American Research Journal of Otolaryngology
Volume 1, Issue 1, 10 Pages

AMERICAN RESEARCH JOURNALS
An Academic Publishing House

Research Article Open Access

Congenital Cholesteatoma in Children: A Retrospective Review of 21 Cases

Clarós P.[1]*, Ribeiro I.[2], Clarós A.[1]

[1]Clarós Clinic. Barcelona, Spain.
[2]Scholarship Clarós Clinic, Barcelona, Spain
clinica@clinicaclaros.com
Dr. Pedro Clarós: orcid.org/0000-0002-7567-0370
Dr. Andrés Clarós: orcid.org/0000-0001-6084-3470

Introduction

Cholesteatoma is an abnormal growth of squamous epithelium in the skull base, most frequently in the middle ear and mastoid.[1] It was initially described by House, in 1953. [2]Although benign, its progressive growth can cause erosion and destruction of bone and other structures within the temporal bone. Three types are described: congenital, primary acquired and secondary acquired. [3]Diagnostic criteria have been established to distinguish these entities. Patients with congenital cholesteatoma (CC) must have (1) a mass medial to the tympanic membrane, (2) a normal and intact tympanic membrane, and (3) no previous history of ear discharge, perforation, or ear surgery.[4]

Epidemiology

It is very difficult to determine the incidence of congenital cholesteatomas alone, but studies limited to cholesteatoma in children show an approximate incidence of 5 to 15 per 100.000 children[5], with the congenital type accounting for 1 to 5 percent.[6] It's more frequent in boys and the average age of diagnosis is 5 years, although presentation in adults is not uncommon.[7]

The most usually affected site is the anterosuperior quadrant (ASQ), but nearly half of the cases involves ≥2 quadrants.[6,8] The ossicular chain is frequently involved, resulting in conductive hearing loss (CHL), and the mastoid is affected in 23% of cases.[7] Rarely, cholesteatomas can present bilaterally, in 4% of cases, usually in boys.[9]

Table1. *Location of congenital cholesteatoma*

Location	Kojima[10]	Friedberg[11]	Potsic[7]	Lim[3]
ASQ	3, 2%	31%	39, 5%	29, 2%
PSQ	16%	5%	11, 6%	23, 6%
≥2 quadrants	80, 8%	64%	48%	45, 8%
Ossicle Affectation	Unspecified – 42%			
Malleus + incus	7, 9%	-		8, 3%
Incus	9, 5%	7,5%		11, 1%
Incus + stapes superstructure	50, 8%	37,5%		50, 0%
Stapes superstructure	3, 2%	-		-
Malleus + incus + stapes superstructure	9, 5%	-		-
None	19%	55%	58%	38, 9%

ASQ: anterosuperior quadrant; PSQ: posterosuperior quadrant

As for classification, Derlacki and Clemis first described congenital cholesteatomas as into petrous pyramid, mastoid or tympanic.[12]Potsic first[13], and Nelson later[14], described staging systems to help correlate presentation with recurrence rates. (Table 1)

Table2. *CC staging systems*

Potsic Staging System			Nelson Staging System		
Stage	**Description**	**%**	**Type**		**%**
1	Single quadrant with no ossicular or mastoid involvement	40	1	Mesotympanum with no incus or stapes erosion	15
2	Multiple quadrants with no ossicular or mastoid involvement	14	2	Mesotympanum or attic with ossicular erosion but no mastoid extension	59
3	Ossicular involvement but no mastoid involvement	23	3	Mesotympanum with mastoid extension	26
4	Mastoid extension	23			

Etiopathogenesis

Several theories try to explain the etipathogenesis of congenital cholesteatomas, but it is still a matter of debate. The most widely accepted is the epithelial rest theory, initially suggested by Teed in 1936[15], who observed epidermal appendages within the middle ear mucosa. Later, Levenson suggested these occurred during normal development.[16] However, according to this theory, these epithelial rests may fail to resorb by week 33 of gestation[17] and squamous inclusion cysts may arise, leading to a cholesteatoma.

