Conjoined Twins with Mirror-Image Clefts of Lip and Palate

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Siamese or conjoined twins, a rare phenomenon, have stimulated the interest of laymen and scientists for centuries. Such twins are non-separated monozygotic twins and are considered to be the product of a single fertilized ovum. Many authorities consider this type of twins "double monsters".

The etiology of conjoined twins is not fully understood. Genetic factors are recognized as causative in the occurrence of monozygotic twins, but there is some evidence that they may occur as a result of the embryonic environmental factors. Some animal experiments have shown that retardation of growth during the early developmental stages of an embryo can be produced by reducing the temperature or the oxygen concentration in its environment (*2, 9*). This retardation may cause normal twins, conjoined twins or a single monster. Consequently, it is possible that growth retardation in the human embryo may produce conjoined twins. Gedda considers that double malformations arise from a delayed and incomplete division of a twin embryo at the time when only one system of membranes has been formed (*4*).

The twins may be joined at different body parts (thoracopagus, craniopagus, omphalopagus, pygopagus, and ischiopagus) and the degree of joining may vary greatly. They are usually joined by the same parts of the body or sometimes by the mirror-image parts. In case of asymmetrical conjoined twins, one twin is normal and the other is incomplete and attached to the normal one as a parasite.

Various estimations of the general incidence of conjoined twins have been published. Such figures, for newborns, range from 1:6,000 to 1:82,000 (*1*). Dinner (*2*) is of the opinion that the incidence is 1:60,000 births and Santon (*12*) gives the figure of 2:220,000 for Australia. These statistical evaluations are of a relative value bacause the data from many Asiatic countries, where the majority of conjoined twins are born, are not available.

The most common type of conjoined twins is thoracopagus where the

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twins are joined in the sternal region. Of 117 analyzed cases, Robertson (*11*) found that 73% were thoracopagus, 19% were pygopagus, and 2% were craniopagus.

In Yugoslavia, a country of 20 million people and a birth rate of approximately 420,000 per year, two pairs of conjoined twins were born within a period of seven months. The first pair, which was thoracopagus, was born in July, 1967, and the second, of the craniopagus type, in February, 1968. This report is concerned only with the first pair. The special interest of this twin pair is the presence of clefts of the lip and palate in both members.

A number of cases of mono- and dizygotic twins with the same or different types of clefts have been presented but the only record of a cleft in conjoined twins is that reported by Robertson and McKenzie (*11*). In their case, one member had a unilateral cleft of the lip while the other had no cleft condition but only a preauricular tag of skin.

The first pair of Yugoslav conjoined twins was born with mirror-image clefts of the lip and palate. Therefore it was thought that this rare phenomenon would be of interest to all those concerned with cleft lip and palate.

Case Report

History of the Case. The female twins were born from the first pregnancy by Cesarean section at a small provincial hospital. They were immediately transferred to the Institute for Child and Mother Health of Serbia for further care and eventual surgical separation. The twins were placed in an incubator. Under the direction of a specialized medical team, radiological, biochemical, genetic and other investigations were undertaken. Photographs of the twins and impressions of their upper jaws were obtained. The twins died at three months of age due to a respiratory infection and after that an autopsy and thorough investigation was carried out.

There was no history of clefts or other congenital anomalies in either parent's family, but on the mother's side there was a history of twins. The mother, age 21, had a full term pregnancy and did not suffer any disease during the pregnancy.

General Description of the Twins. The twins were joined from the manubrium of the sternum to two centimeters below the umbilicus (Figure 1). They shared a single umbilicus and a common umbilical cord. Each twin had her own normal external sexual organs. Deformities of legs and hands were not found. The position of the heads was such that the twins faced each other. There was no deformity of external ears present.

The autopsy revealed a common fused heart and liver as well. Severe abnormalities of the great blood vessels were also present. Each member had a single kidney and ureter. On the basis of these findings it was concluded that the surgical separation of the twins would be impossible.

