The osseous external auditory canal
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Citation for published version (APA):

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General introduction and outline of thesis
INTRODUCTION

The external auditory canal (EAC) and its diseases have received relatively little attention when regarding the amount of research done within the field of otology. Still, acquired ear canal pathology affects the patient in many ways. Common symptoms include otalgia, discharge, itch and hearing loss. These symptoms can substantially impair daily functioning and therefore resolving them will greatly improve the quality of life.

Overview of surgical techniques addressing the external ear canal and its acquired diseases has been done thoroughly and complete but no outcome measures are given to compare the advocated techniques (1). A remarkable variety of surgical approach/techniques, with seemingly contradictory rationales, exists. Currently personal preference seems to prevail in which technique is used. Therefore evidence regarding the rationale of each used technique is needed in order to define the most effective and safe technique. Surgery will inevitably alter the shape of the canal and its acoustic properties. Unfortunately the effects of these changes, both beneficial and detrimental, are largely unknown.

NORMAL ANATOMY AND (PATHO)PHYSIOLOGY OF THE EAC

The external ear consists of the auricle (pinna) and external auditory canal. The EAC commonly has a length of 2.5 cm (2,3) and its medial ending is defined by the tympanic membrane. In a ‘normal’ anatomical situation the EAC is comprised of a lateral fibrocartilaginous part and a larger medial osseous part. The EAC is known to have a complex anatomy with a large inter and intra-individual variability (4). Although many shapes are shown in anatomy books varying from almost completely cylindrical (5) to highly curved shapes with several isthmus (6) no uniform shape or classification of shapes have been described. Two isthmus have been described in the axial plane, the first in the cartilaginous part at the meatal introitus and the second located in the osseous part of the EAC where the mandibular joint bulge is present. In the coronal plane usually only one isthmus is defined which is located in the osseous external auditory canal (OEAC) and is the result of the bulging of the inferior part of the tympanic bone (4,6). Many authors describe that the OEAC has an ascending lateral part which is followed by a descending part in this plane [figure 1] (7-13). This rounding is due to the shape of the tympanic part of the temporal bone. The shape results in a recess just lateral of the tympanic membrane which has been given several different names. We refer to this region as the pre-tympanic recess (PTR) (14). Other names given are pretymppanic sulcus (7-9), tympanic sulcus (10), pretympanic sinus (11,12) and inferior tympanic recess(13). The PTR can be formed in such a manner that direct otoscopic examination of this region is limited and clinical cleaning of this area is hampered.

The epithelial lining of the EAC has unique characteristics [figure 1]. The skin of the cartilaginous part consists of sebaceous and apocrine glands and hair (15,16). These glands produce ear wax covering the lateral part of the EAC and forming a protective lipid layer with a pH of 6.1(17). It protects the EAC against micro-organisms (18-20). The skin of the OEAC lacks subcutaneous tissue and adheres directly to the perist of the external auditory canal.
Figure 1. Anatomy of the external auditory canal in coronal and axial plane
These two factors make the skin of the bony external auditory canal extremely vulnerable and hard to replace. No ear wax is produced in this part and it is self-cleansing by lateral epithelial migration towards the external environment (21). The features of the epithelial lining mentioned above enable the external auditory canal to fight water-borne pathogens (thus reducing the risk of infection) and to prevent accumulation of debris. In acquired pathology of the EAC these functions are affected and a disbalance in the natural homeostasis will be the result (17,22).

**THE ROLE OF THE EAC**

The function of the EAC is to enable the transfer of sound from the concha to the tympanic membrane and to act as a resonant tube (23). The frequencies that are influenced by this resonance function are those who are important in speech perception. Another possible function of the EAC is the protection of the delicate middle and inner ear structures from the external environment (water resistance, trauma) by enabling the inner ear to be located deeper in the temporal bone (24). Furthermore the aforementioned ear wax production has protective abilities against infections and needs an anatomical location before the ear drum.

The EAC also plays a role in the diagnosis and treatment of middle ear pathology. It allows otoscopic evaluation of the tympanic membrane and can offer a window in the surgical approach of the middle ear.

