The Correlation Between Pre and Postoperative Hearing Level with High Resolution Computed Tomography (HRCT) Findings in Congenital Canal Atresia (CAA) Patients

A. Asma · A. W. Abdul Fatah · A. H. Hamzaini ·
A. Mazita

Received: 28 June 2010 / Accepted: 20 December 2011 / Published online: 10 January 2012
© Association of Otolaryngologists of India 2012

Abstract In managing patient with congenital aural atresia (CAA), preoperative high resolution computed tomography (HRCT) scan and hearing assessment are important. A grading system based on HRCT findings was first introduced by Jahrsdoefer in order to select appropriate candidates for operation and to predict the postoperative hearing outcome in CAA patients. The score of eight and more was considered as a good prognostic factor for hearing reconstruction surgery. However previously in our center this score was not used as the criteria for surgical procedure. This study was conducted at Center A to evaluate the correlation between pre and postoperative hearing level with HRCT based on a Jahrsdoefer grading system in patients with CAA. All records and HRCT films with CAA from January 1997 until December 2007 at Center A were evaluated. The demographic data, operative records, pre and post operative hearing levels and HRCT findings were analyzed. Hearing level in this study was based on a pure tone average of air-bone gap at 500 Hz, 1 kHz and 2 kHz or hearing level obtained from auditory brainstem response audiometry. This study was approved by Research Ethics Committee (code number, FF-197-2008). Thirty-two ears were retrospectively evaluated. The postoperative hearing

level of 30 dB and less was considered as successful hearing result postoperatively. Of the six ears which underwent canalplasty, three had achieved successful hearing result. However, there was no significant correlation between preoperative hearing level (HL) with HRCT score and postoperative HL with HRCT score at 0.05 significant levels (correlation coefficient = -0.292 , $P = 0.105$ and correlation coefficient = -0.127 , $P = 0.810$) respectively. Hearing evaluation and HRCT temporal bone are two independent evaluations for the patients with CAA before going for hearing reconstructive surgery.

Keywords High resolution computed tomography · Canal atresia · Stapes · Pure tone audiometry · Canalplasty

Introduction

Congenital aural atresia (CAA) is an abnormality in which the external auditory canal failed to develop [1]. It occurs in one of every 20,000 births [2]. Unilateral atresia is three times more common than bilateral atresia and right ear is more commonly affected than the left. CAA has male predominance. CAA commonly coexists with microtia. The embryologic precursors of the auricle are the axonal hillocks, a series of ectodermal elevations derived from the first and second branchial arches located along the rostral end of the branchial apparatus. The axonal hillocks fuse with each other around the primitive ear canal. Each of the six hillocks is responsible for forming a distinctive part of the auricle.

Early hearing assessment in children with CAA is very important in determining the hearing threshold. Bone conduction pure tone audiometry (BC PTA) and audiometry brainstem response (ABR) are routinely used [3]. An

A. Asma (✉) · A. Mazita
Department of Otorhinolaryngology Head and Neck Surgery,
Faculty of Medicine, Universiti Kebangsaan Malaysia, Jalan
Yaakub Latif, 56000 Cheras, Kuala Lumpur, Malaysia
e-mail: asmaent@yahoo.com

A. H. Hamzaini
Department of Radiology, Universiti Kebangsaan Malaysia,
Kuala Lumpur, Malaysia

A. W. Abdul Fatah
Department of Otorhinolaryngology, Sungai Buloh Hospital,
Sungai Buloh, Malaysia

ABR should be obtained for several reasons. The first is to establish the presence of a functional inner ear. In very young patients this is accomplished with multichannel air and bone conduction ABR. In cases of bilateral atresia, ABR will also allow us to determine the ear with greater cochlear function by examining the response obtained in wave I, which is indicative of ipsilateral cochlear status. The second reason for early ABR is that if an intact inner ear is present, the child can be fitted earlier with a bone-conducting device. A third reason for early ABR is to assess the functional status of the contralateral ear in unilateral atresia. The range of hearing loss is usually varies and based on degree of ossicular abnormality [4]. Conductive hearing loss is frequently observed secondary to bilateral atresia even sensory neural hearing loss (SNHL) has been reported.

