Bilateral Oblique Facial Clefts and Amniotic Bands: A Report of Two Cases

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The oblique facial cleft, amniotic bands (called also annular bands), and intrauterine amputations are rarely seen and belong in the category of medical oddities. For this reason, these two very unusual cases are presented together.

Bilateral Oblique Facial Clefts

According to Gorlin and Pindborg (*3*) the oblique facial cleft (meloschisis) is exceedingly rare, and when it does appear, the occurrence is more frequently bilateral. Like the lateral facial cleft, no hereditary tendency has been demonstrated, and when it occurs unilaterally, the left side is more frequently involved.

The oblique facial cleft is the result of incomplete fusion of the median nasal, lateral nasal, and maxillary processes. Such fusion occurs below the nostrils. When the oblique cleft extends beyond the nostrils and into the eye, it may have traumatic origin. It is thought that amniotic adhesions to the face of the developing embryo might produce tears which have a variable relationship to the nasolacrimal duct (*4*).

The oblique facial cleft does not follow a definite pattern although there is usually associated cleft lip and palate. The nose may or may not be involved and there may be extension of the cleft through the eye. The facial cleft may be associated with encephalocele, hydrocephaly, involvement of the nasolacrimal duct, lower lid coloboma, other facial clefts, amniotic bands, and deformities of the extremities.

Case I

T.J. was a white female patient who expired of bronchopneumonia at the age of four years. At birth, after a full term, she weighed 6 pounds 4½ ounces. There are three normal siblings in her family. Her body was fairly well developed and appeared to be well nourished. Her heart, lungs, abdomen, and genitalia were

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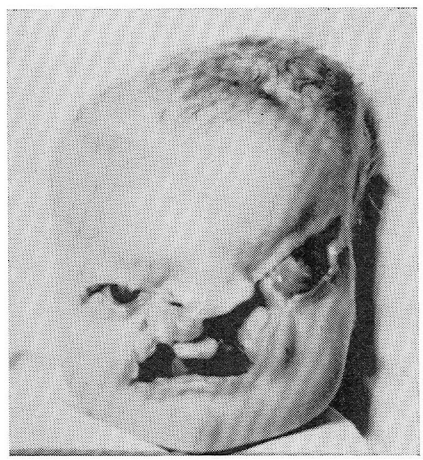


FIGURE 1. Face of the patient at birth showing hydrocephaly and bilateral oblique clefts extending into the eyes.

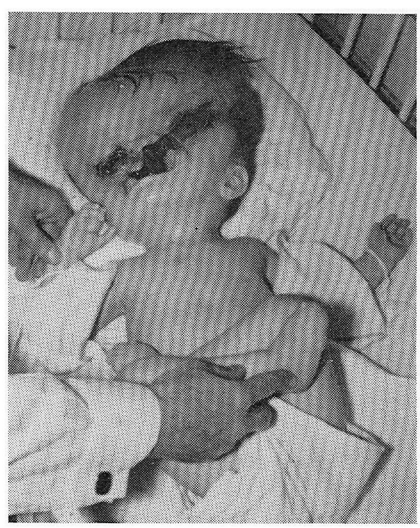


FIGURE 2. Same patient approximately two years of age. Note the left nasolacrimal gland, the premaxilla with deciduous central incisors, and deformities of the extremities.

normal in development and function. Her first teeth appeared at eight months. She was severely mentally retarded and was never able to walk or talk.

There were extensive deformities of the face and cranium including hydrocephaly (Figures 1 and 2). The anterior fontanel remained open and in the region of the posterior fontanel of the distorted cranium there was an opening of more than four inches in diameter.

The palatal processes were absent, creating a wide cleft of the palate associated with bilateral clefts of the alveolar process. The prolabium was missing and the premaxilla was exposed. The nose was incompletely developed. The cleft on the left side extended superiorly beyond the eye to the brow. Both eyes were exposed

due to the absent lids, and the corneas gradually became opaque in spite of protective medicaments. The nasal turbinates and the left lacrimal gland could be seen through the clefts.

The extremities also were malformed. All the digits were grossly deformed, with the exception of the left little finger and the right thumb. There was syndactylism of the right hand. The fingers of the left hand appeared to be constricted at the distal ends and the thumb was underdeveloped. The constrictions of the fingers of the left hand resembled cases seen in ainhum or intrauterine amputation. The right foot showed adactyly and the toes of the other foot were malformed.

