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ORIGINAL ARTICLES

CONGENITAL MEATAL ATRESIA

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ABSTRACT

This interventional study was carried out in ENT Department of Combined Military Hospital Rawalpindi, a tertiary care hospital, from January 1993 to December 2000, to determine the sex incidence, laterality and type of congenital meatal atresia, and possible etiological factors; and secondly to determine the results of surgery in terms of hearing gain and incidence of complications while comparing trans-mastoid approach with the anterior approach. 42 patients of both sexes with congenital meatal atresia, up to the age of 25 years were selected by non-probability sampling. A detailed history and a detailed physical examination showed consanguinity as the biggest possible etiological factor (40.4%), male to female ratio of 2.5:1, unilateral to bilateral ratio of 1.6:1, type II atresia in 50%, type I atresia in 35.7% and type III atresia in 14.2%. X-ray mastoids and CT scan showed favourable anatomy in 36 patients who were selected for surgery. Canalplasty with tympanoplasty was carried out in them, via trans-mastoid approach in ten and direct anterior approach in 26 patients. Pre and post-operative Audiological Brainstem Response and Pure Tone Audiometry (over 4 years of age) showed successful surgery in 19 (52.7%) patients having post-operative hearing thresholds of up to 25-30 dB. Success rate was 61.3% with the anterior approach as compared to 30% with the trans-mastoid approach. Post-operative complications as re-stenosis (16.6%), otorrhoea (11.1%) and facial paralysis (2.7%) occurred in 11 (30.55%) patients. Complication rate was 50% with trans-mastoid approach and 27% with anterior approach. Consanguinity must be studied in greater detail as an important possible etiological factor.

Keywords: Congenital ear canal atresia, canalplasty, trans-mastoid

INTRODUCTION

Congenital Meatal Atresia is one of the frequently encountered congenital anomalies that cause not only cosmetic deformity but also the functional disability of hearing loss resulting in poor development of speech and communicative as well as educative and cognitive skills. It further results in psychological problems as well. It is therefore of immense importance to recognize this disability at an earlier stage for an early audiological rehabilitation so that these children can lead as normal life as possible with a minimum possible handicap [1].

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Congenital meatal atresia in association of varying degrees of pinna anomalies occurs as a result of failure of formation of the first and second branchial arch structures [2].

Mostly the etiology is unknown but aural malformations may occur as a result of factors like intrauterine infections, ischemic injury (hemi facial microsomia), toxin exposure (thalidomide, alcohol, phenytoin, tretinoin), radiation exposure or a positive family history. In only 5 to 10% of such patients the defect is inherited or recognized as part of a syndrome such as the CHARGE association, the VATER association, Goldenhar syndrome,

Treacher Collin's syndrome, Pierre Robin's syndrome etc [3].

Aural atresia occurs in approximately 1 in 20000 live births. Atresia is usually unilateral, but may be bilateral in 30% of the cases, occurring more commonly in the males and in the right ear [4]. Despite the fact that inner and middle ear develop separately, inner ear abnormalities coexist in 12 to 50% of the cases.

Aural atresia is classified into three types by Cremers, Oudenhover and Marres in a refined version of Marquet's classification [5]. In type I atresia (mild) the pinna is normal or near normal and the canal is stenosed and atretic laterally and cholesteatoma may develop in this stenosed canal. In type II atresia (medium) pinna is rudimentary, the canal is atretic completely and the facial nerve always adopts an abnormal course. This type is the commonest of the atresias occurring in more than half of the patients, and may be further subdivided into type II a and type II b. In type III (severe) the pinna is almost always absent, canal is atretic, tympanic cavity is small or absent and mastoid is not pneumatized [6].

Assessment is carried out as soon as the patient is brought in, to assess the severity, type and laterality of the hearing loss and to provide an early audiological stimulation [7]. An early screening Brainstem Evoked Auditory Response through a multi-channel device is the investigation of choice [8]. Conditioned Free Field Audiometry or Pure Tone Audiometry may help to determine the overall hearing levels.