Aguilar and Jahrsdoefer introduced grading system of candidacy for surgery for CAA patient based on HRCT findings [5]. Few parameters are considered and each of the parameter is scored with point (Table 1). Based on Jahrsdoefer scoring system, best result of corrective operation is when a score is 8 or more, fair chance for patient with score 7, marginal for score 6, poor candidate for hearing improvement for patient with score less than 6. A grade of 5 or less disqualifies the patients for surgery. A numeric of 8 signifies 80% chance of restoring hearing to normal or near normal level postoperatively.

High resolution computer tomography (HRCT) play central role in evaluating surgical candidates for correction surgery [6, 7]. Evaluation is done by overlapping CT sections of 1.5 mm thickness in coronal and axial projections. CT scan is useful in preoperative assessment by providing an information regarding status of atresia. With CT scan, an assessment of the ossicular chain, the nature of the atretic plate (whether bony or soft), the position and course of the facial nerve, the degree of external canal development and mastoid pneumatization is possible. The presence of cholesteatoma, which has a slight predilection for

stenotic ears, can also be ruled out. Current recommendations for the timing of CT scan is at approximately 4 years when mastoid pneumatization is most complete and allows adequate growth of temporal bone before surgery [3, 8].

In our centre, the size of the pneumatized middle ear cavity and the ossicles status based on HRCT temporal bone would determine patient's selection for canalplasty. Previously the candidacy of hearing reconstruction surgery was not based on Jahrsdoefer scoring system. Therefore we in this study we used Jahrsdoefer scoring system to provide a quantitative value in order to correlate with pre and post-operative hearing threshold.

Materials and Methods

All records and HRCT films with CAA from January 1997 until December 2007 were evaluated. The demographic data, operative records, pre and post operative hearing levels and HRCT findings were analyzed. Hearing level in this study is based on a pure tone average of air-bone gap at 500 Hz, 1 kHz and 2 kHz or hearing level obtained from ABR audiometry. HRCT findings were scored according to Jahrsdoefer scoring system by one senior radiologist in order to prevent interpersonal bias. Each patient Jahrsdoefer score was then compared with pre-operative hearing level, postoperative hearing level and air-bone gap. In this study, patients with acquired aural atresia, stenosis and congenital cholesteatoma were excluded in this study. All data were encoded and entered into the SPSS for analysis. Statistical analysis was performed using Pearson correlation study for relation between pre and postoperative hearing level and Jahrsdoefer score.

Result

Thirty-two ears of 26 patients who fulfilled the selection criteria were selected for this study. The patient's age at presentation ranged from 18 months to 30 years old with mean of 26 years. Other than CAA, there were patients who had other associated abnormality (Table 2). Majority of CAA patients (21 patients) had associated microtia. One patient had microtia with cleft lip and palate. One patient had microtia and maxillofacial defect. Table 3 shows an average of preoperative pure tone threshold and 18 ears had average preoperative hearing level which can be categorized as severe hearing loss. The best HRCT score according to Jahrsdoefer Scoring system was ten, which is maximum score whereas the worst score was three. Based on this score, four ears had a score of ten and six ears had a score of three (Table 4).

Table 1 Jahrsdoefer scoring system

Parameter	Points
Stapes present	2
Oval window open	1
Middle ear space	1
Facial nerve	1
Malleus—incus complex	1
Mastoid pneumatization	1
Incus—stapes connection	1
Round window	1
Appearance of external ear	1
Total	10

Table 2 Associated abnormality

Abnormality	Number of patient	%
CAA alone	3	11.5
CAA with microtia	21	80
CAA with microtia, cleft lip and palate	1	3.8
CAA with microtia and maxillofacial defect	1	3.8

Table 3 An average preoperative pure tone threshold of the ears with CAA

Hearing level (dB)	No. of ears	Percentage (100%)	Grading of hearing loss
50	1	3.1	Moderate
56.67	3	9.4	
60	6	18.8	
63.33	1	3.1	
63.34	1	3.1	
66.67	2	6.3	Severe
68.33	1	3.1	
70	8	25	
75	1	3.1	
76.67	3	9.4	
80	1	3.1	Profound
83.33	2	6.3	
85	1	3.1	
90	1	3.1	
Total	32	100	