Description of the Orofacial Region. Both twins presented complete clefts of the primary and secondary palate, on the left side in twin A and on the right side in twin B (Figure 1). This type of similarity in identical twins is known as "mirror-image similarity". Although they both had the same morphological type of cleft, it was noticeable that they did not have the same type of maxillary arch deformity. Twin A had a small underdeveloped maxillary segment on the cleft side which was rotated to the median line and overlapped by the larger maxillary segment of the noncleft side (Figure 2). The proximal part of the larger segment was slightly deviated mesially and towards the noncleft side. In twin B, the maxillary segment on the cleft side was better developed than in twin A and almost in a normal position within the arch. The anterior portion of the larger segment was severely deviated to the noncleft side and the gap between the proximal parts was three times wider than in twin A (Figure 2).

Discussion

Twins with clefts are of special interest to the biological scientists. Both monozygotic and dizygotic twin pairs have been reported to the present time with either a heterogeneous or a homogeneous cleft condition (*3, 6, 7*). The frequency of similarity and dissimilarity of cleft types within monozygotic and dizygotic twin groups may be of etiologic significance. In addition, monozygotic twin pairs, in which only one member presents a cleft condition, provide a unique opportunity for the comparative study of growth and development.

The study of conjoined twins may provide additional embryologic and genetic information. A survey of the literature on conjoined twins revealed approximately thirty well-documented reports. Only one case of conjoined twins with defects of the first arch has been reported by Robertson and McKenzie (*11*).

It is well known that monozygotic twins sometimes show mirror-image similarity. Kraus and Oka (*5*) even described a pair of monozygotic separated twin fetuses of 20 weeks gestational age showing mirror-image clefts of lip and palate. In his twin collection, Pruzansky has two pairs of monozygotic separated twins with mirror-image unilateral clefts of the primary and secondary palate (Figure 3). This phenomenon is considered to be the result of a relatively late separation of the embryos after some asymmetry has developed (*9*). The mirror-image similarity is a confirmatory sign of monozygosity (*8*). It occurs more often in conjoined twins than in monozygotic separated twins (*9, 10*). If we assume this, then it follows that the mirror-image clefts in this report are genetic in nature. However, the history of this case reveals a pedigree free of clefts or other congenital abnormalities. Another possible explanation is that the mirror-image clefts may have been caused by some kind of

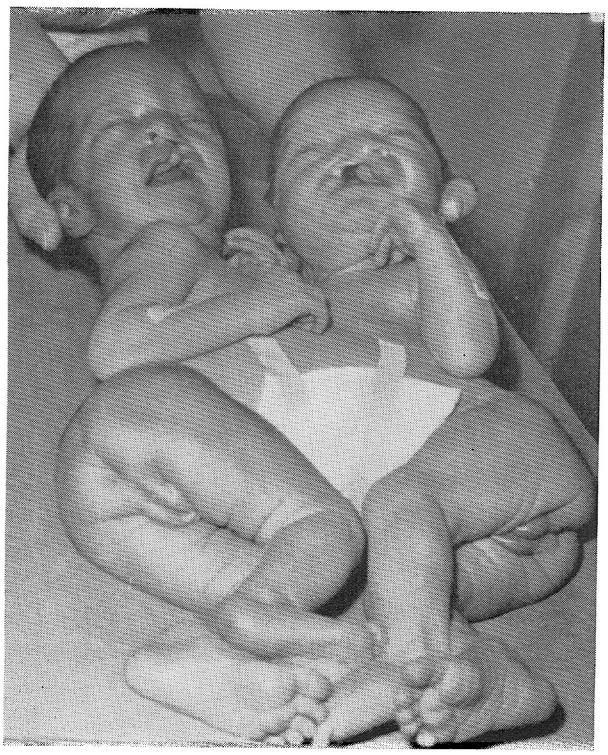

FIGURE 1. Complete unilateral mirror-image clefts in conjoined twins: twin A, left and twin B, right.