Some authors defend this theory is not enough to explain all cases of congenital cholesteatomas. Eavey suggests a squamous debris of amniotic origin[18], Peron an epithelial metaplasia.[19] Some authors even defend there can be an implantation of epithelial cells from the tympanic membrane (TM) following infection or microperforation.[11]

Mirko published a provocative paper suggesting that the retractions of the eardrum, common in children, may be repeatedly fixated to the malleus or incus and then loosened again. During these processes an inclusion of keratinized squamous epithelium may occur, leading in a few cases to cholesteatoma formation.[20]

Histologically, a cholesteatoma, also called keratoma, is an epidermal inclusion cyst of stratified squamous epithelium that forms saclike accumulation of keratin within the middle ear. We can define 3 components: perimatrix, consisting of granulation tissue overlaying dense fibrous connective tissue; matrix, referring to keratinizing, cytologically bland stratified squamous epithelium; and cystic contents, contained within a luminal area of abundant, laminated and anucleate keratin.[21] CCs have less connective tissue and more granulation in this area than acquired cholesteatomas and have been known to be more aggressive and with poorer prognosis.[22]

Several markers have been used to try to predict invasion pattern. A recent study shows that CK-17, a high molecular weight cytokeratin, can be an immunohistochemical marker of squamous hyperproliferative disorders, with overexpression in aggressive cholesteatomas.Another known marker is Ki-67, which is a cell-proliferation marker that has been widely used both in neoplastic and nonneoplastic disorders, such as cholesteatoma. It is overexpressed in the cholesteatoma epithelium compared to the normal retroauricular and meatal skin and recent studies showed that cholesteatomas with severe ossicular erosion expressed this marker in an “active” form, whereas less aggressive lesions expressed it in an “inactive” form.[23]

Clinical Presentation and Work-up

Congenital cholesteatomas usually present as a whitish mass in the middle ear, behind an intact TM, with a predilection for the anterosuperior quadrant, just above the eustachian tube. It can cause obstruction and middle ear effusion, with associated conductive hearing loss, which can delay the diagnosis.[7,15]Acquired cholesteatomas usually present with chronic ear drainage despite treatment, but can also present as a new onset hearing loss in a previously operated ear, an atical retraction pocket with or without debris or with granulation in the periphery of the TM. Children who had had tympanostomy tubes or multiple episodes of ear infection are more prone to develop acquired cholesteatoma.[24]

A careful otoscopy, with thourough observation of all quadrants is essential for an early diagnosis. Primary care providers must be alerted to this pathology and, when in doubt, refer the child to a specialist. Traditionally, pneumatic otoscopy was used to add to the sensitivity of the observation, since cholesteatomas do not move with the TM.[25] However, it is usually not available in primary care and its use has been declining even in ENT specialists, who prefer otoendoscopy or otomicroscopy..

As the cholesteatoma grows, it can involve the ossicles, resulting in another cause of conductive hearing loss.[26] TM perforation and chronic ear drainage are late findings. Rarely, it can present with a complication, such as labyrinthine fistula, due to bone erosion, facial palsy or symptoms of central nervous system (CNS) complications, sigmoid sinus thrombosis, epidural abscess or meningitis.[27]

Diagnosis is made with clinical examination, palpation under operating microscope, computed tomography (CT), diffusion-weighted magnetic resonance imaging (DW-MRI) and/or surgical exploration. Some authors defend that for small, localized, unilateral congenital cholesteatomas, pre-operative imaging might be dispensable, but it is usually advocated as part of the work-up, since it can provide prove important as an aid to surgery. Audiometry is also an important part of the pre-operative work-up, since some degree of conductive hearing loss is usually present, either due to ossicular chain involvement or middle ear effusion. In the latter, some authors defend that placing a tympanostomy tube prior to removal and ventilating the ear enhances CT value and improves middle ear conditions at the time of surgery.[26]

Several conditions that alter the TM are differential diagnosis, such as tympanosclerosis, that presents as a white plaque of the TM, but moves with it under pneumatic otoscopy; white foreign bodies; exostoses; inclusion cysts of TM (occupy the middle layer of TM and move with it).[26] Clinical history and physical examination are essential.

