Question mark ear deformity: a case study and surgical technique

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INTRODUCTION

The ear is a crucial facial feature that significantly contributes to one’s appearance, yet few studies have assessed ear aesthetics. It has been said that “the main job of an ear is to sit on the side of the head and not draw attention to itself.” Ears rarely attract attention when they are normal [1]. However, an ear deformity can significantly disfigure an individual’s appearance.

External ear anomalies are diagnosed through clinical examination, taking into account the normal anatomy of the ear. The Cosman ear, also known as the question mark ear, is a congenital auricular cleft between the fifth and sixth hillocks. It was initially described by Vincent et al. [2] in association with urogenital anomalies. Subsequently, Cosman [3] reported two similar cases, noting the resemblance to a question mark. This deformity is primarily observed as a pathological feature of auriculo-condylar syndrome. However, there are also reports of sporadic isolated cases and some instances of familial inheritance without any syndromic association. The deformity can appear as either unilateral or bilateral in sporadic cases, while inherited cases typically present as bilateral deformities. The Cosman ear is characterized by six distinct features, which vary in severity. The primary characteristic of this anomaly is the constriction of the lower auricle at the junction of the lobule and the helix. In severe cases, the ear lobule may be completely detached from the helix; in other instances, there may be only a mild constriction. There can also be transposition of the lobule and antihelix in severe cases. The upper helix might be prominent, and the ear may project anteriorly. The development of the antihelix can be either incomplete or complete. A unique sign of this anomaly is a postauricular tag. Additionally, there is often deformed development of the superior third, which alters the scaphoid fossa and the superior crus [4].

In auricular procedures, surgeons primarily strive to achieve a structure that integrates seamlessly with the patient’s overall appearance, avoiding any features that might attract undue attention. Here, we present our surgical experience in correcting a moderate-severity question mark ear deformity.

CASE REPORT

A 12-year-old boy with a bilateral congenital ear deformity presented to our institute. He was the third child of non-consanguineous parents. There was no significant antenatal history or family history of craniofacial or ear anomalies. The child was first seen at our outpatient department at the age of 6 months for an assessment of a cranial deformity associated with ear malformation. A three-dimensional maxillofacial computed tomography scan revealed mild metopic craniosynostosis. Given the mild nature of the cranial deformity, surgical reconstruction was not considered necessary. The child also had...
Fig. 1. The picture illustrates cleft of the auricle. There is marked helix deficiency and helix is superiorly rotated. There is loss of antihelix definition. There is excess scapha and concha.

Fig. 2. Incision was made along the red line. Elliptical outer skin and cartilage excision of scapha was done (X). Full thickness excision of area marked Y was done, and excised part was discarded. Scapha and antihelix were dissected from inner skin and exposed posteriorly. Stitch was done to fold the antihelix and enhance its definition (arrows). Antihelix was sutured to supra-tragus, by connecting points [d]&[c] to their correspondent points. Helix was advanced down and sutured in continuity with lobe by connecting points a&b to their correspondents.

bilateral ear deformities, for which surgical correction was postponed until preschool age. However, they lost follow-up and returned at the age of 12 after the child, who had been subjected to bullying at school, requested it and stopped his academic year due to social impairment. On examination, each ear exhibited a cleft of the auricle. The helix was deficient and superiorly rotated. There was a loss of antihelix definition, excess scapha and concha, and the lobe was also superiorly rotated. Both the tragus and antitragus were normal. The deformity is shown in Fig. 1. Hearing tests and otoscopic examinations were normal. The patient and his family had a planning meet-

Fig. 3. Illustrates left ear after incision. Scapha reduction was done with elliptical excision of outer skin and cartilage. Full-thickness wedge excision of concha was done above tragus to help advancing helix and antihelix.

Fig. 4. Scapha and antihelix were dissected off the inner skin. A stitch was applied to fold the antihelix and improve its definition.
Surgical technique
Surgery was planned to correct the middle-third deficiency and the upper prominence, as shown in Figs. 2-6. The procedure was performed under general anesthesia. After infiltrating the area with local anesthesia containing adrenaline, markings were made (Fig. 2). Incisions were made along the lower border of the helix and part of the scapha; the outer skin in these areas was cut and discarded. Additionally, a wedge excision of the concha was performed. A triangular incision was made through the pinna, with the excised part shown in Fig. 3. Following the guidelines in Fig. 2, a single suture was used to correct the antihelix fold. The overlying skin was approximated using 5/0 polydioxanone sutures (Fig. 4). The antihelix and the antitragus were approximated (Fig. 5). The helix and the pinna were also approximated. To correct the ear’s prominence, a posterior stitch was applied. The same technique was applied to the left ear. The final result of the left ear is displayed in Fig. 6. After achieving hemostasis, a mastoid dressing was applied to the ear. The patient was kept in the dressing for one week and followed up in the outpatient department. The wound healed without any complications, and the corrected auricles now have a similar shape and size. The patient is satisfied and can now attend school without any concerns.

