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[UCI]I804:48009-200000367747

의학석사 학위논문

소아 선천성 진주종의 연령에 따른 임상적 특징과 수술 후 예후 분석

Clinical Characteristics and Surgical Outcomes of Pediatric Congenital Cholesteatoma According to Age

울산대학교 대학원

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지도교수 안 중 호

이 논문을 의학석사 학위 논문으로 제출함

2021 년 2 월

울산대학교 대학원

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Summary

Objective: In this study, we analyzed the clinical characteristics and surgical outcomes of chronic otitis media with congenital cholesteatoma according to age in pediatric patients.

Study Design: From August 1993 to January 2016, we retrospectively reviewed the records of 186 children (136 boys and 50 girls) less than 15 years of age with pathologically confirmed cholesteatoma. We divided the patients into three age groups and analyzed the difference of clinical variables between them. Audiologic outcome and recurrence rate were also analyzed.

Results: There were no significant difference in gender, side distribution, and reoperation rate according to age. However, there were significant differences in chief complaints, cholesteatoma type and operative methods according to age. In addition, the younger patients had fewer mastoid invasion and significantly higher incidence of intact ossicle than older patients. Ossicle erosion, mastoid invasion and open type cholesteatoma caused worse hearing outcome and high recurrence rate. Trans-canal approach showed good surgical outcome due to limited extent of disease. Preservation of posterior canal wall during mastoidectomy slightly elevated recurrence risk but did not affect hearing outcome.

Conclusion: Delayed detection of pediatric cholesteatoma can lead to extensive disease and aggressive operation which can result in worse hearing outcome and elevated recurrence risk. Physicians should familiarize themselves with the clinical characteristics of pediatric cholesteatoma, especially regarding the age distribution of this condition.

Key words: pediatric, cholesteatoma, middle ear, age.

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INTRODUCTION

Cholesteatoma means a benign epithelial lesion of middle ear and temporal bone which can expand and destruct surrounding structures.[1] The name of cholesteatoma was first described by Johannes Muller in 1838. He believed that cholesteatoma was consisted of fat tissue so that he named the lesion with assembly of the terms including “chole” which means cholesterol, “steat” which represents fat, and “oma” which means tumor. [2] First report of cholesteatoma by De Verney written in 1683 also described the lesion as “steatoma”, which means tumor composed of adipose tissue. [3]

Nowadays it is known that cholesteatoma is not consisted of cholesterol nor fat, but squamous debris and desquamated keratin, surrounded by fibrous matrix. Inflammation commonly exists around the lesion. [4] It is found in empty area of mastoid and middle ear, sometimes in external auditory canal. It mostly presents as unilateral lesion, but rarely bilateral lesions are found.[1]

The annual incidence of cholesteatoma is about 3 per 100,000 children and 9.2 per 100,000 adults and there is a predominance in males of approximately 1.4-fold. It is more commonly found in people younger than fifty years old. [5,6]. Cholesteatoma is classified into two types; congenital type and acquired type. Congenital cholesteatoma is known to be present mainly in pediatric patients, and acquired type is found in both pediatric and adult patients. Congenital cholesteatoma is defined as an expanding cystic mass of keratinizing squamous epithelium which is not expanding out of tympanic membrane. It is

thought to be present from birth and these group of patients must have no prior history of otorrhea, perforation of the tympanic membrane, or previous ear surgery to be classified in this category, while history of otitis media effusion does not exclude congenital cholesteatoma [4] There is no external auditory canal lesion or tympanic membrane retraction in this type. [7] It is usually diagnosed during early childhood or infancy. The incidence of congenital cholesteatoma is reported to be 0.12 per 100,000 children. [8] Prevalence is highest among Caucasians, followed by Africans, and very low in Asians. [9] The lesion can be found in four areas: middle ear (middle ear cavity and mastoid), petrous apex, pontocerebellar angle, and jugular foramen [10]

On the other hand, patients with acquired cholesteatoma can show signs or symptoms such as otorrhea, hearing loss, tympanic membrane retraction or perforation, destruction of the adjacent structures, and image findings such as soft tissue masses, bony erosion of the middle ear and mastoid appear. [1]

Congenital cholesteatoma can be subdivided into open type and closed type. Closed type congenital cholesteatoma is an epithelial cyst and keratin inside the cyst is not exposed out, while open type appears as flat keratinizing epithelium [11], [12]

The pathogenesis of congenital cholesteatoma remains unclear, but the most widely accepted theory is the epidermoid formation theory, which suggested that fetal epidermoid formations persist and eventually expand to produce the pathogenic lesion [13,14]. Continuous irritation of this fetal epidermoid which can be found in antero-superior

quadrant of tympanic membrane induces formation of congenital cholesteatoma, while it is generally absorbed in around thirty third gestational week. This theory can explain antero-superior quadrant congenital cholesteatoma, but another needs of another theory has been suggested because congenital cholesteatoma is also found in the other quadrants of middle ear, although it is mostly located in antero-superior quadrant. [4] Other interpretations include the amniotic fluid contamination theory, which suggested that epidermal cells in the amniotic fluid are introduced through the Eustachian tube into the middle ear, where they lodge and proliferate [15]. This theory is supported by amniotic fluid cellular content consisted of keratinized, non-nucleated squamous epithelial cells and lanugo hair found in ears of fetus and neonates. In addition, it has been suggested that ectodermal tissue from the external acoustic meatus can migrate into the middle ear cavity due to failure of the inhibitory function of the tympanic ring. Injury of tympanic membrane can cause invagination around neck of malleus, and tympanic membrane adherent with malleus or incus can leave keratinized epithelium inside the middle ear which can be changed into cholesteatoma at last [16]. Cholesteatoma can arise in any quadrants of middle ear according to this theory.

Jennings stated that genes which determine morphology of ear can be risk factors of both congenital and acquired cholesteatoma, which can be indirectly proved by evidence from syndromic patients including Turner Syndrome, Treacher Collins syndrome, Down Syndrome, and Focal Dermal Hypoplasia in which frequent cholesteatoma has been

reported.[17] Patients with cleft palate also have been reported to have high incidence of cholesteatoma development, with rate of nearly 6% among the population. Patients with cleft palate face have even worse prognosis to develop cholesteatoma, with 100-200 times greater likelihood than patients with cleft palate.[18] Relationship of allergic rhinitis and cholesteatoma has been also reported, with significantly lower disease free rate of cholesteatoma in allergic rhinitis. [19]

In diagnosis of congenital cholesteatoma, CT does not always prove its ability to discriminate congenital cholesteatoma from other diagnosis such as granulation tissue, cholesterol granuloma and other soft tissue lesions thus showing lower specificity than MRI, but it is a powerful tool of obtaining information of surrounding bony structures. [20] Nowadays diffusion weighted image MRI (DWI-MRI) has been developed enough to differentiate keratin debris collection from other lesions with high specificity, in addition to its ability to identify semicircular canal erosion and invasion or abscess forming in ear structures, which make DWI-MRI a powerful tool of cholesteatoma diagnosis. [21] However definitive diagnosis of cholesteatoma has been made by histopathological tissue confirmation postoperatively.

Cholesteatoma is not responsive to antibiotics and remains in middle ear for several years to grow large enough to destruct middle ear structures. It is estimated that diameter of the mass in congenital cholesteatoma patients grows by 1mm per year in average [22]. It silently grows during childhood and eventually causes conductive hearing loss when it

invades ossicles if not incidentally identified in routine otologic examination.

For these reasons, the cholesteatoma should be removed by surgical management, and the primary goal of cholesteatoma surgery is to completely eradicate the disease with the secondary goal of improving hearing and preventing residual and/or recurrent disease. Compared to adult disease, the management of pediatric cholesteatoma is more challenging. Children have higher rates of residual or recurrent cholesteatoma compared to adults, possibly due to anatomic and physiologic difference. [23-25]. Immature anatomy and dysfunction of the eustachian tube (ET) result in more frequent infections and retraction pockets. In addition, well-pneumatized mastoids in children allow for more extensive disease compared to the more sclerotic mastoid bones in adults. Moreover, ET function improves with age, and children between 3 and 6 years of age have a poorer ET function than those aged 7 to 12 years. [26,27]. Despite of these difference of clinical manifestation and physical background according to age, studies investigating prognosis and treatment outcome of congenital cholesteatoma have mainly focused on extent of disease itself than changing physical background according to age.

There is no clear consensus regarding the surgical management of cholesteatoma in children. The three most popular surgical techniques for the treatment of pediatric cholesteatoma are trans-canal tympanoplasty, intact canal wall mastoidectomy (ICW), and open cavity mastoidectomy (OC). While each present both advantages and disadvantages, surgeons adopt a specific technique based on the location and extent of cholesteatoma

before or during surgery, as well as his or her preference and experience. In our current study, we investigated the clinical characteristics of pediatric congenital cholesteatoma and analyzed differences in surgical findings and outcomes for this disorder according to age.