Without treatment, cholesteatomas keep growing. Depending on location, they can remain asymptomatic for years, or present with a complication or become secondarily infected. The most common agents are *Pseudomonas aeruginosa, Proteus species, Bacteroides and Peptococcus/Peptostreptococcus*.[5] This infection is very difficult to treat, since cholesteatomas have no blood supply and, therefore, systemic antibiotics are of little use. Complications, as mentioned earlier, are due to bone erosion and destruction of intratemporal structures. Conductive hearing loss is the most frequent, but sensorineural hearing loss may also occur.[7] Other cranial nerves may be involved, such as the VII and sigmoid sinus thrombosis, epidural abscess (a surgical emergency) or meningitis are also possible.[28]

Treatment

Surgical removal is the mainstay of treatment. The only absolute contra-indication if the patient's medical condition is so severe that the anesthetic risk and surgical stress overcome the benefits of treatment. In this case, or when patients refuse surgery, medical care is only possible when there is already a MT perforation and regular excision of debris can help control growth and infection. Topical antibiotics and acidification of the middle ear are the medical treatment of choice, although systemic antibiotics may occasionally help.[29] Some authors defend absence of hearing in the contralateral ear might be a relative contraindication for surgery, due to its risks; however, the natural progression of the disease leads to conductive or mixed hearing loss, so intervention by an experienced surgeon may help improve hearing.[30]

The objective of surgical treatment is to remove the disease and infection, if present, making the ear safe for the patient's daily life while trying to preserve or improve hearing.[31]

Small, encapsulated cholesteatomas may be completely excised by simple access to the middle ear, either by intracanal or postauricular access. When needed, ossiculoplasty might be indicated. If removed intact, recurrence

is rare; however, when piecemeal excision was necessary, the risk of recurrence is higher.[26] Otoendoscopes help improve visualization of middle ear recesses, but a study by Hunter et al showed no difference in outcomes, complication rates, recurrence and residual disease.[32]

In more advanced cholesteatomas, with mastoid involvement, mastoidectomy is indicated. Canal wall-up procedure is preferable, whenever possible, since it preserves the anatomy. However, the risk of persistence or recurrence is higher, so most authors defend a second-look 6-12 months later.[33]

Canal wall-down is preferable in more advanced cases or when the patient is unwilling for revision surgery. It is associated with lower persistence or recurrence rates and the association with meatoplasty allows to optimally control the mastoid bowl.[34] Follow-up is for life, with regular cleaning of debris.

Recurrence is related to the extent of disease, ranging from 14% when confined to one quadrant to 67% when mastoid is involved.[7] Stapleton related recurrence with higher initial stage of disease, ossicular erosion, need for removal of ossicles, cholesteatoma abutting or enveloping the incus or stapes or medial to the malleus or incus.[35]

Preservation of the stapes superstructure relates to hearing preservation or improvement and one large series showed that after canal wall-down procedure, hearing remained mostly unchanged, in 55%, improved in 30% and worsened in 15%.[36]

Facial nerve injury, although very rare, is the most dreaded intra-operative complication. Bone erosion can make the nerve dehiscent and therefore, more prone to trauma.[26] Routine nerve monitoring is still controversial, being the skill and anatomy knowledge of the surgeon the most important factors.[37] If injured, decompression and repair, with removal of the injured section and anastomosis may be attempted.

Sensorineural deafness is reported in 1-2% and is more frequent when a labyrinthine fistula is present or when the cholesteatoma lies on the stapes footplate. Dizziness is also described in less than 1%.[38]

Material and Methods

We identified all patients with diagnosed congenital cholesteatoma between the years 1998 and 2014 in our clinic, specialized in ENT and pediatric ENT. We did a retrospective analysis of the following variables: age, sex, symptoms, location, pre and post-operative hearing, type of surgery, imaging, surgery and recurrence. We used IBM SPSS Statistis 23 for statistical treatment of data, using the t-studen test to identify correlations between variables, with a significant p value < 0,05.

