Charakterystyka rehabilitacji funkcjonalnej dzieci z wadami wrodzonymi ucha

Features of functional rehabilitation of children with congenital malformations of ear

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Streszczenie

W pracy porównano charakterystyki kliniczne i diagnostyczne stu dzieci z wrodzonymi anomaliami ucha, atrezją i mikrocją. Podkreślono znaczenie tomografii komputerowej. Przedstawiono wyniki funkcjonalne i kosmetyczne leczenia operacyjnego. Autorka wskazuje na znaczenie wyboru właściwego rodzaju operacji. Poprawa słyszenia u dzieci z wadami wrodzonymi była możliwa jedynie w przypadkach ubytków słuchu z progami w zakresie 55-70 dB bez składowej odbiorczej. Pneumatyczna jama bębenkowa, dobrze zróżnicowane kosteczki, brak patologii ucha wewnętrznego, nerwu twarzowego oraz okienek okrągłego i owalnego mają też bardzo ważne znaczenie.

Słowa kluczowe: mikrocja, atrezja zewnętrznego kanału usznego, tomografia komputerowa kości skroniowej, prognozowanie wyników operacji.

Summary

The authors compared clinical and diagnostic characteristics of 100 children with congenital ear anomalies, atresia and mycrotia. Important role of computed tomography was stressed. Functional and cosmetic results of the surgical treatment were demonstrated. It was suggested that correct selection of surgery type is very important. Improvement of hearing in children with congenital disorders was possible only in cases with hearing thresholds of 55-70 dB without sensorineural component. Pneumatic tympanic cavity, well-differentiated ossicles, absence of pathology of the inner ear, facial nerve and oval and round windows is also very important.

Key words: microtia, atresia of external ear canal, CT of temporal bone, prognosis of surgery outcomes.

Introduction

The subject of the present research were congenital malformations of external and middle ear in combination with face malformations. Traditionally, they are related to syndromes, associated with malformations of the 1st and 2nd branchial arcs (hemifacial microsomia, Königsmark and Treacher Collins syndromes, etc.). Their typical features are narrowing or absence of external ear canal in combination with dysmorphological features of an auricle, i.e. microtia, anomaly of middle and inner ear, pathological changes of facial skeleton as a whole or isolated deformation of a mandible, abnormal development of cervical part of a spinal column etc. [Balysinskaya, Bogomilsky, Orchinnikov 1999; Swartz, Harnsberger 1998; Yeakley, Jahrsdoerfer 1996].

The urgency of a problem of rehabilitation of patients with microtia is stipulated not only by severity of the child's cosmetic defect, but also by marked loss of auditory and speech functions in bilateral abnormalities, which in combination with hereditary syndromes causes incapacitation of a patient.

Diagnosis

In addition to the general clinical inspection, pure tone audiometry or registration of the ABR thresholds and computed tomography (CT) of temporal bones were performed in all patients. A degree of pneumatization of mastoid, condition of auditory ossicles, facial nerve canal, configuration of labyrinth were evaluated

Material and methods

During a period of five years (1999-2004), we performed surgical treatment of 58 patients aged 4-16 years with microtia and atresia of external ear canal. According to classification of H. Weerda [Weerda, Siegert 1995], 4 patients had 1st degree of external and middle ear deformity, 5 patients had 2nd degree and 49 children had 3rd degree. Total number of observations was 72.

According to our data, the patients with microtia had predominantly the 3rd degree of hearing loss (46 cases). The cases with the 4th degree of hearing loss were met almost twice less frequently (22 observations). Two patients had bilateral deafness.

The first problem that we considered during the elaboration of the individual rehabilitational program for patients with microtia was decision about the appropriateness and terms of realization of functional surgery – meatotympanoplasty. In a number of cases, surgical formation of external ear canal was necessary for subsequent rehabilitation of the patients using hearing aids. The key factors in selecting patients for surgical treatment were the data of audiometry and CT of temporal bones [Yeakley 1996].

The analysis of 100 CTs of temporal bones of patients with microtia and atresia of external ear canal has allowed us to find the following features: absence of external ear canal – in all cases, reduction of the size of tympanic cavity – 27 observations, hypoplasia of head of a malleus – 62 cases (Fig. 1a), adhesion of a malleus to an incus or undifferentiated conglomerate – 48 cases and in 12 cases there was an absence of ossicular chain. In 26 cases, the size of antrum was reduced (Fig. 1b) and in 21 cases lack of its pneumatization was found.