Table 4 The patient's HRCT score based on Jahrsdoefer scoring system

HRCT score	No. ears	Percentage
3	6	18.8
4	2	6.3
5	1	3.1
6	1	3.1
7	4	12.5
8	5	15.6
9	9	28.1
10	4	12.5
Total	32	100

Table 5 shows the HRCT score with prediction of postoperative hearing result that could be achieved. If this scoring system is used to select the candidacy for corrective surgery, 18 ears would have best result if operated, four ears will have marginal result and one ear will have poor result. Nine ears will be disqualified due to the total score of five or less. Of 32 ears, only nine underwent

Table 5 The patient's HRCT score with prediction of corrective result based on Jahrsdoefer scoring system

Total score	Prediction of corrective result	No. of ears
≥8	Best result	18
7	Marginal result	4
6	Poor result	1
≤5	Disqualify from operation	9
Total		32

Table 6 Type of operations

Type of operation	No. of ears	%
No surgery perform	23	72
Canalplasty	6	19
Pinnaplasty	2	6
BAHA	1	3
Total	32	100

Table 7 Postoperative hearing level and number of the ear

Postoperative hearing level dB	No. of ear	Percentage
26.67	1	12.5
30	2	25
40	1	12.5
46.67	1	12.5
53.33	1	12.5
55	1	12.5
Total	7	100

Table 8 Pre and postoperative hearing level and HRCT score

Preoperative hearing level dB	Type of operation	Postoperative hearing level dB	HRCT score
70	Canalplasty	26.67	10
66.67	Canalplasty	30	9
76.67	BAHA	30	8
60	Canalplasty	40	9
70	Canalplasty	46.63	9
70	Canalplasty	53.33	10
63.33	Canalplasty	55	9

surgery and seven ears had surgery to improve hearing and two ears had pinnaplasty (Table 6). Three ears had postoperative average hearing threshold of 30 dB or less, which was considered a successful surgery (Table 7). Table 8 shows pre and postoperative hearing level and HRCT score. Statistical analysis using Pearson correlation study shows there was no significant correlation between preoperative hearing level and HRCT score at 0.05 significant

Table 9 Correlation of preoperative hearing level and HRCT score

	Pre operation hearing level	HRCT score
<i>Correlations</i>		
Pre operation hearing level		
Pearson correlation	1	−0.292
Sig. (2-tailed)		0.105
N	32	32
HRCT score		
Pearson correlation	−0.292	1
Sig. (2-tailed)	0.105	
N	32	32

Table 10 Correlation of postoperative hearing level and HRCT score

	HRCT score	Postoperative hearing level
<i>Correlation</i>		
HRCT score		
Pearson correlation	1	−0.127
Sig. (2-tailed)		0.810
N	31	6
Postoperative hearing level		
Pearson correlation	−0.127	1
Sig. (2-tailed)	0.810	
N	6	6

levels. (Correlation coefficient = -0.292 , $P = 0.105$) (Table 9). Statistical analysis using Pearson correlation study showed there was no significant correlation between postoperative hearing level and HRCT score at 0.05 significant levels. (Correlation coefficient = -0.127 , $P = 0.810$) (Table 10).

Discussion

Abnormal EAC can be classified into three types: almost normal, narrowing of the fibrocartilagenous canal and narrowing, tortuosity of both the fibrocartilagenous and bony part of the EAC [2]. CAA represents failure of embryological development. Together with microtia, it may occur as an isolated anomaly or may associate with other anomalies or as a part of other recognized syndrome. Those recognized syndromes which associated with CAA are hemifacial microsomia, Treacher Collins, Brachio-otorenal, Sticklers, Crouzons, Noonan syndrome, CHARGE syndrome (coloboma, heart defects, choanal atresia, growth retardation, genitourinary malformation and ear abnormality), Goldenhar's Klippel-Fiel and Pierre Robin. A diagnosed

patient with CAA warrants investigations to look for other anomalies and occasionally it requires genetic workups to rule out possible associated syndromes.

