Cephalothoracopagus Syncephalus : A Case Report and Literature Review

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Abstract : *Reports of cephalothoracopagus conjoined twins have appeared rarely in the scientific literatures. At Chulalongkorn Hospital, this malformation is rare with an incidence of one in eight sets of conjoined twins. A case of cephalothoracopagus conjoined twins is presented. Mechanism of conjoining and type of cephalothoracopagus are discussed. (Thai J Obstet Gynaecol 1990; 2: 37-42.)*

Key words : cephalothoracopagus syncephalus, conjoined twins, case report

One of the most interesting congenital malformations is conjoined twins. Since the first reported case, conjoined twins have been the object of curiosity of laymen and doctors alike. The rarity and unusual appearance of the malformation contribute to the fascination. Although the diagnosis is most often missed until labour ensues, conjoined twins are occasionally diagnosed antenatally on the basis of certain radiologic criteria and ultrasonographic findings. Conjoined twins occassionally cause delivery dilemmas for the obstetricians.

The cephalothoracopagus variety is particularly rare. A case of cephalothoracopagus conjoined twins is presented.

Case Report

A 29-year-old Thai woman, gravida 3, para 2-0-0-2, who experienced an uneventful pregnancy, had a history of spontaneous labour beginning 4 hours prior to admission. She had received no prenatal care. Her last menstrual period was unknown. The previous two children were normal and there was no history of twins. She did not smoke cigarettes nor drink alcohol. During the pregnancy she had not taken iron supplement and vitamins. She had an unremarkable medical history. On her way to the hospital, she delivered vaginally in the taxi, with cephalic presentation of stillborn conjoined twins.

External Appearances

The twins had a combined weight of 1420 g. Physical examination showed female twins conjoined anterolaterally at the head and thorax, with separate torsos below the umbilicus and with four arms and legs. The twin's head was large ovoid, with two eyes, one nose and one mouth, all facing forward. There were four ears, one low-set ear on either cheek and two opposing ones at the back of the head opposite the facial features. The conjoined twins had two trunks that opposed each other, joined from a single neck and extended to a shared umbilicus. There were separate pelves, four lower extremities and four upper extremities despite the fused trunk. Complete female external genitalia were present in each body. There were no anomalies of the extremities (Figures 1,2).

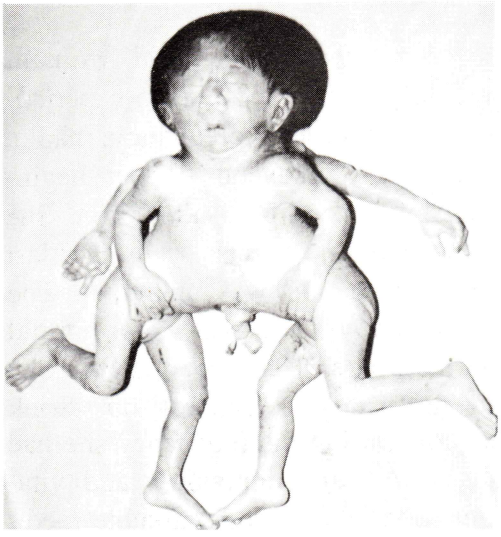

Fig. 1 Anterior view of conjoined cephalothoracopagus twins showing the common ovoid head, two widely spaced eyes, single nose and mouth and low-set ears.

The thorax and abdomen are fused, meeting with a single umbilical

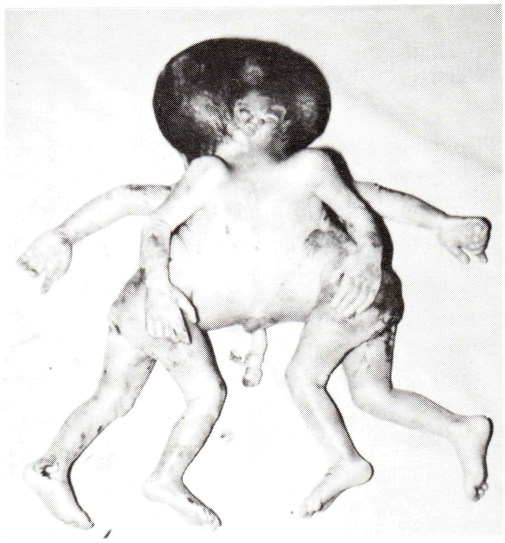

Fig. 2 Posterior view of conjoined cephalothoracopagus twins showing two ears separated by 2 mm were present on the opposite side of the face.