PATIENTS AND METHODS

Patient Selection and Division Criteria

With the approval of the institutional review board, we performed a retrospective analysis of medical and operative records of pediatric patients who were diagnosed with cholesteatoma and who received surgical exploration at the otolaryngology department at the Asan Medical Center, Seoul, Korea. From the patients identified between August 1993 and January 2016, we selected 186 children (136 boys and 50 girls) who were followed for more than 2 years. The mean follow-up period was 47.0 ± 26.8 months. Congenital cholesteatoma was diagnosed based on the criteria of Levenson et al[28], which includes the presence of a whitish middle ear mass behind an intact tympanic membrane, absence of tympanic membrane perforation, and no previous otologic procedures, such as ventilation tube insertion. Presence of effusion in middle ear was not included in exclusion criteria.

Demographic data including age, gender, side of lesion, chief complaints, follow up period, presence of recurrence were reviewed. Patients were divided into three age group; patients less than or equal to three years old, patients whose age were over three years old and less than or equal to seven years old, and patients with age over seven years old for analysis of trend of disease characteristics. Cut-off values of age of each group was set to secure at least 30 number of subjects for each group for their statistical significance and normality of the groups. In addition, patients over 3 years-old showed much better

compliance than patients under 3 years-old so that we could get more detailed clinical information from them by communicating and giving instructions during pure tone audiometry. And it is known that patients under 7 years-old who are preschool aged patients are known to have more immature eustachian tube function, which we referred to determining the cut-off value of latter two groups.[26,27]

Diagnosis, Treatment Method of Cholesteatoma and Data Collection

All patients were examined with tympanic endoscopy in outpatient clinic. Location of the main mass was evaluated, and extraordinary findings in endoscopic findings were reviewed. Preoperative and postoperative hearing thresholds were measured by pure tone audiometry. Pure tone audiometry was possible in 105 patients and 4 frequency average (0.5, 1, 2, and 3kHz) was calculated for each patient. The mean terms of period of follow up pure tone audiometry from operation date was 33.2 months (1.6-168.5). Hearing levels of patients who were unable to get pure tone audiometry test because of lack of compliance underwent auditory brainstem response test for evaluation of auditory function, although it was not analyzed in this study. Impedance audiometry was done for all patients for evaluation of sound conduction status, volume, and pressure in middle ear.

Disease extent was evaluated by non-contrast temporal bone CT scan which is a standard modality of imaging in cholesteatoma; axial and coronal image of both ears were taken and reconstructed into 0.6 mm slice thickness. Typical finding of cholesteatoma of

CT scan was defined as soft tissue density which appears to be mass like lesion which has clear margin or expansile characteristics. Pneumatization of air cell, extent of soft tissue density, erosion of scutum and ossicle were also evaluated.

Presence of ossicle erosion and mastoid invasion was evaluated from intraoperative view, which was reviewed based on operation record. Subtype of cholesteatoma was also evaluated based on intraoperative findings. The lesion was categorized as closed type if the shape of epithelial cyst remains without exposure of keratin. On the contrast, it was categorized as open type if the lesion appeared as flat keratinizing epithelium without formation of an epithelial cyst. (Fig 1.)

Surgical modalities were divided into three categories: trans-canal removal, intact canal wall mastoidectomy and open cavity mastoidectomy. Trans-canal removal was usually performed under microscopic view, but 4 out of 90 cases of trans-canal approach was performed under endoscopic view. Ossicular reconstruction was simultaneously done for the patients if needed.

First follow up and wound evaluation was done about 2 weeks after surgery. The patient visited the clinics with period of about 6 months for otoscopy, and audiologic outcome was also evaluated with pure tone audiometry if possible. However, period of pure tone audiometry after surgery varied according to patient characteristics and surgeon preference. Follow up CT scan was performed in 101 patients with median follow up period of 1.77 years after surgery. The other 85 patients were only followed up with

endoscopic examination. When suspicious recurrence was identified CT scan was done, and reoperation was planned.

Statistics

All values were expressed as a mean ± SD or percentage. Data were analyzed using SPSS version 22.0.

Parametric test was utilized when number of samples in each category was same or above 30 participants or the number of each category is between 10 and 30 with proved normal distribution of data by Shapiro-Wilk test.

To compare clinical characteristics, endoscopic findings, CT findings and intraoperative findings of three age groups, Fisher's exact test were used for analysis. For significant variables, post hoc analysis between each three groups with Bonferroni correction ($P<0.0167$) was performed. Significant difference between the groups in post hoc analysis was descripted on figures.

Perioperative audiometric finding according to clinical variables were analyzed. Unpaired t-test and Mann-Whitney U test were used for comparison of two categories, while ANOVA test and Kruskal-Wallis test were used for comparison of categories more than three. Relationship of age and perioperative audiometric outcome was analyzed by R-squared statistic and general linear models, which were descripted on figures.

For analysis of impact of clinicopathological factors on recurrence of pediatric

congenital cholesteatoma, logistic regression analysis was utilized. Each variable was analyzed for identification of significant factors with p-value under 0.05 in univariate analysis. Multivariate analysis was done for significant factors in univariate analysis to find independent risk factor of recurrence.

RESULTS

Preoperative Clinical Data

Demographic and clinical data of pediatric congenital cholesteatoma were reviewed. (Table 1) The mean age of the study group was 4.9 ± 2.8 years. The number of patients with age less or equal to three years old was 49 (26.3%), age between 3 to 7 years old 106 (57.0%), and that of patients with age over 7 years old was 31 (16.7%). One hundred thirty-six patients were male, more than twice of the number of female patients which is 50 patients. Number of patients was much bigger in age between 2 to 5 with more than 30 patients for each age than other age of patients, but gender proportion did not significantly differ between all number of age. (Fig 2) Right side lesions accounted for 55.4% of patients with 103 cases, while left side lesions accounted for 44.6% with 83 cases. Most patients visited our hospital due to incidental finding of a whitish mass behind the eardrum during otoscopic inspection by local physicians. Another chief complaint included hearing difficulty and referral from other hospital for their otitis media with effusion. Some patients complained of otorrhea and otalgia. There was one patient with facial palsy.

Observers examined tympanic membranes of all patients with endoscopy. (Table 2) Most endoscopic exam showed mass shadow behind the tympanic membrane (TM). (90.3%), while some of the lesions appeared as keratin debris on TM, discharge from middle ear, polypoid lesion and perforation in attic space or pars tensa. Keratin debris appeared in 11.3% of patients, but other lesions accounted for less than 5% each. Location

of the main lesion was anterosuperior area in 37.1% and posterosuperior area in 33.9%. In 7% of patients the lesion occupied space of multiple quadrants. Inferior space lesion accounted for 11.8 % of the patients. But in 19 cases the main lesion could not be identified by endoscopic exam of tympanic membrane and they accounted for 10.2% of the patients. Patients with this 'endoscopically unidentifiable' type visited hospital with chief complaint mainly with hearing disturbance (14 of 19 cases). Other chief complaints included 4 cases of recurrent otorrhea, one case of recurrent otitis media with effusion.

Image findings and Intraoperative Findings

Disease extent and status of temporal bone, middle ear evaluated with CT findings in each patient (Table 3). Sclerotic air cell was present in 9.1% of cases. Most of the patients had soft tissue density in mesotympanum (88.7%) and a lot of patients also had soft tissue density in mastoid or epitympanum (38.2% and 48.9% each).

For management of cholesteatoma, we selected one of three basic techniques, trans-canal tympanoplasty, intact canal wall mastoidectomy (ICW), and open cavity mastoidectomy (OC). Open cavity was performed in 6 cases, while trans-canal approach and intact canal wall mastoidectomy was used in 90 cases each. During middle ear exploration, ossicle and mastoid cavity was evaluated. While malleus erosion occurred in only about 10% of patients, incus and stapes erosion occurred in 50.3% and 40.3% of the patients each. The number of patients who had ossicle erosion in any site was 104,

accounting for 55.9% of the patients. Mastoid invasion was identified in 25.3% of the patients.

Congenital cholesteatoma is divided into closed type and open type. Proportion of closed type was 45.7% (85 cases) and open type was 54.3% (101 cases).

Postoperative Clinical Data during Follow up

Postoperative evaluation of patients who underwent surgery for congenital cholesteatoma was done with CT scan, and 33 cases (17.7%) of recurrent cholesteatoma were identified during follow up. Revision surgery was done and recurrent lesion was pathologically confirmed for the recurred patients. Nine patients (27.3%) underwent transcanal removal of recurrent lesion, 16 patients (48.5%) underwent canal wall up mastoidectomy and remaining 8 patients (24.2%) had to undergo canal down mastoidectomy at last. But among recurrent diseases, there was attic destruction or severe tympanic membrane retraction on 9 patients (27.3%) which could have caused newly acquired cholesteatoma. These lesions were sometimes hardly able to be distinguished from recurred congenital lesion.

Difference of Clinical Characteristics between Age Groups

Age distribution of clinical variables described above was calculated and difference of these variables between three group was analyzed (Table 4).

Gender distribution did not show significant difference between all three age groups, but proportion of male was dominant in all age groups. Side of the lesion was also not related with age distribution. Distribution of chief complaints according to age was analyzed (Fig 3A). Incidental mass occupied most of the chief complaints in the youngest age group and middle age group, while hearing disturbance was the most frequent chief complaint in the oldest group. The patient group older than 7 years-old showed significantly lower proportion of incidental mass and higher proportion of hearing disturbance than both patient groups younger than 7 years-old (Fig 3B, 3C). Other chief complaints including referred otitis media with effusion did not show significant difference between age groups.