Radiological assessment is carried out to find out the degree and nature of temporal bone deformity. Three dimensional CT Scan of temporal bone gives an accurate assessment of the degree of external canal development, the status of ossicles and tympanic cavity, the course of facial nerve, the pneumatization of mastoid and the distance between the glenoid fossa and the mastoid [9,10].

The patients with unilateral atresias usually do not require any early intervention till the consenting age of fifteen years [11]. But recently, early surgery at the age of 5-6 years is being advocated [12]. In cases with bilateral atresias, bone conducting or bone anchored appliances must be provided as early as possible for development of speech and communicating skills [13,14]. Surgery is usually carried out at the age of five to six years, as at this age a child is not only aware of this defect but is also exposed to teasing by his colleagues [15]. Moreover, by this age the rib cage of the child is grown enough to provide cartilage for reconstruction of the pinna in cases of associated anomalies of the pinna [16]. At this age atresia repair may be performed as stage III of the aural reconstruction [17]. However, in cases of non-availability of bone conducting appliances, surgical intervention for meatal atresia can be carried out at an age of eighteen months.

Selection of patients for surgery is made on the basis of a grading system devised by Jahrsdoerfer in 1992 [18,19]. Postoperative success has been defined in terms of postoperative speech reception threshold of 15 to 25 dB. Surgery is always carried out in the ear with good hearing to attain optimal results. Canaloplasty with homograft tympanoplasty is the operation of choice and may be undertaken by two approaches [20]; the conventional trans-mastoid approach starting with the identification of sinodural angle resulting in a canal wall down mastoidectomy and leaving an open cavity [21], and the direct anterior approach, a relatively newer approach described by Jahrsdoerfer, which involves drilling of bone posterior to the mandibular condyle [22]. Successful surgical outcome is found in 12-71% of cases where hearing thresholds of 25-30 dB or below have been achieved [23].

Complications of the surgery include facial nerve paralysis in 1-1.5% cases due to its aberrant course in 25-30% patients [24]. Sensorineural hearing loss occurs in up to

15% cases [25]. Postoperative complications like persistent otorrhoea and re-stenosis of the canal are fairly common, occurring in 8-50% cases [26].

Non-surgical treatment with bone conducting appliances or bone-anchored devices may be considered in those patients who are either unable or unwilling to undergo surgery.

The aim of this study was to determine the sex incidence, laterality and type of congenital meatal atresia and possible etiological factors and secondly to determine the results of surgery in terms of hearing gain and incidence of complications while comparing trans-mastoid approach with the anterior approach.

SUBJECTS AND METHODS

This interventional study was carried out in ENT Department of Combined Military Hospital Rawalpindi, a tertiary care center, from January 1993 to December 2000. All 42 patients of both sexes up to 25 years of age, having unilateral or bilateral congenital meatal atresia, who reported in ENT Department during this period, were selected with non-probability sampling, for the first part of the study. These patients underwent the following protocol:

A detailed history was taken including history of any drug intake, exposure to any toxin or febrile illness or invasive investigations as radiology and sonography during first trimester of pregnancy and family history including consanguinity and incidence of deafness and congenital anomalies in the family. Detailed ENT examination including head and neck examination was performed in each case, and severity and laterality of the abnormality was noted. Facial nerve was also examined in every case.

Audiological Brainstem Response in all the patients and Pure Tone Audiometry in patients above the age of 4 years were carried out. X-Ray mastoid and CT scan temporal bone were carried out to determine the degree of pneumatization of the temporal bone, the extent of stenosis, the condition of the middle ear cavity and the ossicles, the status of the bony inner ear and the course of the facial nerve.

36 of these patients with favorable anatomy of temporal bone and functional cochlea were selected for the next part of the study i.e. surgical treatment. Routine pre-operative investigations like Complete blood count, Urine routine examination and X-ray chest PA were carried out in them.

Surgical intervention in the form of canalplasty with tympanoplasty was performed in all the cases. Trans-mastoid approach was adopted in the first ten cases, while the rest twenty-six cases were operated upon while using the direct anterior approach. Results of surgical treatment were interpreted in terms of gain, no gain or deterioration in postoperative hearing thresholds, and successful surgery was considered in patients having post-operative hearing thresholds of 25-30 dB. Incidence of post-operative complications was also noted in each of the surgical techniques.