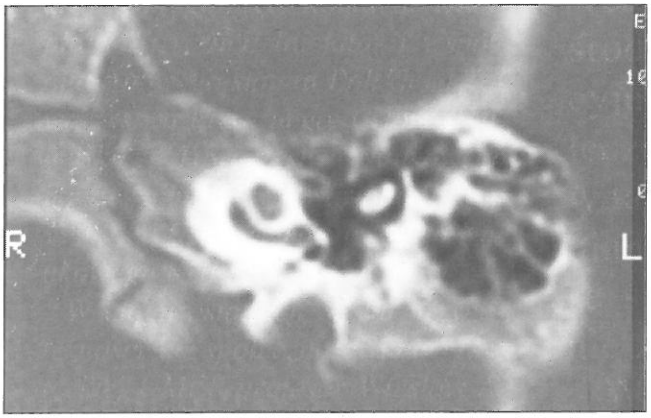

Fig. 1a. CT of patient K.: a hammer and dressing block are determined by a unified conglomeration

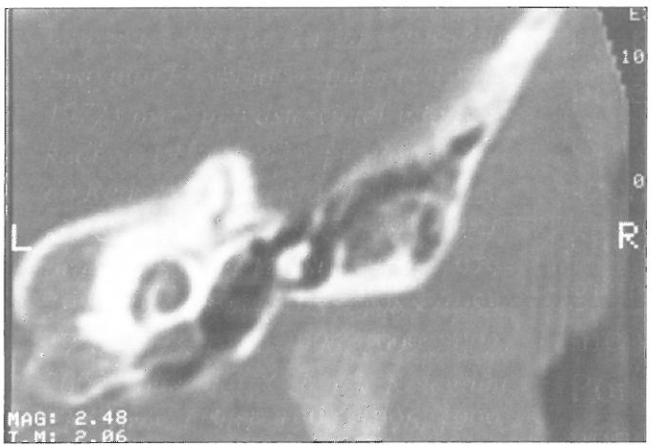

Fig. 1b. CT of patient L.: hypoplasia acoustical bones are determined directly above the head of a lower jaw.

In 45 our observations, the size of oval window was reduced or it was missing, round window was altered in 22 observations, anteposition of sigmoid sinus was found in 29 cases and high position of jugular bulb was observed in 1 case. The lumen of cochlea and semicircular channels was normal in 95 cases.

During surgical forming of external auditory canal and tympanic cavity, it is extremely necessary for the surgeon to know the features of position of facial nerve canal. In our research, disposition of the 3rd part of facial nerve canal was found in 42 observations (Fig. 1c) while in 12 cases separate parts or total facial nerve canal were absent. All above-listed features were confirmed by surgical findings.

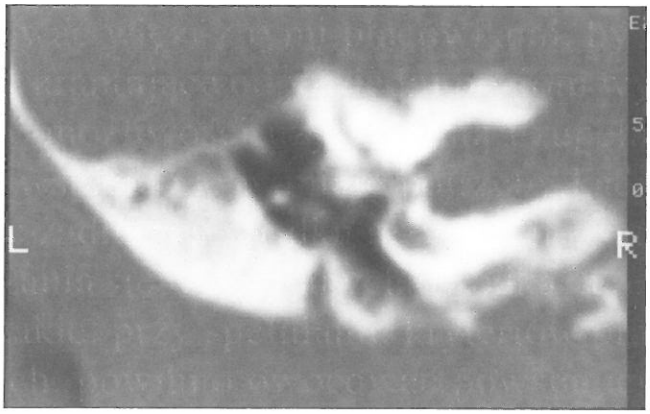

Fig. 1c. CT of patient K. it is determined the displacement of the III part of a facial nerve channel in front

Results

For systematical estimation of CT data of temporal bone in patients with atresia of external auditory canal (EAC) and elaboration of the universal approach to the prediction of the results of meatotympanoplasty we have proposed an universal table of the CT data scoring.

Admittedly less significant for surgery radiological signs were scored 0-1-2. We considered such signs to be the atresia of EAC, the degree of a pneumatization of mastoid, size of tympanic cavity, the state of malleus-incus and incus-stapes complex. In the case of absence of pathological changes, each sign was scored 2, in moderately expressed abnormality (small size or displacement) score was 1. Pronounced pathological changes of some of the listed signs were scored 0.

The state of oval and round windows, position of facial nerve canal, as well as sigmoid sine and jugular bulb are very significant for a solution to the question about advisability of meatotympanoplasty, and also for forecasting intra- and postoperative complications. Each of these signs was scored according to 4-point scale. Normal data corresponded to score 4, moderately expressed pathological changes to score 2 and dangerous to score 0. Thus, maximum score was 26. According to our experience, minimum acceptable score for meatotympanoplasty is 18 (Table 1).