Proportion of keratin debris on tympanic membrane, polypoid lesion, discharge, attic perforation and pars tensa perforation on endoscopic exam did not significantly differ according to age. Mass shadow was present in most patients with age under 7 years-old, but it could be seen in only 64.5% of patients who were over 7 years-old.

Location of the main lesion according to age group was analyzed (Fig 4A). The main lesion was mostly located on anterosuperior quadrant (AS) of tympanic membrane in age group under 3 years-old, which was 61.2% of the group. On the other hand, the most frequent main mass location in middle group was posterosuperior quadrant (PS) (37.7%), which was slightly more than AS (30.2%). This trend was maintained in the oldest age group in which 25.8% of the lesion was on PS and 22.6% in AS. This is because of

decreased proportion of AS lesion in two older groups than the youngest group (Fig 4B). But endoscopically unidentifiable cholesteatoma on tympanic membrane exam greatly increased in the oldest group. Endoscopically unidentifiable lesion accounted for 29.0% of the oldest group, which was significantly higher than two younger groups (Fig 4C).

CT findings including proportion of air cell sclerosis, soft tissue density of mastoid, scutum erosion, ossicle erosion and epitympanic soft tissue density were all related with age increase. Only presence of mesotympanic soft tissue density was not related with age. Operation approach according to age group was analyzed (Fig 5). Trans-canal approach was more frequently used as the age gets earlier, while ICW and OC were more frequently used as the age gets older. Group of age under 3 years old showed significant difference of utilized approach method with the other two groups in post hoc analysis ($P<0.0167$). Latter two groups did not show significant difference in post hoc analysis ($P=0.089$), but the trend that older group gets more aggressive surgery was maintained.

During exploration of middle ear and mastoid in surgery, ossicle erosion and mastoid invasion according to age were analyzed (Fig 6). Mastoid invasion increased elevated as age elevated, though not significant ($P=0.145$) (Fig 6A). Malleus erosion was identified in 19.4% in the oldest group, which was higher rate than two younger groups but not significant. Incus erosion and stapes erosion increased as age elevated ($P<0.001$). Accordingly, overall rate of ossicle erosion increased as age elevated ($P<0.001$) (Fig 6B). Especially, the oldest group showed significant increase of frequency than the other two

groups in post hoc analysis. Open type cholesteatoma also increased as age elevated ($P<0.001$) (Fig 6C). It showed similar prevalence with ossicle erosion grossly, but age group under three years-old showed significantly lower proportion than the other two groups.

Audiologic Outcome in Pediatric Congenital Cholesteatoma Patients

Perioperative audiological outcome of 105 pediatric congenital cholesteatoma was analyzed and air-bone gaps (ABG) were compared between groups divided by various criteria. (Table 5) Overall mean of preoperative ABG was 24.3 dBnHL, postoperative ABG was 19.3 dBnHL and ABG improvement was 5.0 dBnHL in average. Preoperative ABG worsened as age increased, but ABG improvement also increased so that postoperative ABG did not show significant difference according to age (Fig 7). The youngest group did not show significant difference between the other two groups because of insufficient data cases. But there was significant difference of preoperative ABG and ABG improve between the middle group and the oldest group.

Preoperative ABG increased in endoscopically unidentifiable cholesteatoma. But mean postoperative ABG of patients with in endoscopically unidentifiable cholesteatoma is not significantly different from the other patients, with significantly high ABG improvement after operation, which was similar tendency of difference according to age. Patients with open type cholesteatoma, ossicle erosion except malleus erosion, mastoid

invasion showed worse preoperative and postoperative ABG. Significant air-bone gap improvement could be seen in open type and incus erosion. Among operation procedures, ICW and OC showed elevated perioperative ABG than trans-canal approach. Air-bone gap improvement was also higher but not significant. Perioperative ABG was even higher in OC than ICW but preoperative, postoperative and improvement of ABG between ICW and OC was not significantly different (P=0.651, 0.651, 0.284 each).

Risk of Recurrence after Operation of Pediatric Congenital Cholesteatoma

Risk of recurrence for clinical variables was analyzed and odd ratios were calculated. (Table 6) Recurrence significantly elevated as age of the group increased. The youngest group showed only 6.1% of recurrence, while the middle group showed 19.8% and the older group 29.0%. Gender was not significantly related with recurrence rate.

Open type cholesteatoma showed higher risk of recurrence than closed type cholesteatoma. Location of the lesion on tympanic membrane was not related with recurrence risk, but posterosuperior and inferior lesions were more likely to recur than anterosuperior lesions. Lesion on multiple quadrants tended to have even higher risk of recurrence than the other lesions.

When there is ossicle erosion or mastoid invasion of the disease, the risk of recurrence significantly elevated. Among operation procedures, recurrence risk of trans-canal approach (10.0%) was lowest. When ICW and OC were compared, patients who

underwent ICW showed higher (25.6%) and risk of OC (16.7%) was lower. Risk of ICW was significantly higher than trans-canal approach, but risk difference of OC and ICW did not show significance when trans-canal approach is excepted from analysis. (P=0.630)

In multivariate analysis, no significant clinical variables were identified (Table 7). But there was trend of elevated risk in the cases of higher age, presence of ossicle erosion and mastoid invasion. This trend was also found in ICW compared with OC and trans-canal approach. Age group over 7 years-old showed especially elevated risk of recurrence compared to age group under 3 years-old, with odds ratio of 4.21 and p-value of 0.056.

Table 1. Demographic data and clinical results of pediatric congenital cholesteatoma

		Value
Age (years; means ± SD)		4.9 ± 2.8
Age category, No (%)		
≤ 3 y		49 (26.3%)
3-7 y		106 (57.0%)
> 7y		31 (16.7%)
Gender, No (%)		
Male		136 (73.1%)
Female		50 (26.9%)
Side, No (%)		
Right		103 (55.4%)
Left		83 (44.6%)
Chief complaints, No (%)	Incidental mass	125 (67.2%)
	Hearing difficulty	25 (13.4%)
	Otorrhea	8 (4.3%)
	Otalgia	7 (3.8%)
	OME, referral from other clinics	20 (10.8%)
	Facial palsy	1 (0.5%)
Follow-up period (months; means ± SD)		47.0 ± 26.8
Recurrence, No (%)		33 (17.7%)

Abbreviations; SD, standard deviation; OME, otitis media with effusion.

Table 2. Endoscopic findings of tympanic membrane in pediatric cholesteatoma patients

		N (%)
Abnormal lesions on TM exam No (%)	Keratin debris	21 (11.3%)
	Polyp	1 (0.5%)
	Discharge	8 (4.3%)
	Attic perforation	3 (1.6%)
	Pars tensa perforation	5 (2.7%)
	Mass shadow	168 (90.3%)
Location of lesion on TM exam, No (%)	AS	69 (37.1%)
	PS	63 (33.9%)
	AS and PS	13 (7.0%)
	Inferior	22 (11.8%)
	Unidentifiable	19 (10.2%)

Abbreviations; TM, tympanic membrane; AS, anterosuperior quadrant; PS, posterosuperior quadrant.

Table 3. CT findings, intraoperative ossicle and mastoid manifestations, and subtypes of pediatric congenital cholesteatoma.

		N (%)
CT findings, No (%)	Sclerotic air cell	17 (9.1%)
	Mastoid STD	71 (38.2%)
	Scutum erosion	14 (7.5%)
	Ossicle erosion	92 (49.5%)
	Mesotympanum STD	165 (88.7%)
	Epitympanum STD	91 (48.9%)
Operation, No (%)	Trans-canal	90 (48.4%)
	ICW	90 (48.4%)
	OC	6 (3.2%)
Ossicle and mastoid invasion No (%)	Malleus erosion	19 (10.2%)
	Incus erosion	101 (54.3%)
	Stapes erosion	75 (40.3%)
	Ossicle erosion (any site)	104 (55.9%)
	Mastoid invasion	47 (25.3%)
Type, No (%)	Closed	85 (45.7%)
	Open	101 (54.3%)

Abbreviations; STD, soft tissue density; ICW, intact canal wall mastoidectomy; OC, open cavity mastoidectomy.

Table 4. Comparison of age distribution of clinical characteristics, endoscopic findings, CT findings and intraoperative findings.