Results

We identified 21 children with congenital cholesteatoma, 14 males (66,7%) and 7 females (33,3%). All patients denied previous otologic surgery and history of ear discharge. The mean age was 5,19 years (range 2-9 years). There was a mean time delay of 1,23 years between initial presentation of symptoms and diagnosis (range 3 months to 4 years). Nine patients were asymptomatic (42,9%) only presenting alterations at otoscopy (figure 1). Four (19%) also had associated conductive hearing loss (CHL), whereas 8 patients (38,1%) presented with isolated conductive hearing loss. Cholesteatoma was more frequently located in the antero-superior quadrant (ASQ), in 52,4%, and in 38,1% it was in the postero-superior quadrant (PSQ). In 2 cases, it occupied both quadrants. None of the patients had lesions occupying the antero-inferior quadrant or the postero-inferior. All patients had a pre-operative CT-scan, that showed in 85,7% a small localized lesion (figures 2 and 3), in 2 cases (9,5%) a lesion occupying all tympanic cavity and in 1 case cholesteatoma invading the mastoid. The ossicular chain was involved in 11 cases: in 38,1% the incus was the only ossicle affected, in two patients it involved both the incus and the stapes and in another it involved the incus and the malleus. All patients with ossicular involvement had CHL, with a mean air-bone gap (ABG) of 17 dBs (range 12-30 dBs).

The mean age at the time of the first surgery was 7,07 +/-1,9. All surgeries were performed by the same senior surgeon. Conservative techniques, such as transcanal approach (47,6%) and antro-aticotomy via retro-auricular approach (42,9%) were the preferred methods. Only 2 patients needed canal wall-down mastoidectomy with timpanoplasty. We registered no intra or post-operative complications. Post-op hearing evaluation showed that hearing remained stable in patients with no hearing loss or improved to a mean ABG of 8,8 dB in patients who had CHL.

There were 3 cases of recurrence (14,3%), 2 patients after 4 years and 1 patient after 1 year, in patients who had been submitted to antro-aticotomy. The mean age at 2nd surgery was 9,33 years and all patients had a canal wall-down mastoidectomy. In all cases, hearing has worsened to a mean ABG of 40 dB. The mean time of follow-up was 12,19 years, (range 4-21 years). Table 3 shows the characterization of the population.

Table3. *Characterization of population*

Variables	N (%)
Sex Male Female	 14 (66,7%) 7 (33,3%)
Age (years)	5,19 +/- 1,6
Side Right Left	 14 (66,7%) 7 (33,3%)
Time Delay (symptoms vs diagnosis)	1,23 +/- 0,829
Location ASQ PSQ Both	 11 (52,4%) 8 (38,1%) 2 (9,5%)
Symptoms Otoscopy CHL Both	 9 (42,9%) 8 (38,1%) 4 (19,0%)
Ossicular involvement Incus long process Incus + stapes Incus + malleus None	 8 (38,1%) 2 (9,5%) 1 (4,8%) 10 (47,6%)
Pre-op hearing Normal CHL Mean CHL	 10 (47,6%) 11 (52,4%) 17, 8 dB
CT findings + ++ +++	 18 (85,7%) 2 (9,5%) 1 (4,8%)
Age at surgery (years)	7,07 +/- 1,977