**EAC ACOUSTICS AND ITS RELEVANCE**

It is known that sound exceeding 200 Hz undergo pressure transformation between the free field and the ear drum (25). This change can be attributed to several factors like the scattering due to the head, the pinna and the resonant effects of the concha bowl and external ear canal (24). Sound transmission in the human ear canal can be regarded as a rigid cylindrical pipe with one open ending and one closed. This meaning the ear drum and middle ear structures can be modelled as a hard-wall termination (26). However, evidence suggests this model to disregard the visco-thermal losses of the ear canal skin and middle ear condition which were seen to affect the ear canal acoustics as well (27). Several manuscripts have reported a change in resonance acoustics when the osseous external auditory canal (OEAC) is modified surgically (28-31). The creation of a wide OEAC (as is the case with a radical cavity) was shown to decrease the resonant frequency substantially and increase the peak amplitude significantly (32). Although these changes have been reported several times in the past, we were not able to find literature concerning the relationships between objective measurements of the resonances and the subjective sound quality of the resulting sound.

**POSITION OF SURGERY IN ACQUIRED PATHOLOGY OF THE EAC**

In 1950, Work was the first to classify various lesions of the external auditory canal (33). He classified the various conditions of the external auditory canal into three main groups: 1. Lesions
arising directly in the external auditory canal. 2. Lesions or external mechanical forces that involve the external auditory canal secondarily by extension, encroachment or contact. 3. Lesions due to developmental disturbances. Tos pointed out that this classification leads to confusion during the following decades and proposed a new classification (1). In his classification a clear distinction is made between acquired versus congenital aetiology. In this thesis, we focus on acquired disease, excluding congenital atresia or stenosis. In the following paragraphs we will briefly outline acquired EAC diseases/conditions and discuss their definition and treatment. The relevance of surgical intervention will be discussed.

Otitis externa (OE)

Definitions of otitis externa are very difficult to find in the international literature. A definition found in the literature is: Otitis externa is a broad term used to describe an inflammatory condition affecting the ear canal, with or without infection. The inflammation is usually generalized throughout the ear canal and can affect the outer ear (34). The symptoms and signs are easily identifiable. Otalgia, itching, aural fullness with or without conductive hearing loss and jaw pain. Signs of such inflammation are tenderness of the tragus and pinna with or without drainage of the affected ear and oedema or erythema of the ear canal and regional lymphadenitis (35). In clinical guidelines OE is categorized based on the duration and onset of the disease into acute and chronic (35,36,37). In general acute is defined as a duration of less than 3 weeks and chronic as a duration longer than 3 months (35,36,37). In the Dutch guidelines persistent otitis externa (POE) is defined as a OE of more than 3 weeks but less than 3 months (37). Acute otitis externa (AOE) can be secondary to acute otitis media: mucopurulent exudate from the middle ear flowing through an acute tympanic membrane perforation can infect the tissues of the ear canal, creating a secondary otitis externa (35). All clinical guidelines and reviews exclude secondary otitis externa (i.e. being the result of middle ear pathology (cholesteatoma), tympanic membrane perforation, benign and malignant tumors of the external ear canal and radiation induced osteoradionecrosis (ORN)) and the more aggressive necrotizing (malignant) otitis externa (35,36,37).

Moreover, in the literature a distinction is made between diffuse and local forms of AOE (38). Diffuse AOE is a common disease, with a reported incidence in the Dutch population of 12.5 per 1000 patient years and with a slight increase with age (39). Common pathogens of diffuse AOE are Pseudomonas aeruginosa and Staphylococcus aureus but many others have been identified (40,41). Treatment of AOE is conservative (adequate cleaning of the ear canal and topical treatment) and no surgical intervention is indicated (42,43). Eighty percent of patients with AOE has a reported clinical resolution within 3 weeks when treated adequately (38).

Although literature concerning the development of COE is inconclusive in general it is postulated that COE develops from AOE. Many etiological factors have been identified/suggested promoting chronicity in OE. Increased humidity, excessive sweating in warmer climate, water quality and exposure when swimming, alkaline soap usage, trauma from self-cleaning, occlusion due to hearing aid wearing, allergy, stress, genetic predisposition, debris
accumulation, dermatological disease and anatomical shape of the ear canal all have been suggested to play a role in COE (22,44-49). In chronic and recurrent otitis externa swelling and hypertrophy of the skin of the ear canal are persisting and topical treatment becomes ineffective. In these cases surgical management is indicated (37). Many observational studies have shown that surgery leads to high success rates in these cases (50-54). It has been postulated that in COE the skin lining has been subject to hypertrophy and swelling making topical treatment ineffective (1). The principle of surgery therefore is to create a wide and patent ear canal (55). In general two different surgical techniques are described in the treatment of COE: the meatoplasty and the canal(o)plasty. These can be stand-alone, staged or combined procedures. A meatoplasty is performed when a narrowing occurs at the lateral part of the outer ear canal or the introitus (56). A canalplasty, with enlarging of the OEAC, is performed when the medial canal is involved (57). The literature is inconclusive regarding what can be seen as a sufficiently wide ear canal and which determinants contribute to a patent ear canal. It has been recommended to regard surgery as a primary choice in COE (37).