Variables	**≤ 3 y (*n* = 49)**	**3-7 y (*n* = 106)**	**> 7y (*n* = 31)**	***P***
Gender, male	38 (77.6%)	77 (72.6%)	21 (67.7%)	0.619
Side, right	24 (49.0%)	63 (59.4%)	16 (51.6%)	0.428
Chief complaints				
Incidental mass	42 (85.7%)	72 (67.9%)	11 (35.5%)	<0.001
Hearing difficulty	1 (2.0%)	9 (8.5%)	15 (48.4%)	<0.001
Otorrhea	1 (2.0%)	4 (3.8%)	3 (9.7%)	0.240
Otalgia	0	6 (5.7%)	1 (3.2%)	0.224
Referred OME	5 (10.2%)	14 (13.2%)	1 (3.2%)	0.285
Abnormal TM lesions				
Keratin debris	6 (12.2%)	12 (11.3%)	3 (9.7%)	0.939
Polyp	0	0	1 (3.2%)	0.081
Discharge	1 (2.0%)	4 (3.8%)	3 (9.7%)	0.240
Attic perforation	0	3 (2.8%)	0	0.316
P.tensa perforation	1 (2.0%)	2 (1.9%)	2 (6.5%)	0.365
Mass shadow	48 (98.0%)	100 (94.3%)	20 (64.5%)	<0.001
Location of lesion on TM exam				
AS	30 (61.2%)	32 (30.2%)	7 (22.6%)	<0.001
PS	15 (30.6%)	40 (37.7%)	8 (25.8%)	0.399
Multiple quadrant	1 (2.0%)	9 (8.5%)	3 (9.7%)	0.278
Inferior	2 (4.1%)	16 (15.1%)	4 (12.9%)	0.140
Unidentifiable	1 (2.0%)	9 (8.5%)	9 (29.0%)	<0.001
CT finding				
Sclerotic air cell	3 (6.1%)	7 (6.6%)	7 (22.6%)	0.017
Mastoid STD	12 (24.5%)	43 (40.6%)	16 (51.6%)	0.038
Scutum erosion	0	9 (8.5%)	5 (16.1%)	0.024
Ossicle erosion	14 (28.6%)	52 (49.1%)	26 (83.9%)	<0.001
Mesotymp STD	43 (87.8%)	96 (90.6%)	26 (83.9%)	0.567

Epitymp STD	15 (30.6%)	55 (51.9%)	21 (67.7%)	<0.003
Operation				
Trans-canal	36 (73.5%)	47 (44.3%)	7 (22.6%)	
ICW	13 (26.5%)	55 (51.9%)	22 (71.0%)	<0.001
OC	0	4 (3.8%)	2 (6.5%)	
Ossicle erosion (any site)	18 (36.7%)	59 (55.7%)	27 (87.1%)	<0.001
Malleus erosion	4 (8.2%)	9 (8.5%)	6 (19.4%)	0.183
Incus erosion	18 (36.7%)	57 (53.8%)	26 (83.9%)	<0.001
Stapes erosion	13 (26.5%)	41 (38.7%)	21 (67.7%)	0.001
Mastoid invasion	8 (16.3%)	28 (26.4%)	11 (35.5%)	0.145
Type, open	15 (30.6%)	62 (58.5%)	24 (77.4%)	<0.001

Abbreviations; OME, otitis media with effusion; TM, tympanic membrane; P.tensa, pars tensa; AS, anterosuperior quadrant; PS, posterosuperior quadrant; STD, soft tissue density; ICW, intact canal wall mastoidectomy; OC, open cavity mastoidectomy;

Table 5. Perioperative audiometric finding in pediatric congenital cholesteatoma and difference of air-bone gap according to clinical variables.

		Preop ABG	***P***	**Postop ABG**	***P***	**ABG improve**	***P***
Overall (*n*=105)		24.3 (15.6)		19.3 (15.0)		5.0 (14.5)	
Age groups	≤ 3 y (*n*=6)	28.3 (16.4)	0.037†	16.7 (12.9)	0.548†	11.7 (9.1)	0.039†
	3 ~ 7 y (*n*=69)	21.4 (14.2)		18.8 (15.8)		2.5 (12.9)	
	> 7 y (*n*=30)	30.4 (17.2)		20.9 (13.2)		9.5 (17.5)	
Unidentifiable on TM exam	Unidentifiable (*n*=17)	33.9 (15.8)	0.005	18.1 (15.1)	0.721†	15.8 (17.2)	0.001
	Identifiable (*n*=88)	22.5 (15.0)		19.5 (15.0)		3.0 (13.1)	
Location of lesion on TM exam	Anterior (*n*=26)	17.5 (16.3)	0.010	14.0 (14.6)	0.016†	3.5 (9.6)	0.442
	Posterior (*n*=39)	26.6 (14.8)		21.0 (14.8)		5.5 (15.8)	
Type	Closed (*n*=36)	14.8 (10.6)	<0.001	13.6 (14.9)	0.004	1.2 (12.6)	0.05
	Open (*n*=69)	29.3 (15.6)		22.3 (14.2)		7.0 (15.1)	
Ossicle erosion	Normal (*n*=33)	10.2 (9.7)	<0.001	7.8 (9.8)	<0.001	2.4 (9.3)	0.134
	Eroded (*n*=72)	30.8 (13.4)		24.6 (13.9)		6.2 (16.3)	

Malleus erosion	Normal (*n*=92)	24.0 (15.8)	0.589	18.5 (14.8)	0.085†	5.6 (13.9)	0.322
	Eroded (*n*=13)	26.5 (14.9)		25.3 (15.0)		1.3 (18.8)	
Incus erosion	Normal (*n*=36)	10.9 (9.8)	<0.001	10.4 (13.1)	<0.001	0.5 (11.1)	0.01
	Eroded (*n*=69)	31.4 (13.4)		23.9 (13.8)		7.4 (15.6)	
Stapes erosion	Normal (*n*=62)	15.8 (13.0)	<0.001	12.2 (12.4)	<0.001	3.6 (13.3)	0.318
	Eroded (*n*=52)	33.4 (13.0)		26.9 (14.7)		6.5 (15.7)	
Mastoid invasion	Normal (*n*=68)	19.6 (14.9)	<0.001	15.8 (14.4)	0.001	3.8 (15.2)	0.231
	Invaded (*n*=37)	33.1 (13.1)		25.8 (13.9)		7.3 (13.0)	
Operation	Trans-canal (*n*=37)	13.2 (11.6)	<0.001 †	12.3 (14.4)	<0.001 †	0.9 (12.3)	0.056†
	ICW (*n*=65)	30.2 (14.5)		23.3 (14.1)		6.9 (15.3)	
	OC (*n*=3)	34.4 (4.2)		18.9 (8.5)		15.6 (12.1)	

Abbreviations; ABG, air-bone gap; ICW, intact canal wall mastoidectomy; OC, open cavity mastoidectomy

† Mann-Whitney U test between two groups and Kruskal Wallis test between three groups.

Table 6. Univariate analyses of clinicopathological factors on recurrence risk of pediatric congenital cholesteatoma.

Variable	Recurred cases (%)	Risk of recurrence		
		OR	95% CI	*P*
Age groups				
≤ 3 y	3 (6.1%)	1		
3 ~ 7 y	21 (19.8%)	3.79	1.07-13.38	0.039*
> 7 y	9 (29.0%)	6.27	1.54-25.49	0.010*
Sex				
Male	24 (17.6%)	1		
Female	9 (18.0%)	1.02	0.44-2.39	0.955
Congenital chole type				
Closed	8 (9.4%)	1		
Open	25 (24.8%)	3.17	1.34-7.46	0.008*
Location of lesion				
Unidentifiable	3 (15.8%)	1		
AS	7 (10.1%)	0.60	0.14-2.59	0.496
PS	13 (20.6%)	1.39	0.35-5.49	0.641
Multiple quadrant	4 (30.8%)	2.37	0.43-13.04	0.321
Inferior	6 (27.3%)	2.00	0.43-9.42	0.381
Ossicle erosion (any site)				
No	8 (9.8%)	1		
Yes	25 (24.0%)	2.93	1.24-6.90	0.014*

Mastoid invasion				
No	19 (13.7%)	1		
Yes	14 (29.8%)	2.68	1.22-5.91	0.015*
Operation				
Trans-canal	9 (10.0%)	1		
ICW	23 (25.6%)	3.09	1.34-7.13	0.008*
OC	1 (16.7%)	1.80	0.19-17.16	0.609

Abbreviations: OR, odds ratio; CI, confidence interval; AS, anterosuperior quadrant; PS, posterosuperior quadrant; ICW, intact canal wall mastoidectomy; OC, open cavity mastoidectomy

cf) *P* means p-value of odds ratio of the group compared to the first group of each variables on logistic regression analysis.

* Significant p-value ($p < 0.05$) on logistic regression analysis

Table 7. Multivariate analysis of factors on recurrence risk pediatric congenital cholesteatoma.

	Risk of recurrence		
Variable	OR	95% CI	*P*
Age groups			
≤ 3 y	1		
3 ~ 7 y	3.15	0.86-11.53	0.083
> 7 y	4.21	0.96-18.41	0.056
Ossicle erosion	1.37	0.36-5.29	0.648
Mastoid invasion	1.85	0.73-4.69	0.198
Type of cholesteatoma, open	1.71	0.57-5.21	0.340
Operation			
Trans-canal	1		
ICW	1.02	0.25-4.21	0.982
OC	0.49	0.04-6.49	0.589

Abbreviations: OR, odds ratio; CI, confidence interval; ICW, intact canal wall mastoidectomy; OC, open cavity mastoidectomy

cf) If there are only two groups in each variables, *P* means p-value of odds ratio of the group in which the manifestation exists compared to the group that does not. If there are more than three groups, it means p-value of odds ratio of that group compared to the first group of each variables on logistic regression analysis.

