work closely together while planning an ear reconstruction.

A small diameter of the outer ear canal as well as a blocked meatal entrance will put the ear at risk for the accumulation of epithelia in the ear canal.<sup>7</sup> These problems can also be secondary to procedures of the ear canal and/or the pinna, whereby the already smaller external auditory canal is blocked with skin cells.

Both our cases showed a late infection based on the formation of cholesteatoma.



**Figure 4.** CT imaging of the patient described in Case 2. CT imaging of the petrous bone of the second case, axial sequention. A: images 6 months after birth with on the left normal anatomy of the ear. At the right side stenosis of the outer ear canal. B; images at 9 years of age as part of the diagnostic work-up because of persistent otorrhoea. On the right side opacification of the ear canal and middle ear is seen. A subsequent MRI confirmed the diagnosis of cholesteatoma.

If a cholesteatoma is suspected the patient should be referred to the ENT surgeon whereby MR imaging, including DWI sequences, is recommended.<sup>10</sup> Cholesteatoma formation necessitates removal and potentially hinders a favourable outcome of the auricular repair. As demonstrated in both cases, the support of the reconstructed ear was lost due to the salvage surgery by a retroauricular approach needed to remove the cholesteatoma. The result was a non-elevated ear with loss of the postauricular sulcus. We performed an elevation procedure using a piece of Medpor instead of rib cartilage.<sup>11</sup> This had the advantage that we did not have any extra donor-site morbidity. Both reconstructions resulted in a good result and no postoperative infections during 2 years of follow-up.

## Conclusion

Infection after microtia reconstruction can be a sign of cholesteatoma, specifically when the patient does not respond to first choice antibiotic therapy and/or debridement. Close collaboration between plastic surgeon and ENT surgeon is mandatory before and after reconstruction of the auricle.

## Declaration of Competing Interest

The authors declare that there is no conflict of interest.

## Acknowledgements

N/A.

## Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

## Ethical approval

N/A.

## Informed consent

Written informed consent was obtained from all patients.

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