All the patients who underwent surgery were followed up on a regular basis. First follow up visit was performed one week after surgery. Subsequent visits were at 1, 3, 6 and 12 months after surgery.

STATISTICAL ANALYSIS

SPSS version 10.0 was to used to analyze the data. Percentages were calculated for sex incidence, laterality and type of congenital meatal atresia, and possible etiological factors. Chi-square test was used to compare the results of surgical outcome and incidence of complications in both the surgical approaches.

RESULTS

30 (71.4%) patients were males, while 12 (28.5%) patients were females. Thus the male to female ratio was 2.5:1. Twenty-six (61.9%) patients had unilateral atresia while 16

(38.1%) patients had bilateral meatal atresia. Thus Unilateral: Bilateral ratio was 1.6:1. Out of 26 patients with unilateral atresia, 15 patients (57.7%) had right side affected, while left sided atresia occurred in 11 patients (42.3%). Type I meatal atresia was present in 15 (35.7%) patients, while 21 (50.0%) patients had type II meatal atresia and 6 (14.2%) patients had type III meatal atresia.

Consanguinity, being present in 17 patients (40.4%), emerged as the commonest etiological factor. Febrile illness during pregnancy was present in 3 patients (8.3%), while history of drug intake during pregnancy was present in one patient (2.7%).

Pre-operative hearing thresholds between 40 and 70 db were found in 27 patients (64.3%), while 71-100 db in 13 patients (30.9%), and beyond 100 db in 2 patients (4.7%). Air-bone gap ranged between 20 to 40 db.

Surgery was carried out in 36 patients (85.7%) having Type I and Type II atresia. Canalplasty with temporalis fascia tympanoplasty was performed while adopting trans-mastoid approach in 10 patients (27.7%) and direct anterior approach in 26 patients (72.2%).

4 patients (11.1 %) with Type II atresia did not show any improvement in hearing and half of them showed worsened post-operative thresholds. 10-24 dB of improvement in hearing thresholds was seen in 13 patients (36.1%) with Type II atresia. 25-40 dB of improvement was seen in 14 patients (38.8%), of whom 10 patients had Type I atresia while 4 patients had Type II atresia. An improvement in hearing threshold of 41-55 dB was seen in 5 patients (13.8%), all having Type I atresia.

With these improvements in hearing, post-operative hearing thresholds between 25-30 dB (successful hearing) were obtained in 19 patients (52.7%) including all the 15 patients with type I atresia and 4 patients with type II atresia. These successful hearing thresholds were obtained in 16 patients (61.53%) with anterior approach as compared to 3 patients (30%) with trans-mastoid approach. Comparison of post-operative hearing gain between trans-mastoid and anterior approaches is given in table-1.

Complications: Re-stenosis occurred in 6 patients (16.6%), while otorrhoea occurred in 4 patients (11.1%), and facial paralysis occurred in only one patient (2.7%). This patient with post-operative facial paralysis was operated upon through trans-mastoid approach. Overall incidence of complications was found to be 50% (5/10) with trans-mastoid approach while it was 23% (6/26) with the anterior approach. Incidence of post-operative complications through both the approaches is compared in table-2 & 3.

In comparing successful post-operative hearing thresholds with both approaches we get non-significant results since p-value >0.05 using chi-square test. But in comparing incidence of complications in both approaches we get significant results since p-value <0.05 using chi-square test (table-3).

DISCUSSION

Congenital meatal atresia is an anomaly associated with varying degree of conductive hearing loss due to altered anatomy of the external and middle ear which is difficult to correct surgically. Appropriate selection of the patients is very important for successful surgical outcome of the condition [18].

In our study, the incidence of male to female ratio was 2.5:1, which is similar to what is given in the literature. Similarly, the ratio of unilateral to bilateral atresia was found to be similar to that described in the literature. Incidence of right-sided unilateral atresia was found higher as given in the literature [4]. Type II atresia was found to be the commonest type as described in the literature [6].

Consanguinity emerged as the commonest possible etiological factor,

probably because of far greater number of cousin marriages in our society, although it is not mentioned in the literature. Further studies should be carried out to study this etiological factor in greater detail.