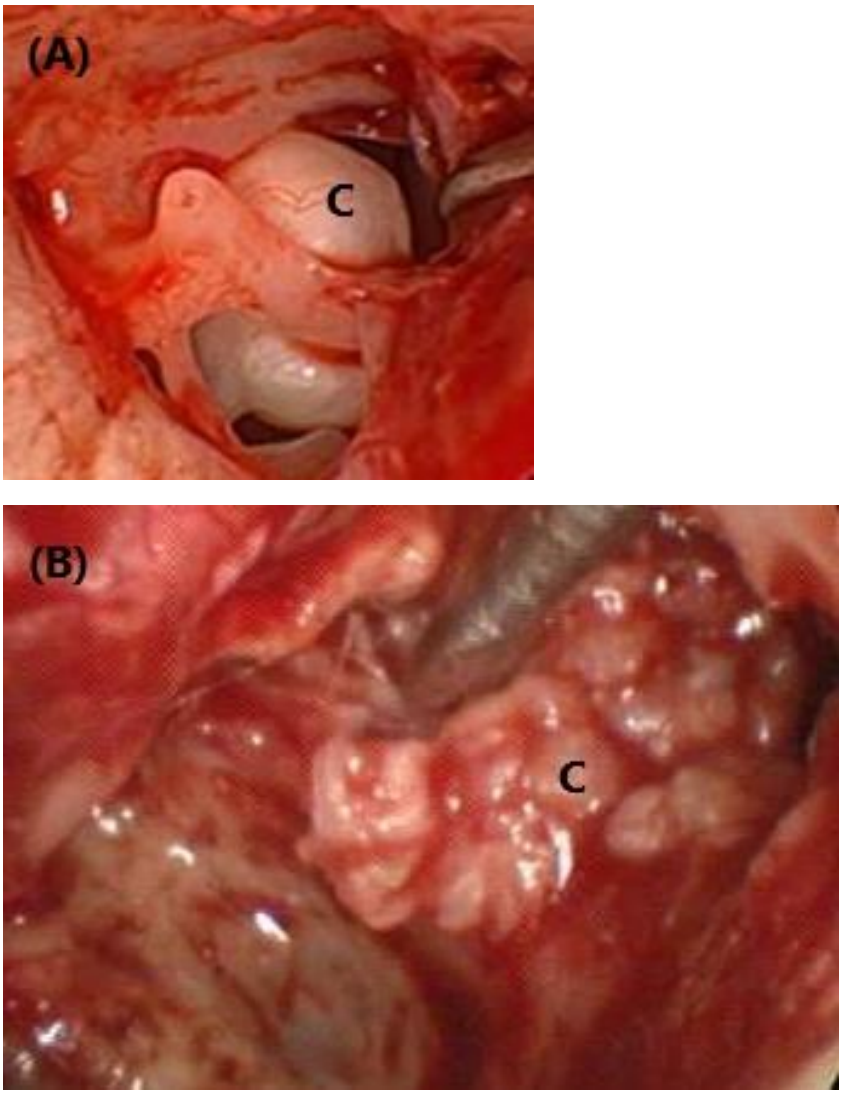

Figure 1. Closed type congenital cholesteatoma (A) and open type cholesteatoma (B) observed by endoscopy intraoperatively. In closed type, cholesteatoma sac (C) has clear and well demarcated margin, maintaining spherical shape. In open type cholesteatoma, however, keratinizing epithelium is protruded out and cholesteatoma sac (C) appears as irregular shaped lesion with poor margin.

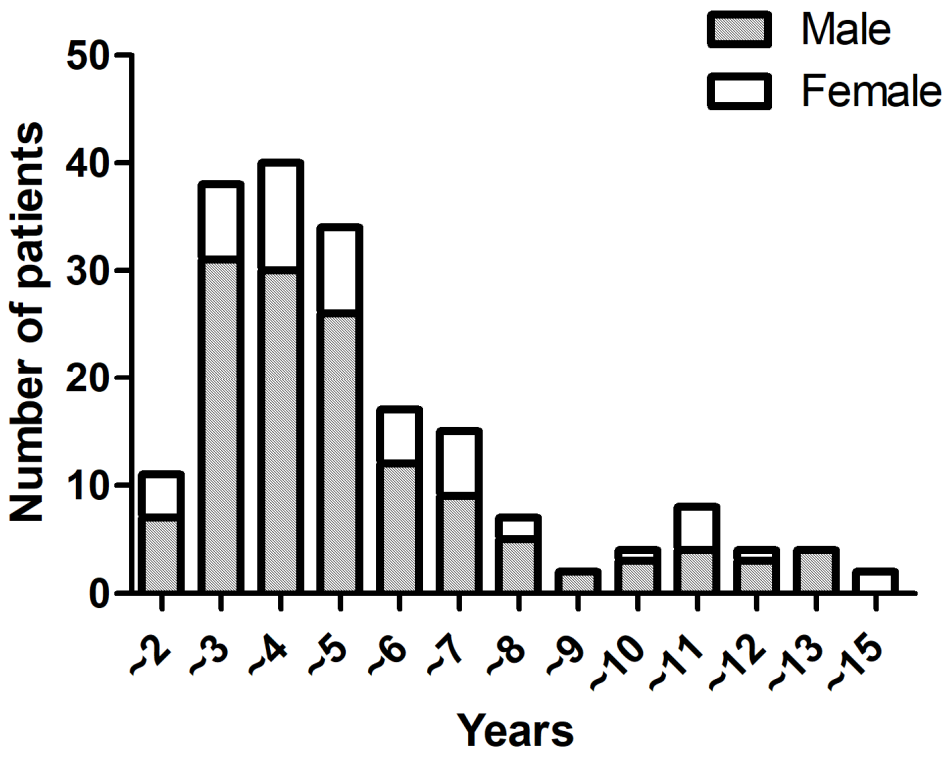

Figure 2. Age and gender distribution of the study patients with pediatric congenital cholesteatoma. The prevalence of cholesteatoma was higher in younger age, with a greater prevalence seen in boys in nearly all age groups.

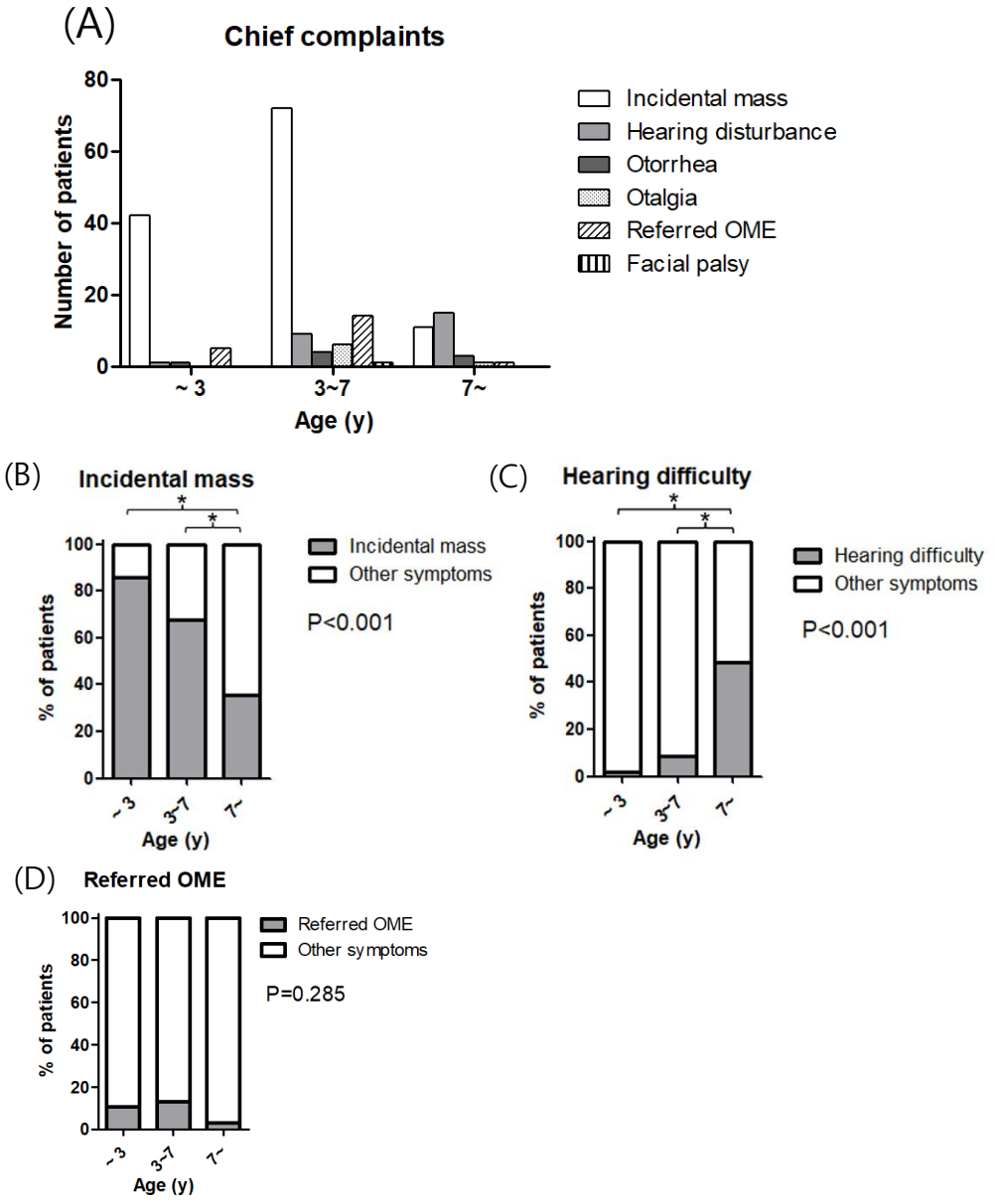

Figure 3. Age distribution of each chief complaints of patients with pediatric congenital cholesteatoma. Patients who complained of incidental mass was mainly younger patients, while those who complained of hearing disturbance was older patients. (P<0.001) Proportion of other chief complaints did not significantly differ according to age.

