Clinical Conference

Anencephaly and Unilateral Cleft Lip and Palate in Conjoined Twins

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Twin girls attached along the thorax and upper abdomen to the level of the umbilicus and possessing apparently one anencephalic head were born to a 28-year-old Puerto Rican woman at 33 weeks of gestation. Four previous pregnancies had ended in three normal deliveries and one spontaneous abortion. No drugs were taken during this pregnancy which proceeded normally until 32 weeks of gestation when she was admitted to the University of Illinois Hospital for investigation of right-sided abdominal pain without bleeding or leakage of amniotic fluid. The uterus was somewhat larger than might be expected for the calculated stage of pregnancy. An abdominal radiograph showed that there was no fetal head, a diagnosis that was confirmed by sonography. Labor started spontaneously one week later. Her membranes were ruptured artificially yielding 4000 ml. of greenish-yellow amniotic fluid. Conjoined twins were delivered by the vertex 17 hours later without difficulty. A heart beat was present immediately after delivery but there were no respiratory efforts. The solitary placenta was described as normal and was discarded.

The appearance of the infants from the front and back is shown in Figure 1. Together they weighed 1210 gm and measured 30.0 cm from crown to heel and 16.0 cm from crown to rump. Both upper and lower extremities, the posterior aspect of both bodies and the anterior aspect below the common umbilicus including the perineum and external genitalia were normally formed and appropriate for the gestational age of the infants. Symmetry of the free and fused components of the twins was a striking feature. Anteriorly, there was a short neck leading into a common thorax with two prominent pectoralis major muscles attached to a midline sternum. The upper extremities were separated from the chest wall by normal axillae. Because of slight rotation of the bodies, the anterior chest wall was broader than the posterior. The rotation was less marked below the costal margins so that the fused anterior abdominal wall was of about the same extent as seen from before or behind. Two arteries and one vein were identified in the single, midline, umbilical cord.

The head, which seemed to be single on first impression, possessed some features of doubling. There were two prominent eyes more or less closed by puffy, edematous eyelids and two flattened, misshapen ears as is usual in uncomplicated anencephaly. A single nose had two nostrils. There was a complete right unilateral cleft lip and palate (Figure 2). The left nostril was complete and communicated with a nasal passage separated from the oropharynx by an intact hard palate. There was a high, midline ridge in the floor of the mouth with two equal parts of a double tongue on either side of this structure. The exact nature of the cranial anomaly will require a detailed dissection of the soft tissue of this region and will be reported later.

As viewed from above, the scalp, calvaria, and brain were absent, the basicranium being covered by a thin layer of vascular tissue, the so-called area cerebrovasculosa of anencephaly (Figure 3). However, there were two foramina magna leading to two spinal columns, the foramina being separated by a prominent bony mass. The basicranium, although highly anomalous, was a single structure with a single pituitary fossa containing two pituitary glands.

Three other cases of anencephaly in twins

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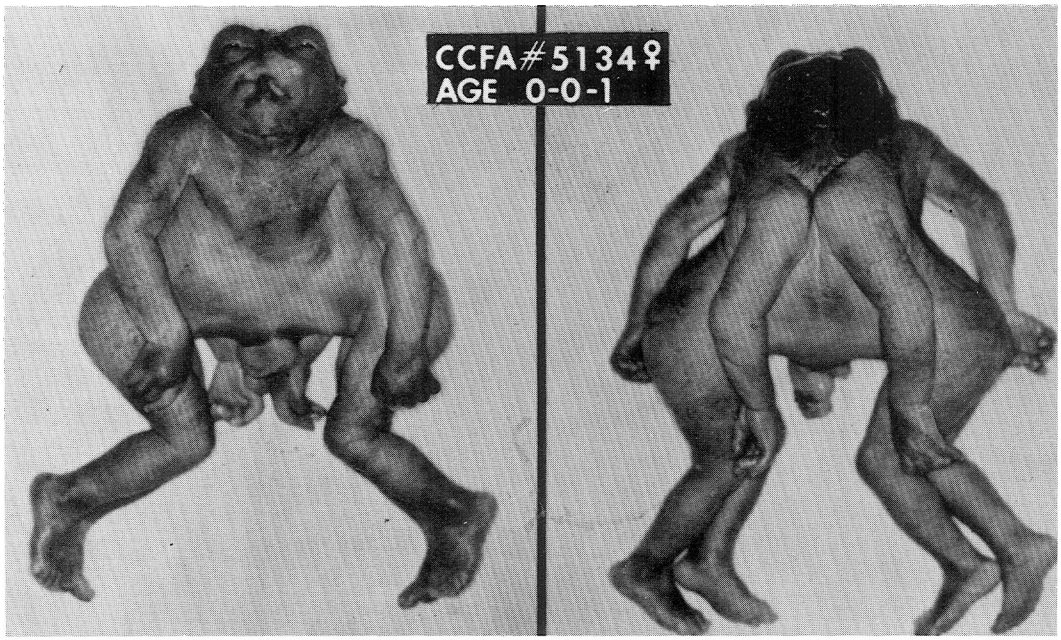