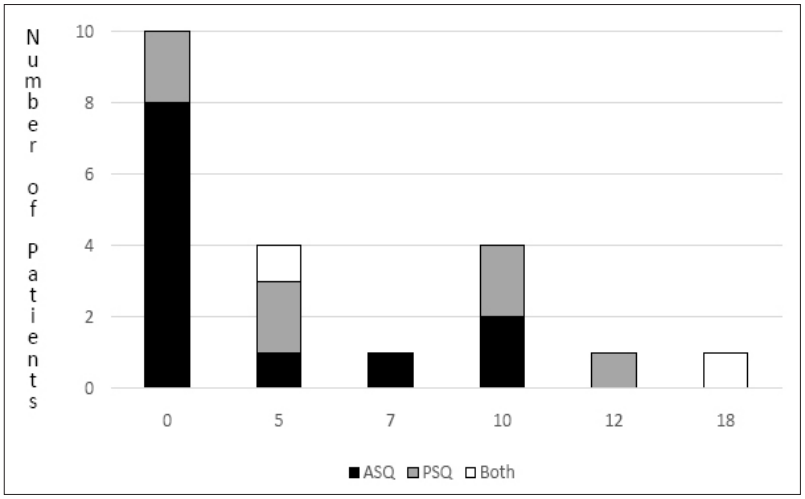
Type of surgery Transcanal Antro-aticotomy Canal wall-down	 10 (47,6%) 9 (42,9%) 2 (9,5%)
Post-op hearing Normal CHL Mean ABG	 10 (47,6%) 11 (52,4%) 8,8 dB
Recurrence	3 (14,3%)
2nd look surgery Canal wall-down	 3 (100%)
Post-op hearing	40 dB
Follow-up (years)	12,19 +/- 4,78

CT findings: + - small, localized cholesteatoma; ++ - cholesteatoma occupying the tympanic cavity; +++ - extension to the mastoid

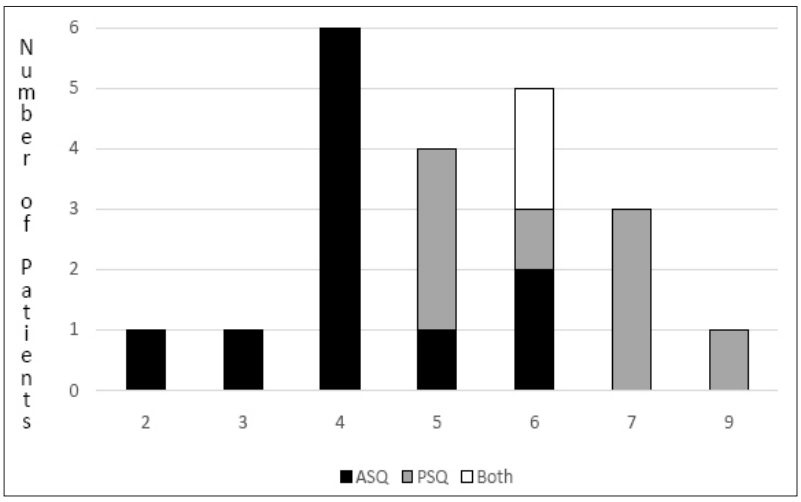
We found that worse pre-operative hearing was related to the presence of ossicular involvement (p 0,000), as well as CT findings (p 0,001), showing that bigger lesions tend to be more symptomatic and easier to identify. We also found a statistical relation between location of the lesion and post-op hearing (p 0,010), with lesions located to the ASQ having better hearing results (graphic 1). Also, as shown in graphic 2, older patients are more prone to have disease localized to PSQ or both, whereas patients aged 4 or less had a localized lesion in the ASQ (p 0,005). This led to more aggressive treatment in the former, with the 2 oldest (aged 7 and 9) needing canal wall-down mastoidectomy (p 0,043) (graphic 3).

The more extensive the lesion, the more aggressive the surgery. We found that location, ossicle involvement and pre-operative hearing, all indicators of the extension of the lesion, correlated to the type of surgery (p 0,019, 0,002, 0,000, respectively).