One of the most radical changes that can be iatrogenically inflicted on the OEAC is radical surgery of the mastoid. Open cavity mastoidectomy is a commonly performed operation for chronic otitis media with or without cholesteatoma (58). Cavities result in clinician dependence as the self-cleansing mechanism of the osseous external auditory canal (OEAC) is disrupted (59) and often they become troublesome (60). Although a troublesome cavity is strictly speaking not a disease of the external ear, it can be regarded as an iatrogenic COE as many presenting symptoms are the same. It has a multifactorial etiology comprising a deep mastoid tip, high facial ridge, open tubal orifice, remaining air cell tracts, inadequate meatoplasty and an inadequate canalplasty (61). Revision surgery is indicated in these troublesome cavities addressing all the aforementioned factors, if applicable.

The following paragraphs will discuss diseases and conditions that present themselves with similar symptoms as OE. These conditions are to be excluded before one can state that a genuine COE is present (37).

Necrotizing (malignant) otitis externa
This condition is defined as an OE with severe osteitis and abscess formation combined with a progression toward the skull base (osteomyelitis of the skull base) (62). It has been recognized as a distinct clinical entity of OE. *Pseudomonas aeruginosa* is the bacterial procreator. Other important etiological factors include immune suppression and raised blood glucose. These characteristics make it often an unresponsive pseudomonal infection of the external ear canal found in elderly diabetic patients. The presenting symptoms are those of an OE but with a more pronounced severe pain (62). The standard of care is not surgical but prolonged use of antibiotics (primarily parental), tight glycemic control and local debridement. Despite this treatment fatalities are still encountered with mortality rates described up to 10% (63). Facial nerve palsy, positive fungal culture, relapse in disease or radiological signs (tegmen, infratemporal fossa or temporomandibular joint erosion) are signs of more severe disease and additional surgery is
advised (63). This surgery comprises of a full mastoidectomy, lowering inflammatory load, and never is limited to only a canalplasty (1).

Osteoradionecrosis (ORN)

This disease is actually a complication following radiation therapy of head and neck malignancies. Defined as a slowly progressive, aseptic, avascular necrosis of bone tissue (64). The ear canal is more susceptible for ORN as it has an unfavorable vascularization and thin epithelial lining of the bone (65). Due to the avascular necrosis the ear canal is more susceptible to an OE. Therefore patients often present themselves with all symptoms similar to a regular OE (64). If one wants to treat the infection in this condition it is necessary to define whether a localized or diffuse form is present as was described by Ramsden (66). A treatment algorithm was supplied by our group, indicating that surgery is only advised in diffuse ORN (67). In such cases a canalplasty is combined with reconstruction of the EAC or obliteration of the mastoid cavity. Diseased bone is removed until living bone is reached. In cases of non-functional hearing a subtotal petrosectomy is advised with complete removal of the EAC (67).

Exostosis and osteomata

As exostoses and osteomata are sometimes difficult to differentiate in clinical practice we will describe them together. Exostosis are broad based bony elevations located in the external auditory canal. An osteoma is a solitary, pedunculated bony outgrowth, usually occurring along the tympanomastoid suture. Exostosis are regarded as localized hyperplasias or outgrowths of the compact bone which is in contrast to osteomas which are true tumours (1). Regarding the etiology it has been hypothesized that in exostosis a predisposition (intrinsic factor) is present in patients which is ‘excited’ by mechanical, chemical or thermal factors (extrinsic factors) (68,69). An association is present between frequent watersports (mainly surfing) and the presence of exostosis (47). The etiology of osteomas remains unknown (70).