FIGURE 1. (CCFA#5134) Ventral and dorsal views of cephalothoracopagus female twins with right unilateral cleft lip and palate and anencephaly.

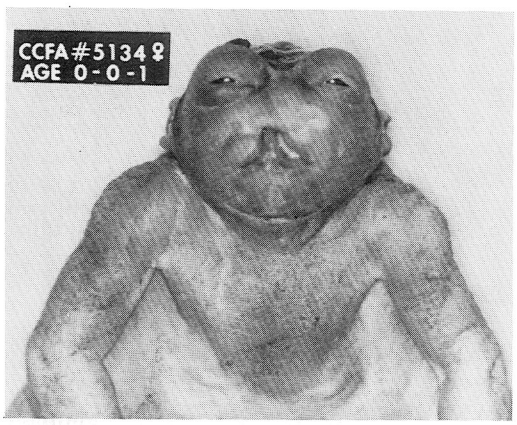

FIGURE 2. Close-up of facies.

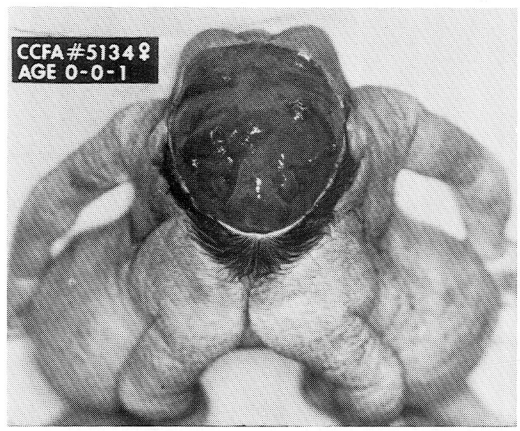

FIGURE 3. As viewed from above, the scalp, calvaria, and brain are absent. The basicranium is covered by a thin layer of vascular tissue, the area cerebrovasculosa of anencephaly.

joined at the head and chest (cephalothoracopagus) have been reported. One pair resembled our infants in that there was a double cleft lip and palate and some suggestion of duplication of the lower jaws (Buchta, 1973). Another pair had a single head but two mouths (O'Toole, 1976), and a third had a single head without doubling but with fusion of the posterior arms (Schwalbe, 1906). The similarity between the cases is quite striking. Furthermore, three out of four have been described in the last four years. Possibly, conjoined twinning may be becoming more common if the recent experience in South Africa (Nelson, Bhettay, and Beighton, 1976) and Atlanta, Georgia, (Hanson, 1975) is repeated elsewhere. If this is the case, the more unusual types of conjoined twins with their variants may be seen more frequently too.

Facial clefting occurs more commonly in thoracopagus twins and in anencephalic singletons than might be expected by chance (Gorlin, Pindborg, and Cohen, 1976). James

(1976) has noted that female zygotes, monozygous twins, of which conjoined twins are an example, and infants with neural tube defects may arise in instances in which fertilization has taken place late in the reproductive cycle. At present, no etiologic agent has been identified.

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