Accordingly, 36 patients (41 cases) with microtia and EAC atresia got score 18 and more and meatotympanoplasty was performed in them. Surgery was performed in cases of bilateral microtia and bilateral atresia at the age of 4 years and older, thus giving our patients an opportunity of early auditory and speech rehabilitation. In unilateral microtia meatotympanoplasty was performed after 7 years of age.

In patients with EAC atresia, the decrease of thresholds after surgical treatment by 20 dB and more was considered as good outcome and in our observations was found in 29 cases.

During the estimation of the efficiency of meatotympanoplasty we also considered the degree of EAC epidermi-

sation, maintenance of its width and depth as well as physiological position of rudimental lobule.

Tab. 1. CT data (congenital malformation of a temporal bone) Patient K.

CT-signs	Score	Right ear	Left ear
Atresia of EAC	2/1/0	0	0
Pneumatization of mastoid	2/1/0	1	1
Size of tympanic cavity	2/1/0	1	1
Malleus-incus complex	2/1/0	0	1
Incus-stapes joint	2/1/0	0	1
Stapes, niche of oval window	4/2/0	0	0
Round window	4/2/0	4	4
Facial nerve canal	4/2/0	4	0
Sigmoid sine, jugular bulb	4/2/0	2	4
Total score	26	12	12

Score 0 – severe degree of pathology
Score 1 to 2 – mild degree of pathology
Score 2 to 4 – norm

Accordingly, in our study good results were obtained in 29 cases, satisfactory – after 8 surgeries and in 4 cases the outcomes were considered unsatisfactory (in 2 patients there was no complete epidermisation of EAC 1 year after surgery and in 1 patient with bilateral pathology the depth of formed EAC was less than 5 mm).

Discussion

We performed the analysis of probable reasons of unsatisfactory meatotympanoplasty results, which demonstrated that in all these cases the forming of osseous ear canal was unsufficiently deep, *aditus ad antrum* and incus were not found, plasty of EAC with a skin flap was not performed.

At the same time, the simple creation of EAC 1.5-2.0-cm deep in patients with bilateral congenital microtia and severe hearing loss was one of the tasks of rehabilitation of 6 patients, who used bone-conduction body-worn hearing aids preoperatively. After surgery, these patients could use in-the-ear or behind-the-ear hearing aids.

One of our observations is presented below.

Patient K., 14 years old, was in St. Vladimir Children Hospital from March 3, 1998 till March 27, 1998. Diagnosis: bilateral microtia of 3rd degree, bilateral atresia of EAC, conductive hearing loss of 4th degree, Treacher Collins syndrome.

Inspection: bilateral hypoplasia of mandible branches and chin, antimongoloid slant. Left ear – the rudimentary auricle displaced forward, missed EAC. Right ear – postoperative scars in EAC location, and the rudiment of an auricle was located behind the scars (from the anamnesis, it is known that at the age of 7 meatotympanoplasty was performed on the right side).

The patient used body-worn hearing aid with bone receiver (K10K).

CT of temporal bones demonstrated following features: right side – bone canal was filled with soft-tissue component from cortical layer to aditus, the ossicles missed, internal ear structures and facial nerve canal had usual location; left side – the EAC missed, hypoplasia of malleus and incus was found, dystopia of the 3rd part of the facial nerve canal to forward was revealed, oval window was thickened (Fig. 1c).

Using our scoring of CT data, the results on both sides were scored 12 (see Table 1), which corresponded to high risk of performance of meatotympanoplasty on any side.

However, taking into account severe bilateral hearing loss, meatotympanoplasty was performed on March 6, 1999 on the right ear with the purpose of subsequent in-the-ear hearing aid fitting. During the surgery, new EAC was formed, the scar tissue was excised, and the upper foot seed grafting was done.

In postoperative period the size of EAC has remained sufficient (2.0 – 1.3 cm) and at present patient can use in-the-canal hearing aid. Subsequently, this patient underwent staged auriculoplasty with silicone framework as well as correction of chin with silicone.

After solving the question of functional rehabilitation of patients with microtia it was necessary to perform cosmetic reconstruction of the auricle. At present we use different materials and techniques for this purpose, which will be described in subsequent papers.

Conclusions

It can be stated that congenital microtia with atresia of EAC accompanied, according to the results of CT of temporal bones, with a lesion of the ossicles, anomaly of the stapes and the facial nerve canal is unpromising for surgical correction of auditory function. Functional effect depends on pre-operative thresholds and CT score exceeding 18.

In unilateral microtia, in the case of assumed low efficiency of the surgery according to audiological data and CT score less then 18, we recommend to limit the treatment to cosmetic surgery.

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