Both conditions are often asymptomatic and are usually an accidental finding during ENT examination (70). Exostosis almost always present themselves bilaterally, and more or less symmetrical between both ears whereas osteoma are almost always unilateral. Both conditions are painless but due their slow growth can give rise to an obstruction of the canal and subsequently result in conductive hearing loss (due to ear wax accumulation) and recurrent or chronic OE. Osteoma, if large enough, could give rise to a pressure feeling and in rare cases temporal pain by a presumable interference with the temporomandibular nerve (71). The accumulation of debris could lead to bony erosion mimicking an external ear canal cholesteatoma (68).

If asymptomatic no treatment is necessary. If symptomatic due to obstruction regular cleaning by a professional is often satisfactory (72). If (recurrent) OE is present conservative treatment often fails and these conditions should be addressed surgically (1). Many observational studies have been conducted in which both conditions are ‘pooled’ and high success rates are reported with varying techniques (72-77). Solitary lesions are often removed with minimally invasive techniques, not always addressing the entire ear canal, and sometimes a chisel is even
sufficient. In more extensive lesions the entire OEAC is addressed. The principle of these surgical interventions is simple. Sufficient removal should be achieved to enable normal physiology to return after healing (1). To which extent removal is necessary to achieve this is up to personal preference as no evidence is present regarding this topic.

External auditory canal cholesteatoma (EACC)

An EACC is a rare benign disease characterized by the erosion of the bony external auditory canal through proliferation of the adjacent squamous tissue (78,79). Secondary to this erosion large plaques of desquamated keratin accumulates. A distinction can be made between an iatrogenic, post traumatic and idiopathic etiology (78). It has an unclear etiology and pathogenesis (80). It can be debated whether nomenclature is adequate and if an EACC is a true ‘cholesteatoma’ of the external ear canal or a separate entity. Symptoms are otorrhea and a chronic, dull pain due to the local invasion of squamous tissue into the bony EAC (81). Conservative treatment has been proposed by removal of keratin and subsequently applying a gauze with salicylate and cortisone ointment (82). This treatment is effective in alleviating symptoms but does not ‘heal’ the condition. True disease resolution is reached in a similar way as that of middle ear cholesteatoma and surgical treatment is necessary for achieving complete removal of diseased epithelium (1). If the disease is solely located in the OAEC a limited canalplasty will often suffice. If extension into the mastoid air cell tracts occurs, reconstruction of the ear canal is needed with or without obliteration or a modified radical cavity (1). The bony erosion is addressed by equalizing it with surrounding bone and the diseased skin should be removed. If more than 50% of the epithelium is involved grafting can be considered (55).

Keratosis obturans

Keratosis obturans has some similarities with the ear canal cholesteatoma, but is now recognized as a distinct clinical and pathological entity. The accumulation of large plaques of desquamated keratin in the ear canal in keratosis obturans doesn't arise secondary to bone erosion (78). Bony ‘remodeling’ is possible but only due to prolonged disease without regular cleaning. In cholesteatoma the skin does lead to primary erosion and not to ‘remodeling’. It also has an unknown etiology. Presenting symptoms are different to EAC cholesteatoma as well. Conductive hearing loss and feeling of pressure are the symptoms if no infection is present (83). Conservative treatment is propagated with regular removal of the keratin plug. Surgery has shown to only alleviate some obstructing symptoms and to enlarge the period between clinical visits but is not effective in truly treating the disease (83). Complete removal of skin and subsequent grafting did show to have good results but has a risk of stenosis and the need for clinical cleaning after such procedures is still necessary (57). Surgery should not be considered as a preferred treatment (1).
Malignant tumors of the external ear canal

Although rare in occurrence both benign and malignant tumors can be found in the external ear canal. The origin of these tumors can be epithelial (squamous cell carcinoma or papilloma, basal cell carcinoma, ceruminous adenocarcinoma) or soft tissue (fibrosarcoma, hemangioma, neurofibroma) or arising from bone and cartilage (chondroma, osteoma, osteosarcoma)(1). If surgery is indicated the oncological principles apply. Radical removal with a considerable margin is propagated (84). Canalplasty alone does not achieve this goal and therefore is deemed insufficient. The treatment of choice is a (sub)total petrosectomy will be performed with or without subsequent radiotherapy (85).

Acquired stenosis of the EAC.

