PEDIATRIC PATHOLOGY

Monocephalus Diprosopus, a Rare Form of Conjoined Twins, and Associated Congenital Anomalies.

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Craniofacial duplication (diprosopus) is a rare form of conjoined twins. A case of monocephalus diprosopus with anencephaly, cervicothoracolumbar rachischisis, and duplication of the respiratory tract and upper gastrointestinal tract is reported. The cardiovascular system remained single but the heart showed transposition of the great vessels. We present this case due to its rarity, and compare our pathologic findings with those already reported.

Key words: Conjoined twins, Monocephalus, Diprosopus, Anencephaly, Congenital anomalies, Rachischisis, Cephalothoracopagus.

Conjoined twins in man is a rare biological event. The incidence is estimated to range from 1:20,000 to 1:50,000 for joined twins, and 1:180,000 to 1:15,000,000 for diprosopia (1). Diprosopus twins, also referred as lateral cephalothoracopagus, are the rarest variety of symmetrical conjoined twins, with approximately 30 cases reported in the world medical literature up to 1987 (1). These cases constitute a spectrum extending from a simple nasal duplication to doubling of the head (dicephalus) on a single trunk with unremarkable limbs.

Case Report

A conjoined male twin was born to a 23 year old G3P2A0 hispanic woman who started her prenatal care after 39 weeks of gestation, completing only 3 prenatal evaluations without apparent complications. In an obstetric sonogram, polyhydramnios was noted as well as a neural tube defect, acrania and other congenital anomalies incompatible with life. The mother decided to have a termination of pregnancy. An autopsy was requested and a permit granted by the parents.

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Postmortem examination revealed a conjoined anencephalic male twin with partial duplication of the face, a single trunk with two arms, two legs, and a single male genitalia (Figures 1 and 2). He measured 36.7 cm from crown to heel, (expected 46.7 ± 4.4 cm), 19.5 cm from crown to rump, (expected 34.3 ± 1.9 cm), and weighed 1850 grams, (expected 2789 ± 520grams). Externally, he presented an anencephalic habitus and a broad neural tube defect that extended from the neck down to the upper lumbar region (cervicothoracolumbar rachischisis) (Figure 3). The head was macrocephalic with a circumference of 29 cm. The calvarium was absent as well as the encephalic mass. Fibrovascular membranes (cerebrovasculosa) were seen covering the base of the cranium. The pituitary gland nor the optic nerves were identified. The partially duplicated face (Figure 2), presented four frog-like eyes, two noses, and two medially fused mouths with anterior and

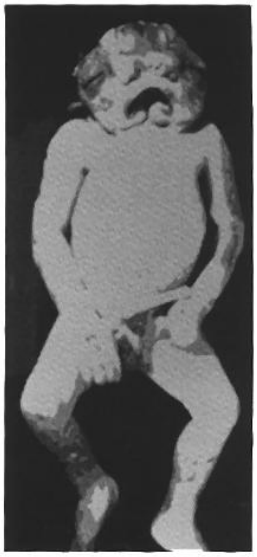

Figure 1. Postmortem photograph, monocephalus diprosopus.Note macrocephaly, craniofacial duplication, a single trunk, and well formed extremities.

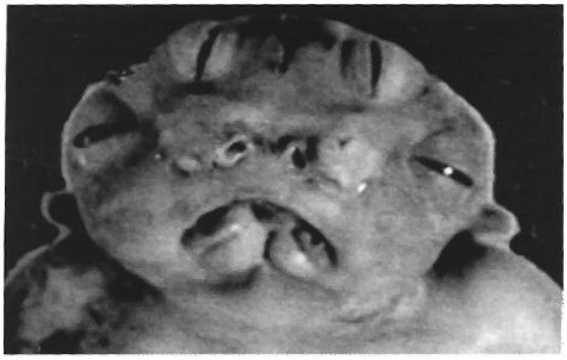

Figure 2. Note partial facial duplication with four frog-like eyes, two noses, and two medially fused mouths.

posterior communication between the two oral cavities. There were two tongues joined in the posterior third. There were two small chins, and two low set and malformed ears (one at each side). The neck was broad and short.

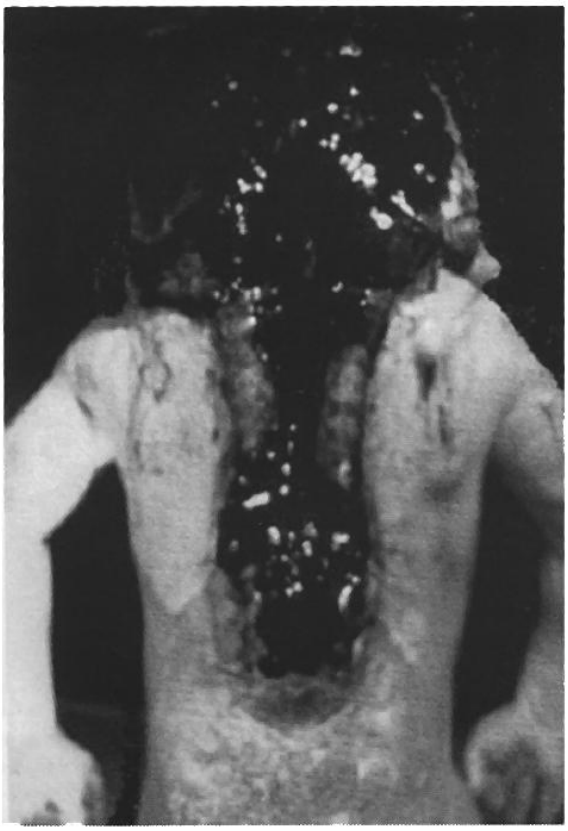

Figure 3. View from the back, note anencephaly, and a broad neural tube defect extending from the cervical region to the lumbar area.