* Significant difference in post hoc analysis of Fisher's exact test with Bonferroni correction (P<0.0167)

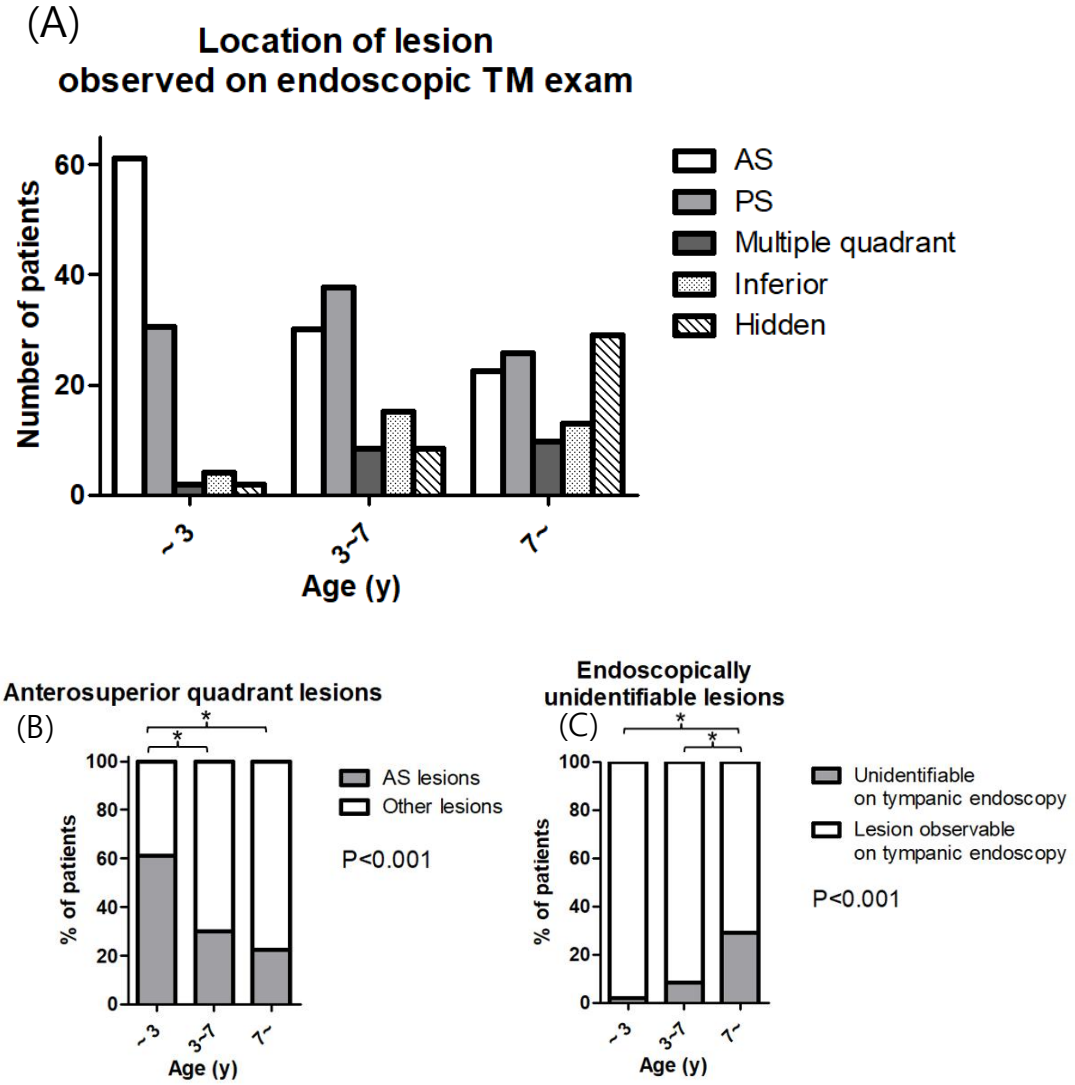

Figure 4. Proportion of each locations of lesions and unidentifiable lesions on endoscopy in pediatric congenital cholesteatoma patients according to age. Proportion of anterosuperior quadrant lesion decreases as age increases. On the other hand, proportion of endoscopically unidentifiable pediatric congenital cholesteatoma increases according to age. (P<0.001) Other location did not show significant relationship with age.

* Significant difference in post hoc analysis of Fisher's exact test with Bonferroni correction (P<0.0167)

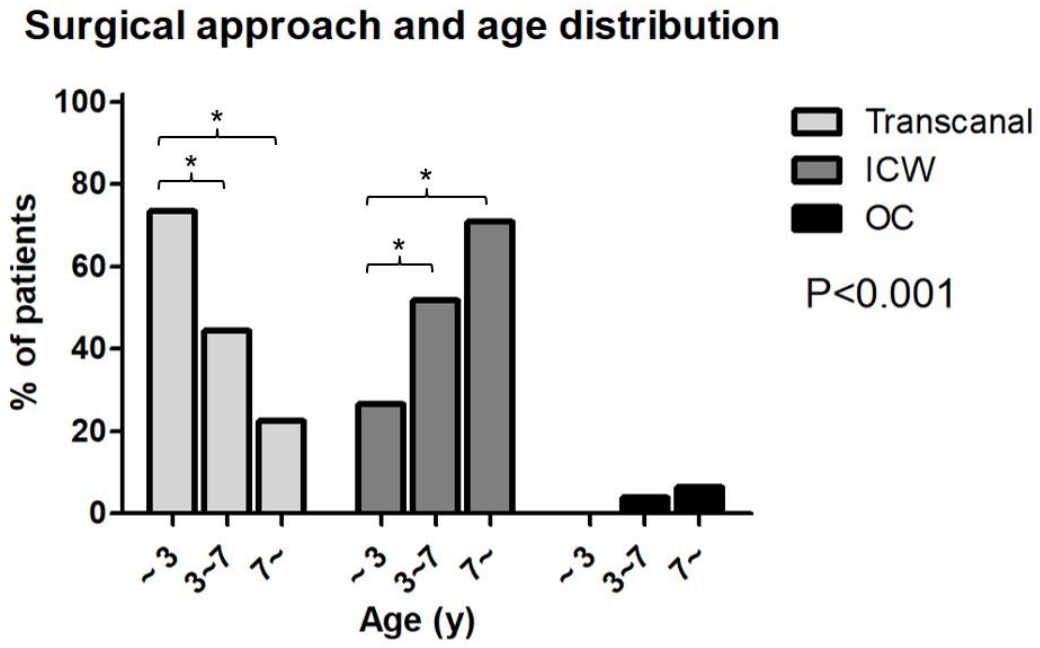

Figure 5. Age distribution of surgical management in congenital pediatric cholesteatoma patients. Trans-canal approach was more frequently used in earlier age, while intact canal wall mastoidectomy (ICW) and open cavity mastoidectomy (OC) was more frequently used in older age. Post hoc analysis with chi-square test and Bonferroni correction method was done. Proportion of transcanal approach significantly increased and that of intact canal wall mastoidectomy significantly decreased in the youngest group than the other two groups. ($P<0.0167$) Difference between the oldest group and group with age 3 to 7 was not significant. Difference of proportion of open cavity mastoidectomy between age groups was not statistically significant.

