

Early Prenatal Diagnosis of Conjoined Cephalopagus Twins

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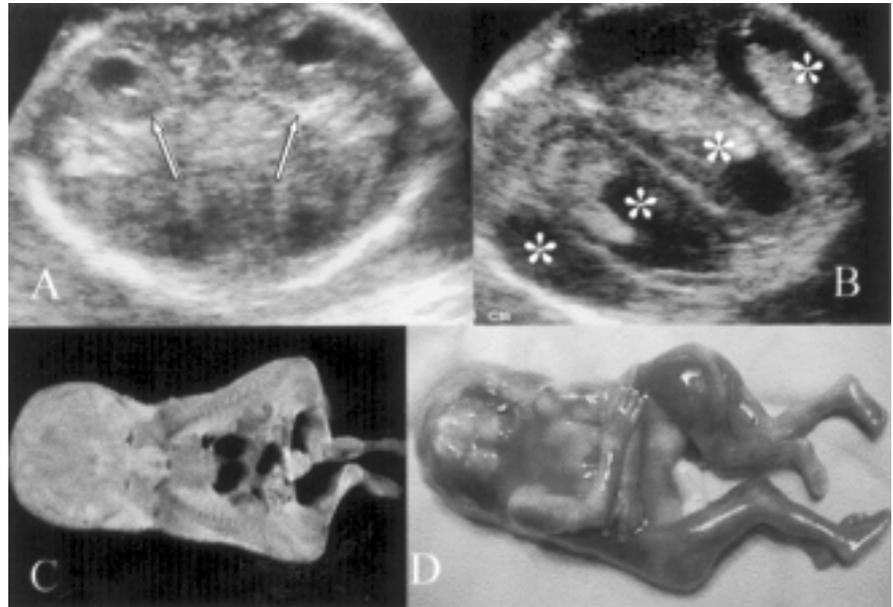
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One in every 50,000–100,000 births is a conjoined twin [1]. It has long been postulated that conjoined twinning is due to fission of the developing embryo. However, based on an animal model in Triton embryos and the study of 1,200 case reports of conjoined twins, Spencer [2,3] recently concluded that conjoined twinning is due to *fusion* of two embryos rather than *fission*. The union occurs either ventrally or dorsally at sites where the surface ectoderm is absent or is programmed to be fused or disrupted (primordia of heart, oropharyngeal membranes and cloacal membranes). The prognosis for conjoined twins depends on the presence of other anomalies, the extent of union of the intracranial, intrathoracic and intraabdominal structures, and abnormal vascular connections.

Patient Description

We report a case of cephalopagus diagnosed at 15 weeks of gestation by vaginal ultrasonography. After being informed of the grave prognosis of this condition, the parents opted for termination of pregnancy. Prenatal ultrasound examination [Figures A,B], MRI findings [Figure C] and a view prior to autopsy [Figure D] of the cephalopagus showed a single large head with four cerebral hemispheres, two cerebelli, two wide-spaced eyes, two ears, one nose, and one mouth with central clefts in both lips. The twins shared a cranium, oropharynx, sternum, liver, parts of intestine and an umbilicus (two veins and two arteries). There were separate hearts, lungs, stomachs, spleens, spinal cords and female genitourinary tracts. In addition, there were four upper and four lower extremities. Our findings were compatible with cephalopagus as described by Spencer [3].



Prenatal ultrasound at 15 weeks of gestation. [A] A single head with two cerebelli (arrows) and [B] four cerebral hemispheres (asterisk). [C] MRI in the coronal plane shows single cranium and oropharynx, conjoined thorax and separate spinal columns. [D] Macroscopic view of the cephalopagus prior to autopsy.

Comment

The embryonic classification of conjoined twins consists of ventral union (87%) and dorsal union (13%). There are altogether eight types of conjoined twins that vary as to the extent and location of their union [2]. The cephalopagus is the rarest type among the ventral union subgroup of conjoined twins and accounts for 11% of all cases, i.e., one in every million births [2]. It consists of a ventral union of the upper half of the body from top of head to upper abdomen. There is one large head and a wide face that results from two conjoined faces rotated through 90 degrees from the vertical axis. Hence, surgical separation of cephalopagus twins is not an option and the prognosis is extremely poor [3]. Therefore, early prenatal diagno-

sis of cephalopagus by either two-dimensional or three-dimensional ultrasound [4] and magnetic resonance imaging [5] are extremely important, so that termination of pregnancy can be offered to the parents as early as possible and trauma to the birth canal via vaginal delivery avoided.

During prenatal ultrasound examination, cephalopagus may be mistaken for a singleton fetus because of superposition of the conjoined fetuses and the extreme degree of fusion. Therefore, it is imperative to scan the fetus in different planes. With advanced ultrasound imaging, even earlier prenatal diagnosis of cephalopagus might become feasible in the embryo. Theoretically, cephalopagus might appear as a "V sign" by ultrasonography. Diagnosis of cephalopagus in the 7th to 8th week of

gestation will allow immediate termination of pregnancy. This is possible also for Orthodox Jews, since pregnancy termination is permitted by Jewish Law until 49 days of pregnancy.

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