Early detection of hearing level must be established as early as few days of life by using auditory brainstem response audiometry. However, the assessment of auditory function in neonates and young children are limited by two factors. First, it is not possible to obtain valid pure tone speech audiometry. Second, the severe hearing impairment associated with CAA creates masking dilemmas [3]. If bilateral abnormal hearing is detected, the child should be fitted with bone conduction hearing aid before the age of 6 months of age to aid them in establishing hearing and normal speech development. In unilateral canal atresia with normal contralateral hearing, hearing aid is not indicated. The most important thing is to counsel the parents regarding the abnormality, the option of treatments available and the time of surgical intervention.

CAA affects males more than females [9]. However in our study, there were equal numbers of male and female: 13 female and 13 male. There were only two races found to have this congenital abnormality in this study. Majority of them were Malays, 18 patients, followed by Chinese, eight patients. There was no Indian population with CAA reported in our study. There were nine Malay patients with right atretic ear canal and three Malay patients with left atretic ear canal. Six of Malay patients had bilateral atretic ear canal. None of the Chinese had bilateral atretic ear canal.

The degree of deformities could be evaluated with HRCT which can be carried out at the age of 4–6 year old or even earlier if cholesteatoma is suspected. Presence of cholesteatoma warrants immediate surgery at any age. The recommended time of auricular and surgical correction for CAA is at 6 year old. Auricular reconstruction must be done prior to hearing reconstruction. The blood supply to the surrounding soft tissue will be compromised if hearing reconstruction is performed first. Surgical correction for CAA itself is one of the most complex and challenging forms of otological surgery. The surgical correction involves maintaining of a canal, removing an atretic plate, identifying and mobilizing the ossicular complex, establishing a stable vibratory and medialized tympanic membrane to provide hearing threshold better than 25 dB.

Jahrsdoefer has performed atresia surgery for the past 25 years. He has introduced HRCT scoring system for indication of surgery in CAA patients. Eighty percent of his patients who scored eight or more base on HRCT scoring will have hearing threshold better than 20 dB over 1, 2 and 4 kHz. Lambert in his series of 69 patients, only 30% of his patients who underwent reconstructive surgery had post-operative hearing improvement better than 20 dB [10]. De la Cruz and Teufert [11] established short term success in closing air-bone gap to less than 30 dB in 58% of 116 cases

underwent primary and revision atresia surgery but reduced to 50% with long term follow-up. Short term follow-up is when the period of follow-up is less than 6 months and long term follow-up is more than 6 months (up to 13.6 years). However, paired comparison analysis found no significant change in ABG from short to long term follow-up for either primary or revision cases.

In our series only seven ears had surgery to improve hearing. Three ears had postoperative average hearing threshold of 30 dB or less, which was considered a successful surgery. The best hearing level for the studied ears was 50 dB which is moderate hearing loss and the worst hearing was 90 dB which is categorized as profound hearing loss based on World Health Organization (WHO) grading of hearing impairment. Four ears had moderate hearing loss, 18 had severe hearing loss and four had profound hearing loss. Most of the studied ears had hearing level of 70 dB preoperatively. The mean of preoperative hearing level was 68.59 dB. Our mean postoperative hearing level was 40.24 ± 11.27 dB. Therefore we learnt from this study that proper selection of patients for canalplasty or BAHA are crucial.

There were soft tissue stenosis seen in 8% of primary surgeries and 3.4% of revision. Lateralization of tympanic membrane occurred in 3.4% of cases. Ossicular chain re-fixation occurred in 11.5 and 6.9% of primary and revision surgeries, respectively [11]. In our study, out of six operated ears, only one ear (16%) had developed stenosis of the ear canal. The patient underwent revision canalplasty to maintain the newly created ear canal.