Although it is a general consensus that bilateral atresias should not be operated upon before the age of 5-6 years [15], and unilateral atresias should not be operated upon before the consenting age of 15 years [11], these issues of timing of the surgery are far from resolved. In this study it has been found out that there are other factors as well, which may influence the timing of the surgery. In this study, four of the patients with unilateral atresia were operated upon before the consenting age due to persuasion of parents as these children with minor pinna anomalies and 'absence of hole' in their ears, were exposed to ridicule of their colleagues in school and were under severe psychological stress. Hence surgery was carried out in these patients to bring them and their parents from psychological stress by improving their physical appearance by 'making holes' in their ears. Thus cosmesis, besides psychological factors may be considered as an important factor for undertaking surgery.

Two of the patients with bilateral atresia were operated upon at the ages of 2 ½ years and 3 years respectively in contrary to the general consensus because of the financial reasons. Although it is generally agreed that surgery should not be carried out before the age of five to six years the topic still remains very controversial. Easy availability and tolerance of the bone conducting or bone anchored appliances is also a factor to be considered before management could be undertaken. In our set up, bone-conducting and bone-anchored hearing appliances are neither easily available, nor they can be afforded economically by a large majority of our patients as they cost over thirty thousand rupees. In contrast, surgery can be easily afforded by most of these patients, as our patients are entitled for free medical and surgical treatment. Hence financial factors have to be considered while planning the management policy.

Table-1: Post-operative hearing gain-trans-mastoid vs anterior approach (n= 36)

Post-operative Hearing Gain	Trans-mastoid Approach		Anterior Approach		P-value
	No. of Patients	%	No. of Patients	%	
No gain	2	20	2	7.7	0.87[NS]
10-24dB	5	50	8	30.7	
25-40dB	2	20	12	46.1	
41-55dB	1	10	4	15.3	
	n = 10		*n = 26*		

NS: Non-significant on chi-square test

Table-2: Types of post- operative complications—trans-mastoid approach vs anterior approach (n = 36)

Post-operative Complications	Trans-mastoid Approach		Anterior Approach	
	No. of Patients	%	No. of Patients	%
Re-stenosis	2	20%	4	15.3%
Otorrhoea	2	20%	2	7.7%
Facial Paralysis	1	10%	Nil	-
	n = 10		*n = 26*	

Table-3: Incidence of post-operative complications trans-mastoid approach vs anterior approach (n=36)

Post-operative Complications	Trans-mastoid Approach		Anterior Approach		P-value
	No. of Patients	%	No. of Patients	%	
Y	5	50	6	23	0.0002*
N	5	50	20	77	
	10		26		

**Significant on chi-square test*

Regarding post-operative hearing thresholds, it was seen that patients with type I atresia had better results as compared to patients with type II atresia. Probably it was due to a combination of more favorable anatomy and relatively better pre-operative hearing thresholds. Moreover, successful surgical outcome i.e. hearing thresholds between 25-30 dB in 52.7% patients, as obtained in this study was comparable with that given in the literature where it is described between 12-71% 23].

Similarly, the incidence of complications was also similar to what is described in the literature [26]. Facial paralysis occurred in only one patient (2.7%) who was operated upon through trans-mastoid approach.

Surgery was carried out via two surgical approaches i.e. Trans-mastoid and Anterior approach, and post-operative results of both approaches were compared. Surgical intervention showed better results with the direct anterior approach as compared to the trans-mastoid approach in terms of better hearing thresholds which is statistically insignificant and lesser complications, which is statistically significant.

CONCLUSION

Congenital meatal atresia is an anomaly that imparts great functional disability as well as cosmetic deformity in the affected child. The prime concern of the otologist is to restore function of hearing so as to enable the child to develop normal speech as well as to acquire binaural hearing.

In this study, consanguinity has emerged to be the greatest etiological factor; hence it needs to be investigated in more detail and marital counseling needs to be carried out in vulnerable cases.

Regarding indications of surgery, besides hearing gain, other factors as cosmetic and economic factors also need to be considered especially in our set up. Moreover, the timing of the surgery should also be flexible.

Further studies should be carried out with increased number of patients to get more accurate results.

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