International Journal of Pediatric Otorhinolaryngology 102 (2017) 174–179

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: http://www.ijporlonline.com/

Check-list for the assessment of functional impairment in children with congenital aural atresia

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ARTICLE INFO

Article history:
Received 26 July 2017
Received in revised form
22 September 2017
Accepted 22 September 2017
Available online 28 September 2017

Keywords:
Congenital aural atresia
Oculo-auriculo-vertebral spectrum
Goldenhar syndrome
Threacher-collins syndrome
Feeding abilities
Speech and language pathology

ABSTRACT

Objectives: Congenital Aural Atresia (CAA) is a deformity of the external ear and it is commonly associated with malformations of middle and inner ear and, in some cases, with other facial deformities. Very few assessment measures exist for evaluating the functional impairment in children with CAA. Purpose of this study is to introduce and describe an assessment Checklist, (nominated FOS Checklist) that covers feeding abilities (F), oralmotor skills (O), communication/language development (S) in children with CAA.

FOS wants to offer a range of assessment providing a profile of the child in comparison to hearing peers and it aims to make clinicians able to identify additional problems and areas of difficulties as well as specific abilities and skills.

Secondary, we want to investigate the presence of correlations between disorders and side of CAA.

Methods: a new Checklist (FOS Checklist) was administered to 68 children with CAA.

Results: Feeding abilities are age-adequate in 94,3% of all patients.

54,4% of all patients are in need for further assessment of their oral-motor skills; delays in language development were found in 44,1% of cases. Orofacial development delays have been observed in 57.2% of subjects among the bilateral CAA group, in 53.9% among the right CAA group and in 53.4% among the left CAA group.

Patients referred for further language evaluation were 42,9% in the bilateral CAA group, 33.3% in the right CAA group and 33.3% in the left CAA group.

According to the χ^2 analysis, referral for further assessment is independent from side of aural atresia.

Conclusions: Subjects with bilateral CAA are more likely to be referred for further assessment, both for oral motor aspects and for speech perception and language development. However, there is not a significant statistical difference between the performances of children with bilateral or unilateral CAA.

FOS Checklist is simple, reliable and time effective and can be used in everyday clinical practice. FOS enable clinicians to identify additional problems and areas of difficulties as well as specific abilities and skills; moreover, FOS allows to determine appropriate referrals and intervention strategies.

1. Introduction

Congenital Aural Atresia (CAA) is a disease constituted by partial or complete lack of development of the external auditory canal, which is commonly associated with conductive hearing loss. Microtia and atresia are rare deformities of the external ear (incidence 1:10000–20000 live births) and atresia is reported in 55%–93% of individuals with microtia [1].

Most cases of atresia/microtia are unilateral (79–93% of cases), with a right-sided predominance (60%), but the abnormality can affect both sides with different degrees [2].

CAA is, in most patients, an isolated condition. In some patients, however, the ear deformity occurs in conjunction with other facial deformities, such as maxillary and mandibular abnormalities.

These deformities can be considered as part of the Oculo-auriculo-vertebral spectrum (OAVS) (http://www.omim.org/), a rare heterogeneous congenital condition in which the head structures, originating from the first and second branchial arches, are incompletely developed on one (85% of cases) or both sides [3].

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https://doi.org/10.1016/j.ijporl.2017.09.024
0165-5876/

Commonly, the disease is characterized by ear and jaw abnormalities (hemifacial microsomia). In most severe cases, the eye or spine are also affected: this condition is known as Goldenhar syndrome. The phenotype in OAVS and Goldenhar syndrome is variable and associated clinical features can involve cardiac, renal, skeletal and central nervous systems.

In addition to this condition, others congenital syndromes occur due to defects involving the ear and structures deriving from first and second branchial arches, including the following: Pierre-Robin Syndrome, Treacher-Collins Syndrome and DiGeorge Syndrome.

Regarding the audiological situation, CAA is especially associated with a conductive hearing loss. However, approximately 10–15% of cases might include a sensorineural hearing loss (SNHL) [4].

The threshold of the conductive hearing loss on the affected side is estimated to be between 40 and 60 dB HL. Patients who have unilateral CAA usually have normal hearing in the contralateral ear; in patients with bilateral atresia, subjects have bilateral conductive hearing loss and early hearing amplification is required to restore the good hearing threshold over the long term.

Different studies show that patients affected by CAA might have an increased risk of speech and language delays; moreover, the associated facial malformations might cause oral myofunctional disorders. It is unclear whether the side might influence development of speech and language [5].