The internal anatomy was somewhat complex. A single normal appearing thymus was present. The airways (Figure 4) were completely doubled, with two medially fused larynges, two tracheae, and two pairs of hypoplastic lungs in each side. The bronchi were abnormally branched.

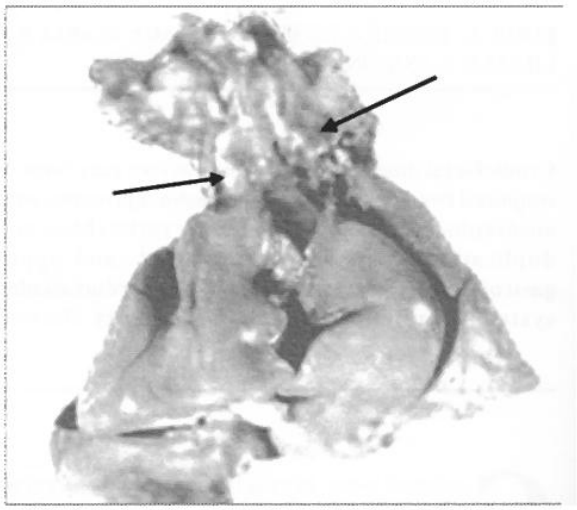

Figure 4. Duplication of the respiratory system. Note two partly fused larynges, tracheae (arrows), and a pair of hypoplastic lungs at each side.

Each pair of lungs presented four pulmonary veins, fused distally into two pulmonary veins with adequate drainage to the left atrium. There was a large thyroid gland seen as a single mass around the two tracheae. A single heart with transposition of the great vessels was present. The left atrium received four pulmonary veins, two from each pair of lungs; originally four that fused into two. The aortic arch was left sided, and showed normal branching of the great vessels. The gastrointestinal tract was duplicated proximally up to the duodenum, where it fused into a single small bowel (Figure 5). The appendix was right sided, and located in the right lower quadrant. The liver, gallbladder, pancreas, and spleen were shared by both twins and otherwise normal. The urogenital tract was also shared by both twins and consisted of a pair of kidneys, ureters and a single bladder, and urethra. A pair of adrenal glands were present, adequate in size, shape, weight, and position.

The monochorionic monoamniotic twin placenta weighed 460 grams after cord and membranes were trimmed, and measured 17 x 15 x 2.2 cm. The umbilical cord had a length of 19.5 cm, and presented two vessels. No abnormalities of the fetal membranes or placental disc were grossly seen. Histological sections confirmed the presence

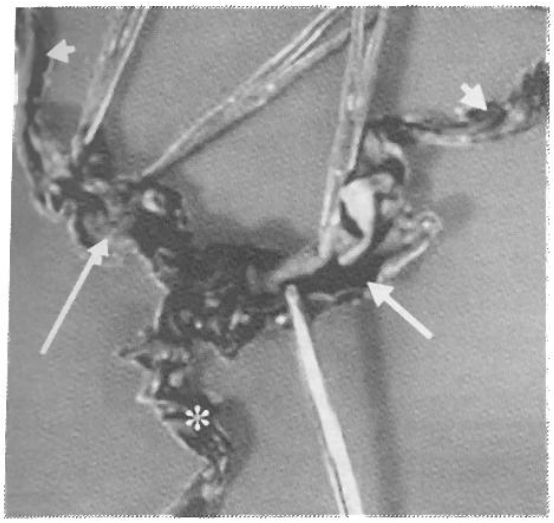

Figure 5. Duplication of the upper gastrointestinal system ending into a single small bowel. Note the two esophagus (arrow heads), two stomachs (arrows) and a single duodenum (*).

of a single umbilical artery, and showed focal villous stromal fibrosis, moderate perivillous and intravillous fibrin deposition, focal villous stromal hemorrhages, and retained muscularized maternal vessels.

Discussion

Conjoined twins in man is a rare biological event with an incidence estimated to range from 1:20,000 to 1:50,000 for joined twins, and 1:180,000 to 1:15,000,000 for diprosopia (1). The most accepted theory of the formation of conjoined twins, including diprosopus, is incomplete splitting of a single embryo between days 13 and 15 postconception. Until now, most evidence indicates that conjoined twins are incomplete separate monozygotic or identical twins (the only true twins) as they are derived from a single ovum. The anatomical variations described are determined by the degree and point of origin of the splitting. Their nomenclature reflects the anatomic site where the twins are joined (See Appendix 1, p. 240).