Amniotic Bands with Congenital Amputations

Amniotic bands and adhesions (sometimes referred to as Streeter's Bands) have often been associated with facial clefts and the phenomenon of intrauterine amputation. According to Streeter (*6*), adhesions of the amnion to the surface of the embryo have never been observed in normal material and there is no evidence that intrauterine amputation is due to amniotic bands or to adhesions or to other mechanical constrictions. Streeter's theory is based on 'focal deficiencies of foetal tissue'.

Bragg (*1*) believes that congenital amputation is germinal in origin and consists of pathological break down of the affected tissues early in uterine life and is characterized by localized hemorrhage.

Amputations in ainhum bear a striking resemblance to the later phases of intrauterine amputations (*5, 6*). As the following case illustrates, the occurrence of intrauterine amputation may be erroneously attributed to strangulation by the umbilical cord.

Case II

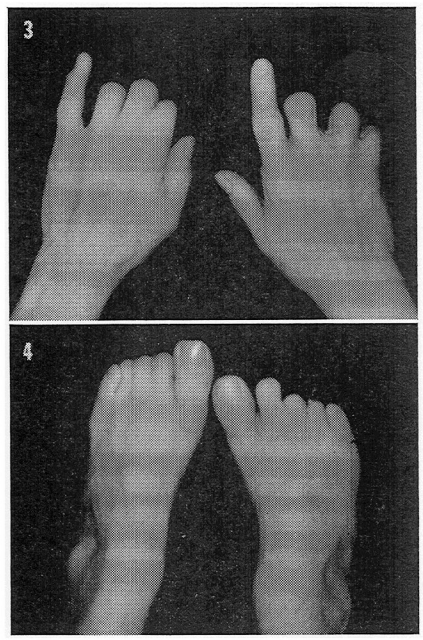
W.D. is a 43-year-old white female. She has two younger siblings. She was delivered prematurely after seven months and weighed 2½ pounds. She has been diagnosed as having congenital, nonspecific cerebral maldevelopment. However, she managed to complete the eighth grade.

There is no family history of mental retardation. She has an illegitimate child who developed normally. She is also handicapped with grand mal seizures. Her maternal uncle also is afflicted with epilepsy.

She was born with bilateral club feet, which required surgical correction. The patient demonstrates complete syndactylism of 2, 3, and 4 on the left foot and incomplete syndactylism of 2, 3, and 4 on the right. The hands present partial amputation of 3, 4, and 5 of the right hand and 2, 3, and 4 on the left with syndactylism of an incomplete nature (Figures 3 and 4).

Her medical record indicates that she was born with a peculiar discolored streak around her head because the umbilical cord had almost strangulated her at birth. (See figures 5 and 6 for reference to the marking.) This is not reasonable since strangulation or amputation by the umbilical cord requires that the tissue proximal to the constriction be normal and the tissue distal to it show signs of disturbance in circulation with necrosis. In addition, most fetuses so afflicted are macerated and retained several weeks after death, before being expelled (*6*).

This peculiar indentation which completely encircles the head is believed to be the result of an amniotic band (*2*). Many times, macerated sheets of epidermis and strands of hyalinized fibrous tissue, which are residue of the localized areas of defective tissue, have been mistaken for amniotic bands (*6*).



FIGURES 3 and 4. Congenital amputations of digits of hands and feet.

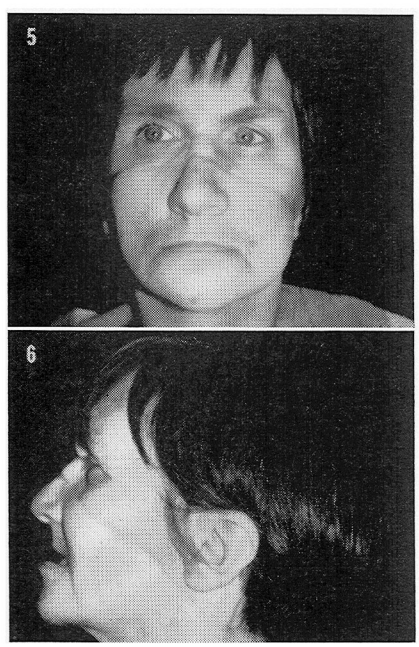


FIGURE 5. Frontal view of same patient showing the scar from an amniotic band crossing over the bridge of her nose.

FIGURE 6. Profile view showing continuation of band around the head.

The patient is completely edentulous, and clinical and radiographic examination of the soft and hard oral tissues reveals normal findings.

Summary

An unusual case of bilateral oblique facial clefts with associated deformities, and a case with amniotic bands and congenital amputations have been presented. Amniotic or annular bands are frequently reported in association with facial clefts and intrauterine amputations, but there is much controversy concerning the disposing factors. These two cases are in the realm of medical oddities.

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