Surgical reconstruction of CAA is easier when the EAC is present, because it acts as a landmark to help the surgeon to reach the tympanic cavity more safely. The presence of EAC also is closely related to the formation of the manubrium. The presence of the manubrium helps to reconstruct the tympanic membrane in its original positions [2]. There are several approaches in atresia surgery: Anterior approach, transmastoid approach and modified approach [10, 12]. Anterior approach is the most popular approach of the three main surgical approaches and it is popularized by Jahrsdoefer and Lambert [10, 13]. In this approach, surgery is performed after pinnaplasty and via post auricular incision. A temporalis fascia graft is harvested and the periosteum is incised at the posterior margin of the temporomandibular fossa and horizontally along the inferior margin of the temporal line. Drilling starts at the remnant of tympanic bone once it is identified and this will lead to atretic plate and middle meatus. Drilling along anterosuperior along the tegmen towards epitympanum is safe since the area is away from the facial nerve. Then the ossicles are visualized and if the ossicles are fixed or not in contact with stapes, they should be removed and replaced by sculptured bony columellar or partial ossicular replacement prosthesis (PORP).

The temporalis fascia graft is laid over the ossicular mass and part of bony meatus. The rest of newly created ear canal is laid with split skin graft which is usually harvested from the medial upper arm.

Whereas in transmastoid approach uses the durra of middle cranial fossa, sigmoid sinus and sinodural angle as landmarks to the antrum. Lateral semicircular canal, atretic plate and facial nerve if possible are identified. The atretic plate is removed to create a cavity on the stapes. The rest of surgery is completed in similar fashion as in anterior approach. A modified anterior approach is performed in patients with thick non-pneumatized atretic plates. In this approach, the sinodural angle is opened with limited dissection anteriorly to expose the posterior antrum identifying the lateral semicircular canal and ossicular mass. In our centre, anterior approach is the method of choice. It avoids opening of the mastoid air cells in order to prevent chronic discharging ear and for a better epithelization of newly created ear canal.

Bone anchored hearing aid (BAHA) is another option of hearing rehabilitation for patients with CAA who have good cochlear function with an unfavorable middle ear for reconstructive surgery. In this study we did not include BAHA as reconstructive ear surgery. BAHA offers a low risk, highly acceptable and predictable method of hearing rehabilitation. BAHA is better than conventional bone conduction hearing aid in terms of speech discrimination in noisy environment.

HRCT Score and Hearing Outcome

Jahrsdoefer scoring was used to score the studied ears. Based on the scoring system, the best score is ten and the worst score is three. Out of 32 ears, four had a score of ten and six ears had a score of three. Most of the studied ears had a score of nine. The mean of HRCT score was 7 ± 3 . If all ears were operated, 18 of them who had a score of eight or more will have a good result postoperatively, four ears with a score of seven will have fair result, one ear with a score of six will have marginal result and nine of them with a score of less than six will have poor postoperative result. Out of 32 ears, only six ears underwent canalplasty, one patient had BAHA and two ears had pinnaplasty for microtia. Other 23 ears did not undergo any form of corrective surgery. The youngest patients who underwent canalplasty were 5 years of age and the eldest was 30 years old. Two patients were operated at the age of 6 years old, three patients were operated at the age of 11 year old. The only patient underwent BAHA operation was operated at the age of 5 year old. Two patients underwent pinnaplasty at the age of 13 and 18 years old respectively. The mean age for canalplasty and BAHA was 11.43 ± 8.62 years.

As only 7 of 32 ears underwent surgical procedure for hearing improvement and therefore it was difficult to comment about the hearing outcome in relation to their HRCT score. Postoperative hearing level which could be achieved in this study was ranging from 26.67 to 55 dB (Table 7). Generally, all the ears which underwent hearing reconstructive surgery had an improvement of the hearing level postoperatively. Out of six ears which underwent canalplasty, only one ear had achieved postoperative reduction of air-bone gap of 26.67 dB from 70 dB air bone-gap. Two other ears which underwent canalplasty had hearing level of 30 dB postoperatively. According to the definition, canalplasty as a hearing reconstruction surgery is successful when the postoperative hearing level is 30 dB or less, so those three ears (37.5%) which were operated could be considered to have a successful hearing reconstruction surgery. The only one ear which underwent BAHA also had achieved a successful postoperative hearing level of 30 dB from 76.67 dB hearing level preoperatively.

