CONGENITAL MALFORMATIONS OF THE EAR

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Malformations of the external ear can involve orientation, position, size and relief pattern of the pinna, anotia may also occur. To make a classification of ear malformations must be considered: size of auricle, shape of the ear and position of the ear. Aim of this retrospective study is to assess the clinical outcome in a series of patients affected by ear defects and discusses the pertinent literature. In the period between January 2001 and December 2010, 35 patients underwent to surgical correction of external ear malformations at the Pediatric Surgery Unit, S Anna Hospital, Ferrara, Italy. Patients included 23 (66%) females and 12 (34%) males. Age ranged from 1 months to 14.5 years with a mean value of 2.6 years at the time of admission. All cases were surgically corrected under general anesthesia. Successful correction of prominent ears and of all others kind of ear malformations requires a precise understanding of the normal anatomy and relationships of the external ear with the face. The surgeon should be familiar with the normal anatomy of the cartilaginous skeleton and the soft tissue features of the external ear. Abnormalities in ear dimensions will reveal themselves at an early age. The rapid development of the ear to approximately 90% of adult dimensions by age 3 years allows for early surgical intervention for auricular anomalies. Many surgeons recommend performing surgical correction of prominent ears when children are aged 3 to 6 years, before the start of school. The goal is to minimize the malformation before the period of socialization to avoid ridicule by other children.

The external ear consists in auricle and external acoustic meatus. The auricle arises from a series of elevations termed "auricular hillocks" around the first pharyngeal cleft. Three elevations on the first pharyngeal arch (i.e. the mandibular arch) and three elevations on the second pharyngeal arch are seen at six weeks. The hillocks are transitory and soon lose their identity to contribute to the various part of the external ear. Early first auricle is placed ventro-laterally but at the end it assumes a dorso-lateral position (1).

Malformations of the external ear can involve orientation, position, size and relief pattern of the pinna, anotia may also occur. Ear malformations may have a genetic acquired background (1). In numerous investigations, especially in studies of inner ear development, various genes as transcription factors, secretion factors, growth factors and cell adhesion proteins have been identified as being responsible (2, 3). However, in all genetically determined malformations one can assume a high frequency of spontaneous genetic mutation. Among the congenital malformations about 30% are associated with syndromes involving additional malformations and functional losses of organs and organ systems. Examples are Threacher-Collins Syndrome or Crouzon Syndrome (2).

To make a classification of ear malformations must be considered:

- Size of auricle: Microtia - small ears commonly associated with hemifacial microsomia; Macrotia - large ear, the auricle is usually very large but very well shaped without malformations; Anotia - absent pinna with an artistic ear canal; most form are associated with

Key words: Ear, skin, cartilage, anomaly, malformation, defect.

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0393-974X (2012)

conductive hearing loss.

- Shape of the ear: Cup shaped - small auricle that grows forward over the meatus; Lop ear - the external ear stands away from the head at a greater angle; Dysplastic ear - abnormally shaped auricles. Usually, associated with chromosomal anomalies; Elfin ears – with sharp form.
- Position of the ear: Melotia - ear located at the cheek due to lack of the midline; Low set ear - ears are very close to each other to the middle.

A newborn with auricle deformities must undergo under detailed investigation of the craniofacial structures. Middle ears should be particularly closely investigated because the development of the external ear usually correlates with that of the middle ear (4).

Over two hundred different techniques have been described to correct these deformities. Many of these techniques have proven successful in their ability to achieve high patient satisfaction, despite the significant variations in these techniques. From this perspective, the otoplasty represents a privileged procedure allowing the surgeon great latitude in his approach and ability to achieve patient satisfaction (5).

Aim of this retrospective study is to assess the clinical outcome in a series of patients affected by ear defects and discusses the pertinent literature.

MATERIALS, METHODS AND RESULTS

In the period between January 2001 and December 2010, 35 patients underwent to surgical correction of external ear malformations at the Pediatric Surgery Unit, S Anna Hospital, Ferrara, Italy.

Patients

Patients included 23 (66%) females and 12 (34%) males. Age ranged from 1 months to 14.5 years with a mean value of 2.6 years at the time of admission.

Treatment

All cases were surgically corrected under general anesthesia.

DISCUSSION

Newborn ear deformations vary in both type and severity along a wide anatomic spectrum. They can produce significant psychological stress including social withdrawal, depression, and/or lack of companionship in the maturing child, motivating both the parents and the patients to seek correction (6, 7).