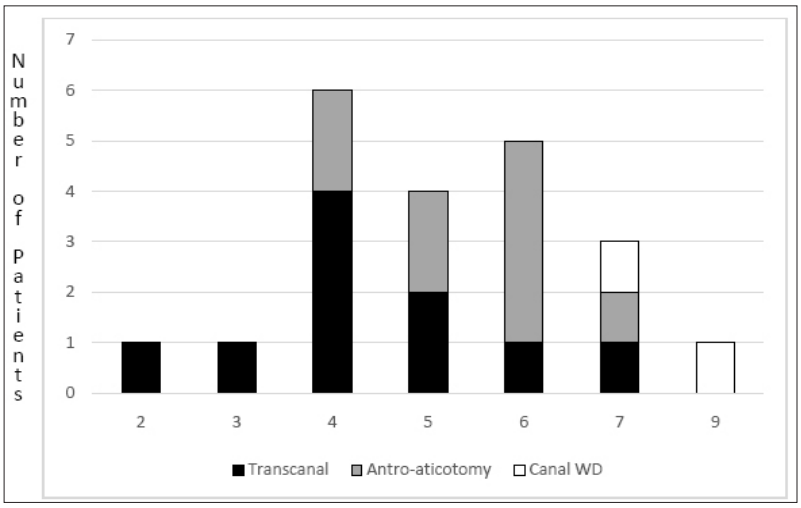
We only found a statistical correlation between recurrence with pre and post- operative hearing (p 0,022 and p 0,021, respectively). No other variables seemed to have an impact in the evolution of the disease (Table 4).



Graphic1. *Relation between location and post-op hearing (dB)*



Graphic2. *Relation between location and age (years)*



Graphic3. *Relation between age and surgical technique*

Table4. *Statistically significant relations found between variables (p<0,05)*

	Age	Pre-op hearing	Ossicular involvement	Location	Symptoms	CT-Scan	Recurrence	Post-op hearing
Pre-op hearing	p 0,786		p 0,000	p 0,061	p 0,001	p 0,017	p 0,022	p 0,067
Age		p 0,984	p 0,563	p 0,005	p 0,000	p 1,219	p 0,093	p 0,016
Type of surgery	p 0,002	p 0,871	p 0,002	p 0,019	p 0,102	p 0,063	p 0,591	p 0,000
Recurrence	p 0,632	p 0,022	p 0,601	p 0,079	p 0,961	p 0,086		p 0,021

Discussion

Our study is in many ways according to the literature. We found a predominance in boys, as previously described[7], and the age of diagnosis also coincides. In most cases, only one quadrant was affected, with a preference for the antero-superior quadrant. This is reported as the primary location of a CC, with posterior extension to the PSQ, with ossicle erosion and finally with mastoid invasion[12] as the natural history of CC. However, our older patients presented mainly with lesion in the PSQ, suggesting that either the origin is different or this may also be a primary site of CC. Lim et al[3] also suggest the latter in their study, since the extent of disease clearly differed

between patients with anteriorly and posteriorly confined lesions and although most early closed-type lesions seemed to arise from the ASQ, some could also originate from the PSQ. All patients with ossicular involvement presented with conductive hearing loss; however, one patient with CHL didn't have ossicular involvement. This can be explained by the presence of effusion and hearing returning to normal after surgery. The incus was the most frequently affected ossicle, and in only 2 cases it was associated with the malleus and stapes, respectively. These patients had the biggest hearing loss, showing that ossicle affectation is the primary and most important cause of conductive hearing loss. Some patients had a significant time delay between the beginning of symptoms and diagnosis. This can be explained because conductive hearing loss is very common in young children, but the most frequent reason for that is effusion. If primary care practitioners are not sensitized to CC, it can remain undetected for a long time, while the patient is being treated for another thing.

All patients were surgically treated by the same surgeon, with no intra or post-operative complications, showing that this is a safe procedure when performed by very experienced hands. Whenever possible, we prefer a closed technique (canal wall-up), either with timpanoplastyvia transcanal approach, in small, localized lesions, or through a retro-auricular antro-aticotomy, with preservation of the anatomy. We believe that with careful excision by the hands of an experienced surgeon, it is possible to achieve good results with low recurrence rates, while preserving or improving hearing and giving children a better quality of life. Results are also better if the cholesteatoma capsule is preserved intact, reducing the chance of recurrence. We only registered 3 cases of recurrence (14,3%), mostly in older patients, with bigger initial lesions and associated CHL. This could be explained because the presence of CHL might be due to ossicular erosion and may relate to a more extended and surgically challenging disease, with higher risk of leaving residual cholesteatoma. This is a very good result when compared to the literature, that puts recurrence between 14 and 67%, depending on the extent of disease[7], which shows that the surgeon skills, experience and work team are key to a good outcome. All were then submitted to a canal wall-down mastoidectomy, with a consequent increase of the ABG to 40dB. In these cases, the patients had all been previously submitted to an antro-aticotomy, so we chose a more aggressive approach, which was successful in controlling the disease. All patients maintained a very long follow-up, with no signs of recurrence.