DISCUSSION
The ear is an important aesthetic feature of the face [5]. Embryologically, it develops from six swellings at the dorsal end of the first and second branchial arches, known as auricular hillocks. Ear deformities may present with various types of clefts between different structures. A cleft in the lower third of the ear results from a fusion arrest between the fifth and sixth hillocks, a condition known as Cosman ear or question mark ear. The severity of a Cosman ear can vary widely, and it is believed that the timing of the fusion arrest determines the severity of the cleft. This condition is more prevalent in boys than in girls, with a ratio of 2 to 1. Congenital ear anomalies occur in 0.8 to 2.4 out of every 10,000 live births and are more common among individuals of Asian and Hispanic descent, with the incidence potentially reaching as high as 55% in the overall population [6]. Ear deformities can have a significant psychosocial impact on children and their parents [7]. A questionnaire-based study was conducted to evaluate the psychosocial morbidity associated with severe congenital and acquired auricular deformities. The survey assessed psychosocial indicators such as anxiety, depression, lack of confidence, embarrassment, and others. The most prominent feelings among the participants were lack of confidence, dissatisfaction, and embarrassment. Depression was present in 55% of the individuals, and anxiety was identified in 52% of the cases. The patients highlighted that a significant portion of their psychosocial challenges stemmed from peer bullying, which explains their strong motivation for surgery [8]. Sixty-four percent of children were predominantly motivated by psychosocial reasons, 28% by concerns about appearance, and 8% by parents worried about the future welfare of their child [9].

The correction of ear anomalies is one of the few widely accepted surgical procedures performed on children for aesthetic reasons. It has been shown to significantly improve self-esteem and happiness [5]. A retrospective questionnaire sent to 101 children revealed that 97% reported an increase in happiness, 92% an increase in self-confi-
dence, 79% an improved social experience, and 100% reported reduced or ceased bullying. The appropriate age for ear correction surgery remains a topic of debate; however, it is generally delayed until after the age of 6 years, when most auricular growth has occurred [9].

There are multiple reconstructive principles for correcting a question mark ear that surgeons must address: one is to restore the normal dimensions and mass of the lobe. Additionally, it is crucial to ensure that the free margin of the lobe is undivided, smooth, and seamlessly continues with the face. For these reasons, when selecting a reconstruction method, the severity of the tissue defect must be considered first [10]. Various techniques have been described for correcting Cosman ear. Currently, there is no standard technique that suits all the different characteristics of Cosman ear. For instance, Vayvada et al. [11] described the reconstruction of the cleft using an onlay conchal cartilage graft, covered with a postauricular local cutaneous flap. Fujiwara repaired a cleft earlobe using the Tennison-Randall triangular flap technique, traditionally used for cleft lip repair [12]. Other techniques include opposing z-plasty [6], V-Y advancement [10], and costochondral graft [13].

Most patients with question mark deformity exhibit ear prominence, which is ideally corrected concurrently with cleft repair. Some techniques may lead to a reduction in ear size; therefore, they may not be appropriate for cases of unilateral Cosman ear, as they could result in asymmetrical ear sizes. In our case, cutaneous and chondro-cutaneous advancement flaps were used to reorient the helix/antihelix and to close the cleft. Additionally, elliptical and wedge resections were employed for the reduction of the scapha and concha.

Using our method, we successfully treated both the protrusion of the upper ear and the cleft, achieving similarly satisfactory results for both ears. The technique described herein is suitable for clefts with or without associated prominence or size discrepancy. When utilizing a local flap, we ensure a dependable blood supply and achieve an excellent color match. This technique is a very effective and simple method, resulting in superior cosmetic outcomes without any post-operative notching. Additionally, no early complications were observed.

In conclusion, we present a rare case of bilateral question mark ears, along with our method for correcting this deformity. Our approach involves using cutaneous and chondro-cutaneous advancement flaps to address both the upper ear prominence and the ear cleft, resulting in satisfactory outcomes.

NOTES

Conflict of interest
No potential conflict of interest relevant to this article was reported.

Patient consent
Parents or legal guardians provided written consent prior to the inclusion of the patient in this report. Additional consent was obtained for the use of his images.

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REFERENCES