In literature, there is a lack of assessment measures capable of characterizing the complex profile of individuals with CAA. Indeed, we believe it was necessary to create it.

The purpose of a good Checklist is to identify those who require further assessment through the analysis of children's performance compared the hearing peers' development.

The aim of present study is to present the newly developed Checklist and to collect the profiles of a group of children with CAA. In addition, we want to verify the presence of correlations between disorders and bilateral or unilateral CAA.

2. Design and methods

2.1. Subjects

Patients affected by CAA, who presented from April 2016 to April 2017 to the multidisciplinary craniofacial clinic of ENT Clinic of University Hospital of Padua (Italy), were examined in agreement with the Italian privacy and sensible data laws (D.Lgs 196/03) and to the ENT Clinic of University Hospital of Padua inside rules.

The local institutional ethics committee approved the study. All experiments were conducted with the informed and overt consent of each participant or their caregiver, in accordance with the Code of Ethics of the World Medical Association (Declaration of Helsinki) and the standards established by the local Institutional Review Board.

Sixty-eight patients (mean age 5 years 10 months; age range 8 months–14 years and 8 months; 27 females) affected by CAA were included in the present study (Table 1).

CAA was unilateral in fifty-four patients, with a predilection for the right side (39 subjects) and 72.22% of patients with unilateral CAA were right-sided. All patients demonstrate complete atresia of the external auditory canal regardless of the grade of microtia (I-II-III).

Thirty-nine patients are affected on the right side, 15 on the left side and 14 showed a bilateral CAA. Two patients had a diagnosis of Goldenhar Syndrome, while three children had a diagnosis of Treacher-Collins syndrome; in the other cases, CAA is described as part of Oculo-Auriculo-Vertebral Spectrum.

All subjects demonstrated a PTA (Pure Tone Average) around 60 dB HL (mean PTA 64.67 dB nHL) in the affected ear and about 20 dB HL (normal hearing; mean 19.69 dB nHL) in the unaffected side. In particular, most of the subjects show a conductive hearing loss (bone conduction threshold: mean 16.29 dB nHL); only two patients demonstrate evidence of a mixed hearing loss and one shows moderate sensorineural hearing loss. Exclusion of this three patients did not substantively change statistics in any category, and they where included so as to reflect the overall CAA population.

All subjects with bilateral CAA (15) have been aided before the age of 6 months. In particular, two patients in bilateral group wore air conduction hearing aids (because they had microtia with partial atresia) and the remaining had bone conduction devices. Only three patients with unilateral malformation received bone conduction hearing aids. In addiction one patient with left atresia wore an air conduction device on contralateral side (right moderate neurosensorial hearing loss).

In patients with device FOS Checklist was administered with hearing aids on.

2.2. FOS checklist

FOS is an acronym for Feeding abilities, Oral-motor skills and Speech perception and language abilities (FOS).

FOS Checklist is a collection of measures which have been chosen to meet the need of identification of difficulties in children with Congenital Aural malformations.

FOS offers a range of assessment providing a profile of the child in comparison to hearing peers. The Checklist focuses on the main areas of development, which can be modified because of the malformation: feeding, oralmotor skills, hearing. The development of audition is not viewed in isolation, but in its role in the development of communication and language by the child.

The Checklist is particularly useful in providing a semi-structured way of looking at development before the period when most standardized tests of linguistic skills can be used. Indeed, some of the measures can be used from early infancy right through to adulthood, providing continuity and the basis for long-term comparisons in the various areas of development. FOS provides useful markers in that early period, guiding decision making about appropriate intervention and providing a tool for continuing assessment and monitoring.

Table 1
Demographic and Audiometric Characteristics of the sample.

	Right CAA (n = 39)	Left CAA (n = 15)	Bilateral CAA (n = 14)
Male:female	24:15	11:4	6:8
Mean age (range), years	5.7 (1–13)	5.1 (1–14)	7.6 (0.8–15)
Etiology (OAVS: Gold: Tr. Col.)	33:2:1	15:0:0	12:0:2
Pure Tone Average dB nHL (range)	66.13 (28.75–100)	60.83 (41.25–77.5)	62.55 (42.5–86.25)

Abbreviations:
CAA, congenital aural atresia; OAVS: oculo-auriculo-vertebral spectrum; Gold: Goldenhar syndrome; Tr.Col.: Treacher Collins syndrome.