Conclusion

Congenital cholesteatoma is a rare condition, that can present as a diagnostic challenge since it can be undiagnosed for years. The only effective treatment is surgery, and the best technique is the one that provides better outcome with minimal impact in hearing and the overall quality of life. Small, localized lesions, with no ossicular chain involvement have better prognosis. In experienced hands, we believe that, whenever possible, a closed technique has little complications and good outcome. Hearing was preserved or improved, and the patients ended up with a safe ear. A very long follow-up is mandatory, to keep the ear stable and to be able to intervene in a timely manner, should a recurrence occur.

References

1. Isaacson G. Diagnosis of pediatric cholesteatoma. Pediatrics 2007; 120:603
2. House HP. An apparent primary cholesteatoma. A case report. Laryngoscope 1953; 63(3):712-13
3. Lim HW, Yoon TH, Kang WS. Congenital cholesteatoma: clinical features and growth patterns. Am J Otolaryngol. 2012; 33(5):538-42
4. Derlacki EL, Clemis JD. Congenital cholesteatoma of the middle ear and mastoid. Ann OtolRhinolLaryngol 1965; 74(3): 706-27
5. Bluestone CD, Klein JO. Intratemporal complications and sequelae of otitis media. In: Pediatric otolaryngology, 4th ed, Bluestone CD, Casselbrant ML, Stool SE et al (Eds), Saunders, Philadelphia. 2003 p.687

6. Bennett M, Warren F, Jackson GC, Kaylie D. Congenital cholesteatoma: theories, facts and 53 patients. OtolaryngolClin North Am 2006; 39:1081
7. Potsic WP, Korman SB, Samadi DS, Wetmore RF. Congenital cholesteatoma: 20 years' experience at The Children's Hospital of Philadelphia. Otolaryngol Head Neck Surg 2002; 126:409
8. Koltai PJ, Nelson M, Castellon RJ et al. The natural history of congenital cholesteatoma. Arch Otolaryngol Head Neck Surg 2002; 128:804
9. Kemppainen HO, Puhakka HJ, Laippala PJ, et al. Epidemiology and aetiology of middle ear cholesteatoma. ActaOtolaryngol 1999; 119:568
10. Kojima H, Tanaka Y, Shiwa M, Sakurai Y, Moriyama H. Congenital cholesteatoma: clinical features and surgical results. Am J Otol 2006; 27(5): 299-305
11. Friedberg J. Congenital cholesteatoma. Laryngoscope 1994; 104: 1–23
12. Derlacki EL, Harrison WH, Clemis JD. Congenital cholesteatoma of the middle ear and mastoid. A 2nd report presenting 7 additional cases. Laryngoscope 1968; 78:1050-78
13. Potsic WP, et al. A staging system for congenital cholesteatoma. Arch Otolaryngol Head Neck Surg 2002; 128: 1009–12
14. Nelson M, et al. Congenital cholesteatoma: classification, management, and outcomes. Arch Otolaryngol Head Neck Surg 2002; 128: 810–4
15. Teed RW. Cholesteatoma verum tympani: its relationship to the first epibrachial placode. Arch Otolaryngol 1936; 24: 455–74
16. Levenson MJ, et al. Congenital cholesteatoma in children. Laryngoscope 1988; 98: 949–55
17. Kayhan FT, Mutlu C, Schachern PA, et al. Significance of epidermoid formations in the middle ear in foetuses and children. Arch Otolaryngol Head Neck Surg 1997; 123: 1293
18. Eavey RD, Camacho A, Northrop CC. Chronic ear pathology in a model of neonatal amniotic fluid ear inoculation. Arch Otolaryngol Head Neck Surg 1992;118:1198
19. Peron DL, Schuknecht HF. Congenital cholesteatoma with other anomalies. Arch Otolaryngol 1975; 101: 498
20. MirkoTos. A new pathogenesis of mesotympanic (congenital) cholesteatoma. Laryngoscope 2000; 110: 1890-97
21. Bassiouny M, Badour N, Omran A, et al. Histopathological and immunohistochemical characteristics of acquired cholesteatoma in children and adults. EJENTAS 2012;13:7–12
22. Nevoux, J, Lenoir, M, Roger, G, et al. Childhood cholesteatoma. Eur Ann Otor, Head and Neck Diseases. 2010;127(4):143-50
23. Hamed MA, Nakata S, Shiogama K, et al. Cytokeratin 13, Cytokeratin 17, and Ki-67 expression in human acquired cholesteatoma and their correlation with its destructive capacity. ClinExpOtorhinolaryngol. 2017;10:213–20
24. James AL, Papsin BC. Some considerations in congenital cholesteatoma. Curr Opin Otolaryngol Head Neck Surg. 2013;21:431–9
25. Semple CW, Mahadevan M, Berkowitz RG. Extensive acquired cholesteatoma in children: when the penny drops. Ann OtolRhinolLaryngol. 2005;114:539