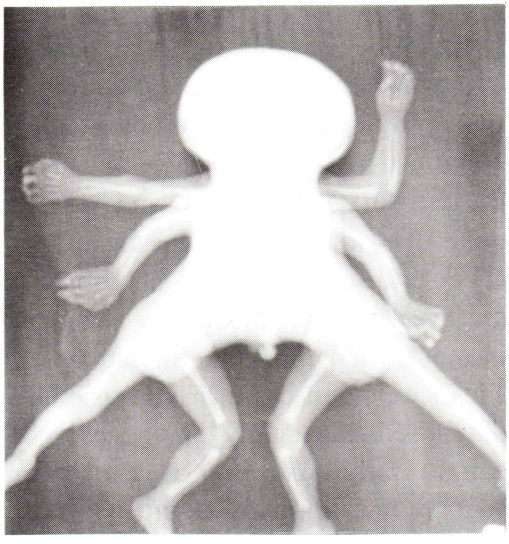

Fig. 3 Post mortem total body roentgenogram of conjoined cephalothoracopagus twins, anteroposterior view. Note the common head with two spines extension laterally, the fusion of the chest and abdomen and the four upper and lower extremities.

cord.

The total body roentgenogram confirmed the two separate spines (Figure 3).

Placenta

The placenta weighed 500 g. The placental plate appeared normal. There was one anmiotic sac.

The autopsy was not performed because of the mother's refusal.

The maternal postpartum course was unremarkable. Grief counseling was initiated. She was ready for discharge from the hospital on the third postpartum day.

Discussion

The exact incidence of conjoined twins is not known, but estimates of conjoined twinning have varied from 1 in 33000 to 1 in 165000 births[1-5]. An especially rare form is the cephalothoracopagus, with fused head and thorax. The actual incidence of cephalothoracopagus is vague. The malformation is said to occur in one of every three million births or one in 58 conjoined twins[6]. Data were analyzed on 228434 births delivered at Chulalongkorn Hospital over the period 1975-1989. The analysis identified 8 set of conjoined twins, for a crude incidence of 1 per 28500 births.

Stillbirths	2 cases(25%)
- Thoracopagus	1 case
- Cephalothoracopagus	1 case
Livebirths	6 cases(75%)
- Thoracoomphalopagus	2 cases
- Thoracopagus	1 case
- Ischiopagus tetrapus	1 case
- Dicephalus dipus triradius	1 case
- Dicephalus tripus tetrabrachius	1 case

Three types of cephalothoracopagus twins have been described[7]:

1. deradelphus : with one face and two ears

2. janiceps : with two faces on opposite side of the head

3. syncephalus : with a single face and four ears, two of which are on the back of the head.

This case appeared to be of the last variety. A review of the available English literatures shows that only a few cases of cephalothoracopagus syncephalus have been reported[4,5,8-16] (Table 1).

The most papers reported a single cortex, two brain stems, two thoracic cavities, two hearts, four lungs and a single gastrointestinal tract that bifurcated at the level of the small intestine. Most cases had two livers and normal genitourinary tracts. Some cases of triplet pregnancy associated with cephalothoracopagus have also been reported[16,17].

Mechanism of conjoining

Embryologically, conjoining results from a twinning event occuring between days 13 and 15 after fertilization[18]. The location of the connection and the degree and nature of organ duplication will depend on the area affected by the twinning event.