Many factors have been suggested as predisposing or responsible for the development of the conjoined twins such as difficult closing of the rostral neuropore, aging of the ovum, environmental factors, maternal age, and chromosomal abnormalities, among others. However, the exact nature of the formation of conjoined twins is uncertain, and many of these factors have no correlation with twinning. Other studies have suggested possible "epidemics" (2), while others propose clustering (3). The sex of most conjoined twins is female (4-6), although in a study performed in Latin America during 1967-1986, an even distribution (12 M, 11F) was reported (3). They are more common in nonwhites than whites (4). Diprosopus twins, also referred as lateral cephalothoracopagus, are the rarest variety of symmetrical conjoined twins, with approximately 30 cases reported in the world medical literature up to 1987 (1). These cases constitute a spectrum extending from a simple nasal duplication, facial duplication, to doubling of the head (dicephalus) on a single trunk with unremarkable limbs. The pathogenesis of diprosopus is not well understood, but the factors that play a role in diprosopus are probably similar to the ones that affect monozygotic twinning (1). The diprosopus variety presents a high incidence of neural tube defects suggesting the possibility of a common mechanism or relation between these two events (6-9). Cardiac malformations and anencephaly are also frequently associated anomalies.

Other congenital anomalies reported in the literature associated with diprosopus include diaphragmatic hernias (7,8), vertebral fusion(8), duplication of the vertebral column, and holoprosencephaly (1), cheilognatho-palatoschisis (presence of a cleft in the lip, upper jaw, and palate) (8), and gastroschisis (9). A case with hydrocephalus was reported by Chervenak et al, but this association is rare (7). The gamut of defects is extense, varied, and not constant, reflecting the complexities of embryogenesis. Our case is similar to one reported by Moermann et al in 1983 (10). Their case in addition to the duplication of the respiratory and gastrointestinal tract, presented duplication of the thymus, pancreas, and spleen.

Early prenatal diagnosis is possible by obstetric ultrasonography. With early detection, the option of a therapeutic abortion may be offered to the parents.

Resumen

La duplicación craniofacial (diprosopus) es una forma rara de gemelos siameses. Presentamos un caso de gemelos siameses tipo diprosopus monocéfalo con anencefalia, raquisquisis cervicotoracolumbar, y duplicación de su sistema respiratorio y gastrointestinal. Su sistema cardiovascular era sencillo y compartido por ambos siameses pero presentaba transposición de los grandes vasos. Presentamos este caso por ser muy raro y comparamos nuestros hallazgos patológicos con los de los escasos casos reportados en la literatura médica.

Appendix 1

Terms and Definitions of Conjoined Twins

Craniopagus:	joined at cranial vault, two separate bodies except at head.
Dicephalus:	two distinct heads, one body.
Diprosopus:	partial craniofacial duplication, one body.
Ischiopagus:	joined at the ischia.
Omphalopagus:	joined at the umbilicus.
Pagus:	from Greek pagos, that which is fixed.
Pygopagus:	joined by lateral and posterior surface of coccyx and sacrum—back to back.
Parasite:	one completely developed viable twin, and one partially developed nonviable twin.
Thoracopagus:	joined at thoracic wall.
Xiphopagus:	joined at the xiphoid process.
Thoraco-omphalopagus:	joined at the thorax, and abdomen.

Acknowledgement

The authors wish to thank Mr. Manolín Fernández for his technical advice with the illustrations.

References

1. al Muti Zaitoun A, Chang J, Booker M. Diprosopus (Partially duplicated head) associated with anencephaly: A case report. Pathology Research and Practice 1999;195:45-50.
2. Herring SW, Rowlatt UF. Anatomy and embryology in cephalothoracopagus twins. Teratology, 1981:23:159-173.
3. Castilla EE, López-Camelo JS, Orioli IM, Sánchez O, Paz JE. The epidemiology of conjoined twins in Latin America. Acta Genet Med Gemellol, 1988;37:111-118.
4. Milham S. Symmetrical conjoined twins: An analysis of the birth records of twenty-two sets. J Ped, 1966;69:643-647.
5. Edmonds LD, Layde PM. Conjoined twins in the United States, 1970-1977. Teratology, 1982;25:301-308.
6. Riccardi VM, Bergmann CA. Anencephaly with incomplete twinning (Diprosopus). Teratology, 1977;16:137-140.
7. Chervenak FA, Pinto MM, Heller CI, Norooz H. Obstetric significance of fetal craniofacial duplication. A case report. J Rep Med 1985;30:74-76.
8. Pavone L, Camera G, Grasso S, Gambini C, Barberis M, Garaffo S, Russo S. Diprosopus with associated malformations: Report of two cases. Am J Med Gen, 1987;28:85-88.
9. Fontanarosa M, Bagnoli G, Cioloni P, Spinelli G, Curiel P. First trimester sonographic diagnosis of diprosopus twins with craniorachischisis. J Clin Ultrasound, 1992;20:69-71.
10. Moermann P, Fryns JP, Goddeeris P, Lauweryns JM, Assche AV. Aberrant twinning (diprosopus) associated with anencephaly. Clin Genet, 1983;24:252-256.