26. Sie KC. Cholesteatoma in children. PediatrClin North Am. 1996;43:1245
27. Smith JA, Danner CJ. Complications of chronic otitis media and cholesteatoma. OtolaryngolClin North Am. 2006; 39: 1237
28. Liu JH, Rutter MJ, Choo DI, Willging JP. Congenital cholesteatoma of the middle ear. ClinPediatr (Phila). 2000; 39: 549
29. Ricciardiello F, Cavaliere M, Mesolella M, Iengo M. Notes on the microbiology of cholesteatoma: clinical findings and treatment. ActaOtorhinolaryngol Ital. 2009; 29(4):197–202
30. Myung HY, Byung CK, Hong JP, Tae HY. Middle Ear Surgery in Only Hearing Ears and Postoperative Hearing Rehabilitation. Korean J Audiol. 2014; 18(2): 54–57.
31. Shirazi MA, Muzaffar K, Leonetti JP, Marzo S.Surgical Treatment of Pediatric Cholesteatomas. The Laryngoscope. 2006; 116(9): 1603–1607.
32. Hunter JB, Zuniga MG, Sweeney AD, et al. Pediatric Endoscopic Cholesteatoma Surgery. Otolaryngol Head Neck Surg. 2016; 154(6): 1121-7
33. Dawes PJ, Leaper M. Paediatric small cavity mastoid surgery: second look tympanotomy. Int J Pediatr Otorhinolaryngol. 2004; 68(2): 143-8
34. Roland PS, Meyerhoff WL. Open-cavity tympanomastoidectomy. OtolaryngolClin North Am. 1999; 32(3): 525-46
35. Stapleton AL, Egloff AM, Yellon RF. Congenital cholesteatoma: predictors for residual disease and hearing outcomes. Arch Otolaryngol Head Neck Surf. 2012; 138(3): 280-5
36. Vartiainen E, Vartiainen J. Hearing results of surgery for acquired cholesteatoma. Ear Nose Throat J. 1995; 74: 160
37. Heman-AckahSE, GuptaS, LalwaniAK. Is facial nerve integrity monitoring of value in chronic ear surgery? The Laryngoscope. 2012; 123(1): 2–3
38. Busaba NY. Clinical presentation and management of labyrinthine fistula caused by chronic otitis media. Ann OtolRhinolLaryngol. 1999; 108(5): 435-9

Citation: **Clarós P., Ribeiro I., Clarós A.** "*Congenital Cholesteatoma in Children: A Retrospective Review of 21 Cases". American Research Journal of Otolaryngology. 2019; 1(1): 1-10.*