Unusual Cardiac Malformations in Conjoined Twins: Thoracopagus Twins with Conjoined Pentalogy of Cantrell and an Omphalopagus Twin with Atretic Ventricles

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Abstract. Two unrelated cases of conjoined twins were found to have cardiac malformations that apparently have not been reported previously. In one case, thoracopagus twins had an extensive thoracoabdominal wall defect that resulted in ectopia cordis of a conjoined heart along with evisceration of the shared liver and intestine along with one spleen. These malformations, accompanied by defects in the sternum, diaphragm, and supraumbilical abdominal wall, constitute a conjoined pentalogy of Cantrell. In the second case, the heart of one of omphalopagus twins consisted of a solid ventricular mass with only a minute aortic cavity but no atroventricular communication—an ineffective heart that could develop only in a conjoined or chorioangiopagus twin. In both cases, a common atrium lay in the primitive (embryologic) position caudal to the ventricles.

Key words: Conjoined twins — Pentalogy of Cantrell — Cardiac malformations

It has been postulated that all conjoined twins arise from the secondary fusion of two homologously oriented, early monozygotic embryonic discs, with the site of union determining the type of twins [4]. Both omphalopagus and thoracopagus are thought to result from union at the rostral extremity of the discs near the end of the second week after fertilization (perhaps stage 7 or 8), when the primordia of the septum transversum and the heart are just outside this portion of the disc (Fig. 1a). Minimal fusion between two rostrally oriented embryonic discs at this

Fig. 1. Stylized representation of embryos at approximately stage 8 (a, early third week), stage 9 (b, late third week), and stage 10 (c, early fourth week after fertilization). Shown are the amniotic cavity above the embryonic disc, the cardiac primordia (C) and septum transversum (the dark area adjacent to the heart), the vertically striped oropharyngeal and cloacal membranes, the stippled body stalk, and the yolk sac open ventrally.
very early stage might involve only the septum transversum, giving rise to omphalopagus (united in the diaphragms, perhaps also in the pericardium, but not in the hearts), whereas more extensive fusion would also result in comingling of the cardiac primordia, as seen in the single multiventricular heart of thoracopagus. Both these types of twins develop on a single yolk sac and later will share some portion of the intestinal system, with the union later coming to involve the lower thoracic and upper abdominal walls.

The somatic union of thoracopagus and omphalopagus may be identical, but (by definition) the former always share a single abnormal heart, whereas the latter always have two separate hearts, perhaps sharing a single pericardium but without any patent intercardiac vascular connection [3]. The distinction is important in that thoracopagus only very rarely survive, with or without surgery, whereas omphalopagus usually do.

Although both omphalocoeles and cardiac anomalies (of either conjoined or individually owned hearts) are common in ventrally conjoined twins, neither of the malformations reported here were found in a recently completed review of 1261 cases of conjoined twins (Spencer, *Conjoined Twins*, in press). In only one instance was the heart reported to have been exposed—a case in which the heart protruded through an abdominal defect, presumably the result of caudal traction by herniated abdominal viscera; in this case, a newborn female had a supernumerary forequarter attached to the ventral thoracic wall (omphalopagus twins consisting of one normal fetus and the remnants of a parasite) as well as gastrochisis with evisceration of the intestine, liver, and heart, the latter lying 1.5 in. caudal to the costal margin. Neither the thoracic wall nor the diaphragm were described, but the former must have been intact as the parasitic limb was attached here, and the ventral portion of the latter must have been defective to permit the heart to escape from the thorax. The infant was tied spread-eagle on a box and a 30-minute operation performed at home without anesthesia (in 1891). After suturing the 3-in. wound resulting from the removal of the parasite, the eviscerated heart, liver, and intestine were simply replaced within the body and held in place with a binder. The wounds healed per primum and the infant was completely well by the age of 10 weeks [1]. In another case (not conjoined twins), a fetus *in utero* had typical ectopia cordis of a heart without chambers and with the atrial portion caudal to the ventricular mass [6].

Genetic studies (not performed in the cases described here) only rarely have been reported in conjoined twins, but significant developmental malformations are more common in the right than the left twin and in males rather than in females (Spencer, in press).

**Terminology**

The following terms are used to identify each individual conjoined twin and to distinguish between shared and individually owned structures:

1. The broader conjoined aspect of union is referred to as the “anterior” and the narrower aspect as the “posterior,” with the words always enclosed in quotation marks;
2. In relation to the “anterior” aspect, the fetuses are identified as the twin-on-THEIR-right (RT) and twin-on-THEIR left (LT);
3. Dorsal and ventral refer to the individual twin;
4. Rostral and caudal may apply to either shared or individually owned structures; and
5. Shared viscera may result from secondary fusion (the single conjoined heart in the thoracopagus reported here) or from failure of division (the unlobed portion of the upper small bowel in both cases).

An embryologic aberration not seen in singletons but common in conjoined twins is “division and diversion” [5]. Median (midline) structures at the site of union are not simply buried but instead are divided in the median, each half diverted laterally and then united to a half from the other embryo. This 90° axial rotation results in two structures at right angles to the usual location, often quite normal, but with each twin owning half of each (such as the livers in case 1 reported here).

**Case 1: Conjoined Pentalogy of Cantrell**

These female thoracopagus twins were those of a 25-year-old gravida III whose previous pregnancies resulted in a miscarriage at 5 weeks and a 39-week healthy female infant. Abnormal maternal serum α-