A stenosis is a narrowing of the canal of varying severity and aetiology. Tos divided the acquired stenosis in four aetiological groups: 1. Postinflammatory. 2. Posttraumatic 3. Postoperative and 4. Neoplastic. (1). Stenosis can be located medially or laterally, can be web shaped or diffuse, and can be circumferential or non-circumferential. Slight stenosis are clinically unimportant. If diffusely present most clinicians regard this entity as a narrow ear canal and often don't refer to it as a disease at all. A stenosis can lead to symptoms, as it can interfere with normal physiology of the EAC and can result in a chronic refractory otitis externa (86). Web shaped stenosis can be seen after surgery and inflammation and is commonly regarded as a undesired situation. Although such small stenosis seldom lead to problems when regular clinical cleaning is performed. Conservative treatment will normally suffice but in the more severe cases surgery is the only real problem solving option. In these cases a canalplasty is necessary (1).

Acquired atresia of the EAC.

Defined by Tos as an intraluminal sequelae of either intra- or extraluminal processes of varying aetiology (1), nomenclature regarding this disease is as varied as its aetiologies. Post-traumatic, post inflammatory and iatrogenic atresia are several entities that have close resemblance to one another without any clear distinguishing parameters. A distinction is made within the acquired atresia group between solid and membranous acquired atresia but these can present themselves in all etiological subgroups. As membranous atresia is very rare (1) we will discuss only the more common disease of a solid acquired atresia. This condition is most commonly referred to as Acquired Medial Canal Fibrosis (AMCF). Iatrogenic blunting of the tympanic annulus region could be regarded as the mildest form of this entity. Giving rise to conductive hearing loss and a feeling of fullness surgical solutions have been described for this condition. Some older reports have described modified radical cavity surgery as a solution (87,88). Current surgical principle is the complete removal of the fibrous plug by elevating it from the bony ear canal, annulus fibrosis and the lamina propria of the ear drum (1). The usage of skin grafts and flaps have been propagated and many advise to enlarge the bony ear as well (a complete canalplasty) (73,89-91).
Surgical management of the OAEC has a clear role in treatment of varying diseases of the external ear canal. But not much evidence is currently present regarding the topic. More research therefore is warranted.

AIM AND BRIEF OUTLINE OF THIS THESIS

The main goal of this thesis is to investigate three aspects of the OEAC in the treatment of COE: surgery, shape and (the effects on) sound. Currently insight is lacking regarding which surgical technique/approach is most safe and effective. Furthermore, the knowledge to which extent the shape needs to be altered during surgery is insufficient. Also, the alteration of shape of the OEAC after surgery will affect the resonance function of the OEAC and therefore the perceived sound quality. More data regarding the effects of these alterations on perceived sound quality will improve pre-operative counselling and minimise patient discomfort.

We asked ourselves whether surgery is a safe and effective way of addressing pathology of the OEAC. Furthermore we wanted to know which techniques can be advocated. We reviewed the literature and identified manuscripts regarding surgery of acquired diseases of the OEAC in order to answer these questions (Chapter 2.1). We report on the retrospective analysis of the results of the canalplasty technique used in our centre to evaluate its effectiveness and safety (Chapter 2.2). This technique enables adequate widening of the bony ear canal due to the creation of a pedunculated skin flap (resulting in a maximisation of exposure and maximal preservation of skin and eliminating the need for grafting). As this technique cannot be used in revision radical cavity surgery (due to the extensive alterations of the OEAC making) we propose a novel canalplasty technique in such cases and the results are shown in Chapter 2.3.

Next we will focus on the shape of the OEAC. Which aspects of the shape are relevant in acquired inflammatory disease of the OEAC? To our knowledge there are no standardised and validated classifications regarding the shape of the OEAC. We therefore suggest a novel method of objectively determining the dimensions of the pre-tympanic recess (PTR) using CT scans (Chapter 3.1) and evaluate whether the PTR plays a role in chronic otitis externa (COE) and troublesome radical cavities. (Chapter 3.1 and 3.2)

The third part of the thesis focuses on the perceived sound quality when the shape of the OEAC has been surgically altered. In order to investigate whether surgically altering the shape has a perceivable effect at all a proof of concept study was performed. We describe a way of filtering sound to simulate a real ear condition. Using this technique we evaluated several conditions of normal and altered ear canals (radical cavities) (Chapter 4.1). Finally we used pre and postoperative conditions of our canalplasty and posterior wall reconstruction in radical cavities techniques (described in Chapters 2.2 and 2.3) to evaluate if and how the perceived sound quality is affected (Chapter 4.2).
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