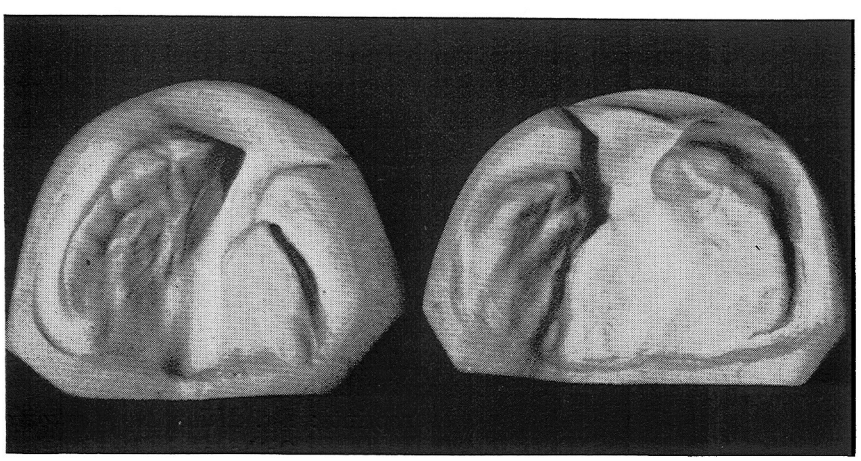

FIGURE 2. Plaster models of the upper jaw showing different arch deformities in the twins.

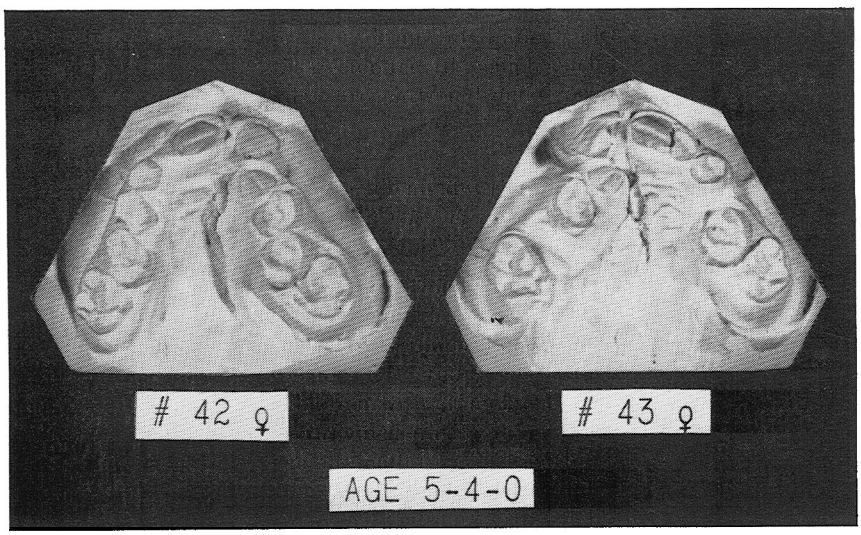

FIGURE 3. Mirror-image unilateral clefts in separated monozygotic twins (by courtesy of Dr. S. Pruzansky).

chromosomal mutation, but chromosomal studies failed to support this view. A third explanation would relate to intrauterine environmental influences. As conjoined twins have identical genotypes, they are more likely to react similarly to the same environmental factors than twins with dissimilar genotypes.

The dissimilar maxillary arch deformities found in these twins may be the result of different tongue positions during both pre- and postnatal life. It was observed that twin B positioned her tongue between the maxillary segments for the duration of her postnatal life. One may assume that this position was maintained prenatally as well. This tongue position may have contributed to the increased intersegmental distance which was three times larger than in twin A. On the other hand, the absence of the tongue between maxillary segments in twin A may have contributed to the arch collapse observed in this twin. This possible influence of the tongue emphasizes the importance of nongenetic factors.

Summary

A case of thoracopagus conjoined twins with mirror-image clefts of lip and palate is described. The etiology of the condition and the factors contributing to the dissimilar arch deformities present are discussed.

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