Correlation Between Average Preoperative Hearing Threshold and HRCT Score

All 32 studied ears underwent HRCT temporal bone. The findings on the scan were scored by using Jahrsdoefer scoring system. Statistical analysis using Pearson correlation study showed there was no significant correlation between preoperative hearing level and HRCT score at 0.05 significant level (Correlation coefficient = -0.292 , $P = 0.105$) (Table 9). Based on this study, no prediction of the status of the middle ear could be made based on the hearing level which is established at preoperative evaluation.

Correlation Between Average Postoperative Threshold Level and HRCT Score

Based on statistical analysis using Pearson correlation study showed there was no significant correlation between preoperative hearing level and HRCT score at 0.05 significant levels (Correlation coefficient = 0.129 , $P = 0.782$) (Table 10). In this study we found that the scoring system cannot be used as a predictor for the outcome of the surgery even though in Jahrsdoefer series he had a successful reconstructive surgery for 80% of his patients who scored eight and more based on his scoring system.

Conclusion

This study clearly shows that the preoperative hearing level of patients with CAA unable to tell us the status of middle

ear. HRCT scoring system based on Jahrsdoefer scoring system is also unable to determine the outcome of the hearing reconstruction surgery. Therefore both audiology testing and radiology investigation are two independent evaluations before hearing reconstruction surgery.

Limitation

It is difficult to compare this result from various series because this retrospective study is limited by small number of cases since most of the old files and CT scan images were no longer available for review. Improper documentation in some of the traceable files also contributes further limitation in the study.

References

1. Kountakis SE, Helidonis E, Jahrsdoefer RA (1995) Microtia grade as an indicator of middle ear development in aural atresia. *Arch Otolaryngol Head Neck Surg* 121:885–886
2. Ishimoto S, Ito K, Yamasoba T, Kondo K, Karino S, Takegoshi H, Kaga K (2005) Correlation between microtia and temporal bone malformation evaluated using grading systems. *Arch Otolaryngol Head Neck Surg* 131:326–329
3. Jahrsdoefer RA, Yeakley JW, Hall JW, Robbins KT, Gray LC (1985) High resolution CT scanning and auditory brain stem response in congenital aural atresia: patient selection and surgical correlation. *Otorhinolaryngol Head Neck Surg* 93:292–298
4. Jahrsdoefer RA, Jacobson JT (1995) Treacher colins syndrome: otologic and auditory management. *J Am Acad Audiol* 6(1):93–102
5. Jahrsdoefer RA, Yeakley JW, Angular EA, Cole RR (1992) Grading system for the selection of patients with congenital aural atresia. *Am J Otol* 13(1):6–12
6. Gassner EA, Mallouhi A, Jaschke WR (2004) Preoperative evaluation of external auditory canal atresia on high resolution CT. *Ame J Radiol* 182(5):1305–1312
7. Swartz JD, Faerber EN (1985) Congenital malformation of the external ear and middle ear: high-resolution CT findings of surgical importance. *Ame J Radiol* 144:501–506
8. Yeakley JW, Jahrsdoefer RA (1996) CT evaluation of congenital aural atresia: what the radiologist and surgeon need to know. *J Comput Assist Tomogr* 20(5):724–731
9. Teufert KB, De la Cruz A (2004) Advances in congenital aural atresia surgery: effects on outcome. *Otorhinolaryngol Head Neck Surg* 131(3):263–270
10. Lambert PR (1998) Congenital aural atresia: stability of surgical results. *Laryngoscope* 108(12):1801–1805
11. De la Cruz A, Teufert KB (2003) Congenital aural atresia: long term results. *Otorhinolaryngol Head Neck Surg* 129(1):121–127
12. El-Hoshy Z, Abdel-Aziz M, Shabana M (2008) Congenital aural atresia: transmastoid approach; an old technique with good results. *Int J Pediatr Otorhinolaryngol* 72(7):1047–1052
13. Lambert PR (2006) Congenital aural atresia. In: Bailey BJ, Johnson JT (eds) *Head & neck surgery otolaryngology*, 4th edn. Lippincott Williams and Wilkins, Philadelphia, pp 2027–2040