Newborn ear deformations occur commonly and have been documented with incidence ranging from 15% to as high as 30% of the United States population (8). This may translate into nearly a quarter of a million affected newborns for the year 2011. Deformations range phenotypically and include: lop, stahl, constriction, cupping, prominent ear, helical abnormality and non-typical deformations. Ear deformations, unlike malformations such as microtia, contain all major anatomic components of the adult ear but may be arranged in apparently abnormal and/or non-aesthetic configurations (9). This division between deformation and malformation recalls the embryologic origins of the ear. The ear derives from a fusion of the 3 anterior and 3 posterior hillocks of the first (mandibular) and second (hyoid) brachial arches, respectively, gaining

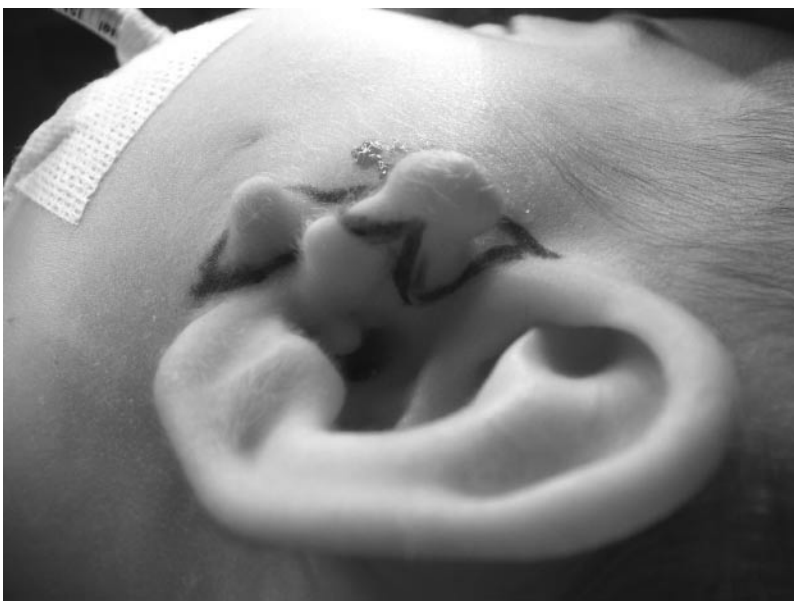

Fig. 1. *Abnormally shaped auricle*

much of its appearance by week 8 of gestation (10).

Impedances in embryological auricular development is thought to be due to stapedial artery malformation and/or impaired flow, producing microtia (ears with missing parts)(9). However, the underlying causation of auricular deformations (ears with all, but odd parts) is not yet fully understood (10).

Successful correction of prominent ears and of all others kind of ear malformations requires a precise understanding of the normal anatomy and relationships of the external ear with the face. In some instances, significant differences exist among different races and the values cited can be considered generalities (11).

The surgeon should be familiar with the normal anatomy of the cartilaginous skeleton and the soft tissue features of the external ear. The ear occupies a zone from the brow superiorly to the base of the columella inferiorly. The base of the tragus begins approximately a single ear length, lateral to the lateral orbital rim. The average length of the adult ear varies from 5.5 to 6.5 cm. The width of the ear is approximately 50% to 60% of the length. The vertical axis is inclined 15 to 30° degrees posterior (7).

Characteristics of cartilage should also be considered: Gibson and Davis (12) originally described the principle of cartilage bending away from cut surface. Stenstrom (11) has eloquently described the biological principles of cartilage scoring. He showed that: 1) auricular cartilage bends away from the scored surface; 2) bending properties are intrinsic to the cartilage and are independent of the perichondrium; 3) deeper furrowing results in more profound bending.

Abnormalities in ear dimensions will reveal themselves at an early age. The rapid development of the ear to approximately 90% of adult dimensions by age 3 years allows for early surgical intervention for auricular anomalies. Many surgeons recommend performing surgical correction of prominent ears when children are aged 3 to 6 years, before the start of school. The goal is to minimize the malformation before the period of socialization to avoid ridicule by other children (13).

Autogenous costal cartilage is still considered as an ideal material for framework fabrication in microtia reconstruction. Many surgeons have adopted the Nagata approach, the Brent approach or variations of the two in their work. With these employed techniques, auricles reconstructed by experienced surgeons have proven to be aesthetically promising. However, with regards to the harvesting of the costal cartilage, the underdevelopment of the chest wall donor site, alopecia of the scalp, and scarring of the postauricular-mastoid region are still considered problematic aspects of these approaches. It is generally accepted that prominent ears should be corrected through a combination of sculpting and suture techniques, according to the individual shape and the quality of the ear prominence. Moreover, most of the cryptotia malformations show not only embedded upper auricles, but also associated adhesions of the upper auricular cartilage. So, their correction should therefore resolve both deformities (14).

From the clinical evaluation and anatomical basis, to the formulation of surgical strategy, correction of the malformed ear continues to be one of the most complex and debatable topics in reconstructive surgery (15).

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