Conjoining, according to one theory, is the result of the incomplete separation of two embryonic axes in

Table 1 Caphalothoracopagus syncephalus : comparison of anatomical variations in reported cases (numbers indicate numbers of each organ present)

	Grundfast[5] 1950	Bartlett[8] 1959	Fahmy[9] 1966	Carlson[12] 1975	Furuhashi[13] 1980	Delprado[14] 1984	Benbridge[15] 1987
Central nervous system							
Cortex	-	2	-	1	1	3	1
Brain stem	2	2	-	2	2	2	2
Thorax							
Heart(s)	1	2	1	2	2	2	1
Connections	-	CA	-	CA	Heart to aorta	CA	-
Lungs	4	4	2	4	2	4	4
Gastrointestinal tract							
Esophagus	2→1	1	-	1	1	1	1
Stomach	1	1	1	1	1	1	1
Small intestine	1→2	1→2	-	1→2	1→2	1→2	1→2
Large intestine	2	2	2	2	1	2	
Pancreas	1	2	-	-	-	1	2
Liver	2	2	1	2	-	2	1
Genitourinary tract	NAD	NAD	-	NAD	NAD	1-NAD 1-H	NAD

H=Hydronephrotic, CA=Communicating artery, NIL=No connection, NAD=No abnormality detected

one ovum after the embryonic disc has formed[19]. On the basis of the mechanism of somite development, would duplicate themselves later than the more central areas. That theory is supported by the observation that conjoined twins are fused more frequently in the midaxial than proximal or distal areas. Sturrock and McKenzie[20] suggested a "collision" theory as the primary fault in the development of the cephalothoracopagus. The two cranial ends of the embryonic monozygotic twin axes lie close enough to meet or even fuse. The head regions and brain (or brains) are thus fused into one head, with various degrees of body fusion in a caudad direction. Such an explanation is attractive in terms of the findings in this case.

The prenatal diagnosis of conjoined twins is thus important for several reasons. It allows correct planning of the site and type of delivery, referral to a center where appropriate obstetric and pediatric surgical facilities are available[21,22]. The most important first step in making the diagnosis is awareness of its possibility. All sets of twins when examined with ultrasound should be shown to be separated from each other. If separation is suspected but cannot be confirmed, careful examination with realtime ultrasound allows full assessment of the site and type of conjoining, it being the method which shows the soft tissue conjunction. It also allows shared organs to be seen, and both babies can be carefully screened for other system

abnormalities. A number of sonographic and roentgenographic features of ventrally fused, conjoined twins have been described and listed (Table 2).

Table 2 Roentgenographic and sonographic features of ventrally fused conjoined twins[22-25]

Findings	Roentgenography	Ultrasonography
Fetal body parts on the same level	X	X
Constant relative fetal position	X	X
Fetal extremities in unusual proximity	X	X
En face fetal position	X	X
Bibreech, less commonly bicephalic presentation	X	X
Hyperextension of one or both cervical spines	X	X
Nonseparable, continuous external skin contour		X
Single heart sound by Doppler		X
Solitary large liver and heart		X
Multiple shared omenta		X
Solitary umbilical cord with > 3 vessels		X

Once the diagnosis of conjoined twins is established, the method of delivery should be based on the potential for infant survival, their size, type of joining, and parental attitudes[21].

When the diagnosis of conjoined twins is made before viability, the option of pregnancy termination should be offered to the parents. After viability, serial examinations are indicated to monitor fetal growth and the development of hydrops, and to detect fetal demise. The method of delivery depends upon the prenatal assessment of the likelihood of survival [26]. Although vaginal delivery is possible[27], dystocia occurs frequently[28]. Vaginal delivery should be reserved for stillbirth and for variety that are incompatible with life. A destructive procedure (embryotomy) can be considered for stillbirths. Caesarean section is the method of choice to maximize survival of the twins, because it decreases the risk of birth trauma and hypoxia. A vertical uterine incision is recommended. In cephalic / cephalic presentation, the heads should be delivered before the rest of the bodies. The same principle is applied to breech / breech and cephalic/ breech conjoined twins[26].

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