* Significant difference in post hoc analysis of Fisher's exact test with Bonferroni correction ($P<0.0167$)

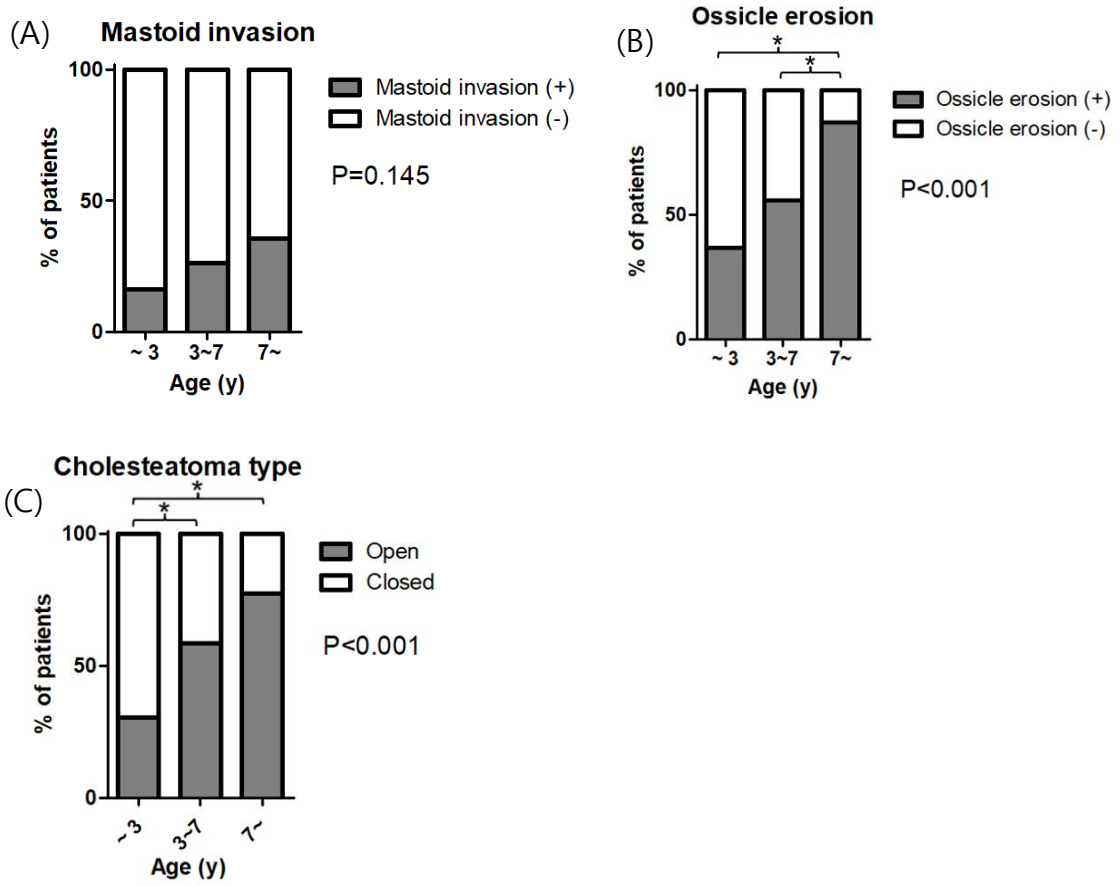

Figure 6. Mastoid invasion, ossicle erosion and type of pediatric congenital cholesteatoma according to age groups. Mastoid invasion rate had increasing tendency according to age, but it was not significant. (P=0.145) Ossicle erosion and open type cholesteatoma showed significant increase according to age. (P<0.001) Ossicle erosion especially increased after 7 years old, and open type cholesteatoma after 3 years old.

* Significant difference in post hoc analysis of Fisher's exact test with Bonferroni correction (P<0.0167)

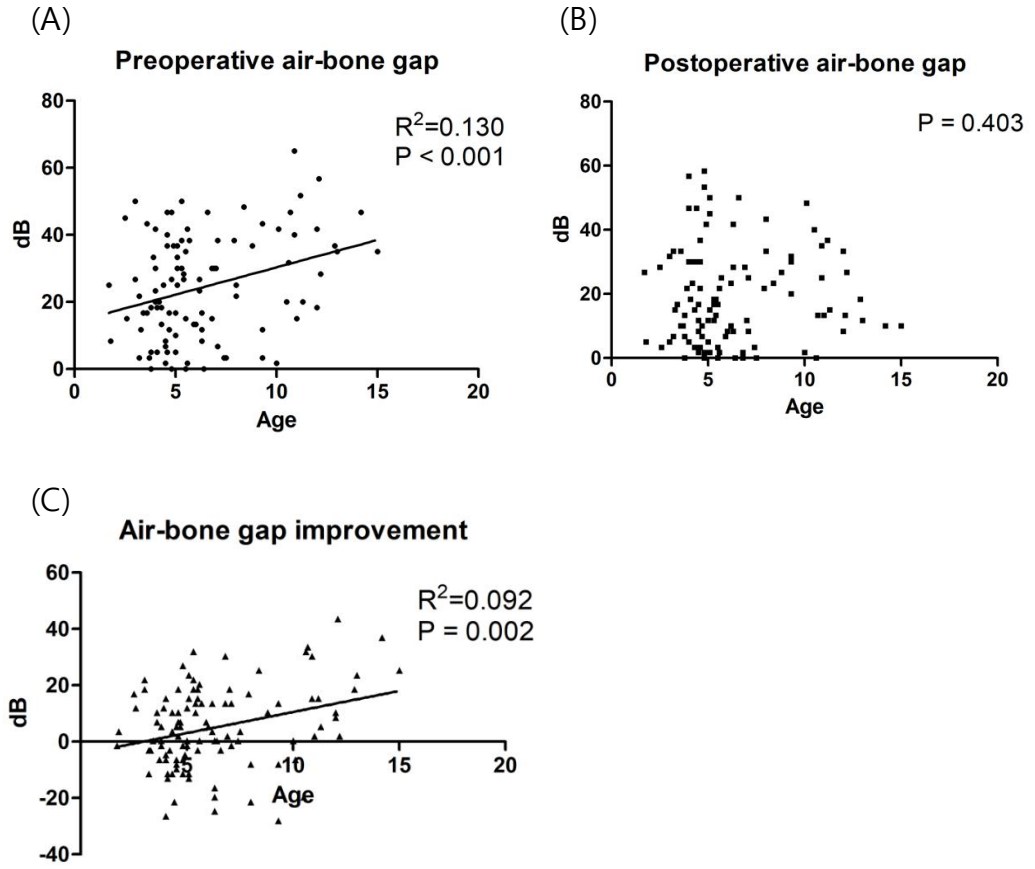

Figure 7. Air-bone gap of preoperative and postoperative period measured by pure tone audiometry. Preoperative air-bone gap shows aggravating tendency as age increases (P<0.001), while postoperative air-bone gap shows no specific tendency. (P=0.403) Air-bone gap improvement tends to be higher in patients with increased age. (P=0.002)

DISCUSSION

The primary goal of cholesteatoma surgery is to eradicate disease completely and improve hearing. In general, cholesteatoma is more aggressive and has a higher recurrence rate in children than in adults. [29-32] Roger et al. reported that four factors were related with residual cholesteatoma in children: invasion of the posterior middle ear, presence of ossicular erosion, skill of the surgeon, and the presumption of incomplete ablation. [33] Stangerup et al. found correlations between outcome and age, poor ET function, cholesteatoma extent, and ossicular erosion. [34] In our present study, the overall recurrence rate of cholesteatoma after first operation was 17.7%, which is slightly lower than the values of 19~25.6% which were previously reported. [30,35] When the patient group with attic destruction or severe tympanic membrane retraction during postoperative follow up who were suspicious to be obtained newly acquired cholesteatoma is excluded (9 patients), actual recurrence rate can be calculated as 12.9%.

Our current data showed that the prevalence of congenital cholesteatoma was highest in children of age two to five and decrease after five years old in our study cohort. Its incidence is increasing due to developing diagnostic tools such as microscopy and endoscopy, and regular otologic examination of children. [9] More screening and early diagnosis of pearly, whitish mass behind the intact tympanic membrane with otoscope or otoscope might lead to higher prevalence of congenital cholesteatoma in younger children. Main chief complaint of patients in our study was incidental mass found in local clinics.

Potsic et al. also reported that the most common presentation was asymptomatic mass lesion, accounting for 82% of the cases. [36] Patients who suffered from hearing difficulty as main chief complaint drastically increased after 7 years old. Considering that ossicle erosion increased gradually according to age, the reason of this drastic increase is that the symptom could remain silent because the patients lacked ability to communicate with their parents or did not realize their own symptom in age younger than 7 years-old. Patients who visited hospital for incidental mass might also have combined undetected hearing loss or other symptoms.

About 10 percent of congenital cholesteatoma was could not be identified via otoscopic examination. They are more likely to remain undetected until they grow up, which explains elevated proportion of this endoscopically unidentifiable type in older patients. These patients visited the hospital with chief complaint of hearing disturbance, recurrent otorrhea and recurrent effusion. Diagnostic tests should be considered in active manner for the patients with these symptoms.

Presence of cholesteatoma in multiple quadrant was found in only 7% of patients and showed no significant relationship with age, hearing level or recurrence in our study, though it showed higher odd ratio than the case in which the lesion was on other locations. Patients with lesion in only anterosuperior quadrant showed tendency of lower recurrence rate than the other lesions, though not statistically significant. Potsic et al. reported that when the lesion existed in any quadrant except the anterior-superior quadrant it was a

predictor of residual cholesteatoma, with extensive disease more common in those cases. [37] In addition, our data also suggested better prognosis of audiologic outcome of anterosuperior quadrant lesions than posterosuperior quadrant lesions. (Table 5)

CT findings of older congenital cholesteatoma patients showed more extensive disease with increased sclerosis, erosion of ossicle and surrounding bone, extension to epitympanic space and mastoid, thus they tended to need more aggressive surgical procedure. About 54% of patients had open type cholesteatoma and it significantly increased with age. Andrea et al. also reported similar tendency according to age, but only 26% of open type cases identified among the study group. [38] Several hypothesis on pathogenesis of open type cholesteatoma which can explain the tendency has been suggested including rupture of closed cholesteatoma or relationship between formation of closed type cholesteatoma and middle ear inflammation, but it still remains unclear. Many closed type cholesteatoma patients were excluded from the study due to short follow-up period, so prevalence of open type should be identified in more precise prospective study.

Preoperative audiologic outcome was worse in patients with invisible lesion, lesion in posterosuperior quadrant, open type, erosion of ossicles and invasion of mastoid. Aggressive disease and late detection of the lesion can affect hearing level. Despite of bigger ABG, significant ABG improvement could not be achieved in patients with posterosuperior quadrant lesion, stapes erosion, and mastoid invasion. According to Stapleton et al., perioperative hearings level of congenital cholesteatoma were

significantly correlated with elevated Potsic stage, which is indicator of extent of disease including invasion of ossicle and mastoid, while they obtained excellent audiometric results of under 20 decibel hearing level in 77 percent of the patients. [39]

Trans-canal approach, mostly performed in patients with limited extent, was done for patients with good hearing levels and showed best hearing outcome without deterioration of hearing level. OC and ICW showed worse preoperative hearing level and postoperative outcome than trans-canal approach. Kobayashi et al. also stated that there was no hearing deterioration after trans-canal surgery of congenital cholesteatoma, although hearing assessment was only available seven patients. [40] ICW and OC did not show significantly different hearing outcome in our study.

Preoperative air-bone gap increased with age, as patient with older age is more likely to have aggressive disease. Patients with endoscopically unidentifiable cholesteatoma also showed similar tendency of higher air-bone gap than the other cholesteatoma patients, but their postoperative air-bone gap was not inferior to that of other cholesteatoma patients. However, air-bone gap improvement of some patient with older age was drastic, which resulted in good hearing level under 20 dBnHL. Concurrent ossicular prothesis during first surgery was more frequent in older patients, which could affect the result. However, it suggests that the patients diagnosed late can also achieve as good audiological outcome as younger patients via ossicular reconstruction. This result emphasizes the importance of regular otologic and audiologic evaluation in pediatric patients and consideration of CT

scan for patients with conductive hearing loss or recurrent otorrhea who are refractory to medical treatment to rule out endoscopically unidentifiable cholesteatoma.

Our study could not suggest independent prognostic factor of recurrence in multivariate study, but it suggested that the patients with increased age, ossicular erosion, mastoid invasion, open type cholesteatoma and canal wall up mastoidectomy can elevate the risk of recurrence. Stapleton et al. also stated that extent of primary lesion, ossicle erosion and necessity for ossicle removal were positively related with residual disease of congenital cholesteatoma. [39] And patients with age over 7 years-old showed especially elevated risk of recurrence than age under 3 years-old with p-value of 0.056 while other factors not showing significance. This can imply the fact that patients with higher age are actually late-detected patients of congenital cholesteatoma, and this late detection caused all other adverse changes including ossicular and mastoid damage, cholesteatoma rupture which led to aggressive operation procedure, and the importance of early detection should be emphasized for lowering the risk of recurrence.

Among surgical management of pediatric congenital cholesteatoma, transcanal tympanoplasty plays a major role in early stage. Recent endoscope-guided dissection enabled more benefit for prevention of residual cholesteatoma and facilitates a minimally invasive approach. James at al. reported 19% of recurrence rate in transcanal approach of pediatric cholesteatoma, 24% in microscopic surgery and 15% in endoscopic surgery. [41] We also introduced endoscopic device in lots of cases, and our result showed 10.0%

recurrence rate in transcanal approach, suggesting efficacy of minimally invasive transcanal approach for managing pediatric cholesteatoma. The frequency of transcanal approach decreased as the patients' age increased, since increased extent of disease raises the need for mastoidectomy approach.

There is always a question whether to preserve posterior canal wall or not during mastoidectomy procedure. Alexander et al. reported 89.5% of canal wall preservation rate during mastoidectomy in pediatric age with achieving better audiometric outcomes and easier postoperative care. But the author suggested that need for revision surgery was higher in the canal wall-up mastoidectomy group (51%) compared with the canal wall-down group (21%). [42] In our result, only 3.2% of patients underwent open cavity mastoidectomy and 16.7% of them should have had revision operation. Patients who underwent intact canal wall mastoidectomy accounted for 48.4% of total patients and showed revision rate of 25.6%, which was slightly higher tendency of occurrence of recur than that of open mastoidectomy technique. Multivariate study showed lower risk of recurrence in open mastoidectomy, but it was not statistically significant. As recurrence rate and hearing prognosis of ICW and OC is not drastically different, less invasive, and more functional treatment can be achieved with little concern of recurrence if careful patient selection is proceeded. Chadha et al. suggested not only less ossicular damage and better preoperative hearing level as good prognostic factor of better hearing postoperatively, but also canal wall up mastoidectomy compared with canal wall down

procedure. [43] Our result did not show worse hearing outcome in OC compared to ICW, and canal wall down approach should also be considered as a treatment option for complete eradication of very extensive disease with ossicle erosion and mastoid invasion or patients who failed after canal wall up mastoidectomy, despite the need of regular follow up for cleaning the cavity after surgery.

There are several limitations on this study. Although we indirectly measured extensiveness of the disease with invaded quadrants, ossicular damage and mastoid invasion as Potsic's stage stated, we did not measure exact size of the mass. Patients with minimally aggressive disease were likely to be lost during follow up, so the prevalence of aggressive disease is inaccurate. Follow-up period of pure tone audiometry was irregular, so it was not precise enough to clarify prognostic factors of audiologic outcome. Audiologic evaluation of patients under three years-old was insufficient because of lack of number of patients who can obey command in this age group. This study was retrospective and other neglected confounding factors could exist. Intraoperative findings are reviewed based on operation record, but the information in the record can be subjective and can contain imprecise data according to the recorder. We considered the disease recurred if cholesteatoma was identified in revision surgery, but newly acquired cholesteatoma due to attic destruction or severe tympanic retraction is not actual recurrence of congenital cholesteatoma. But combined congenital cholesteatoma also cannot be excluded in the cases that newly acquired lesions seem to be present. And sometimes distinguishing these

cases were impossible because of insufficient data. So we assumed all of these cases recurrence. So actual demographic data and analysis on risk of recurrence can differ from actual manifestation

CONCLUSION

In our current analyses, there was a significantly higher rate of open type cholesteatoma, mastoid invasion of cholesteatoma and damaged ossicles in older children compared to younger children. Delayed detection of pediatric cholesteatoma can lead to ossicular damage and aggressive operation which can result in worse hearing outcome and elevated risk of recurrence.

Our study also suggested efficacy of minimally invasive transcanal approach for managing pediatric cholesteatoma and preserving posterior canal wall during mastoidectomy can be a reasonable option when mastoidectomy is needed.

Physicians should familiarize themselves with the clinical characteristics of pediatric cholesteatoma, especially regarding the age distribution of this condition.

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국문 요약

목적: 본 연구는 소아 선천성 중이 진주종 환자에서 연령대별로 나타나는 임상적 특징과 수술적 치료의 예후 차이를 확인하고 분석하는데 그 목적이 있다.

방법: 1993 년부터 2016 년까지 선천성 중이 진주종으로 본원에서 확진 되어 수술적 치료를 시행 받은 15 세 이하의 환아 186 명 (136 명의 남아, 50 명의 여아)을 대상으로 후향적 의무기록 분석을 진행하였다. 환자를 나이에 따라 세 그룹으로 나누어 그룹에 따른 임상적 소견의 차이를 분석하였으며, 다양한 임상적 소견과 청력 평가 및 재발률 간의 관계를 분석하였다.

결과: 성별, 환측 방향, 그리고 재발률은 나이에 따라 유의미한 차이를 보이지 않았으나 주 증상, 진주종의 종류, 그리고 수술적 치료 방법은 나이에 따른 유의미한 차이가 있었다. 나이가 적은 환자 군에서는 나이가 많은 환자들보다 적은 유양동 침범, 유의미하게 높은 비율의 정상 이소골 소견이 확인되었다. 이소골의 미란, 유양동 침범 및 개방형 진주종이 존재할 경우에는 청력 예후가 떨어졌고 재발률도 증가하였다. 경외이도 접근법을 이용한 수술은 상대적으로 좋은 예후를 보여주었다. 유양동 절제 시 외이도 후벽을 보존하면 재발률이 다소 높았으나 청력의 예후에는 영향이 없었다.

결론: 소아 중이 진주종의 진단이 늦어지면 병변의 광범위한 침범으로 더 적극적인 수술방법을 요하게 되고 이는 간접적으로 청력과 재발률의 악화를 초래할 수 있다. 본 질환의 진단과 치료에 있어서 환자의 연령에 따른 임상적 특성에 대한 고려가 충분히 이루어져야 하겠다.

중심단어: 소아, 진주종, 중이, 연령