

A Morphological Study on Conjoined Twins in Korea[†]

Je G. Chi,* Jeong Wook Seo, Hye Kyung Lee, Geung Hwan Ahn,
Yoon Sung Lee, Sung Sik Shin, Eun Sil Yu, Sang Yoon Kim
and Kyung Ja Cho

Department of Pathology, College of Medicine, Seoul National University, Seoul 110, Korea

= Abstract = General morphological patterns of major organ systems in various types of conjoined twins are described. Sixteen pairs of conjoined twins used in this study were five thoracopagi, four dicephalic twins, four cephalothoracopagi, a craniopagus, a dicephalus dipygus and a heteropagus. The fusion of skeletal systems was closely related to the external morphology. The central nervous system was always duplicated. Nose and oropharynx patterns were determined by the number of faces, but the lower respiratory organs were always duplicated. The digestive tract was related to the skeletal union but the level of union and duplication varied. The liver was frequently fused in the cases of truncal union except in cephalothoracopagi. Genitourinary system patterns were related to the number of legs. Cardiovascular systems showed five distinct types of vascular union. The umbilical vessels were related to the number of trunks, but variations existed. In the fusion of symmetrical conjoined twins, there appeared to be certain rules that can be summarized as follows. The twin pairs tended to face each other. The fused portion was in the frontal plane, whereas the rest of the bodies were in lateral planes. Conjoining of the bodies in side-by-side pattern was associated with the same inner sides. In Korea, we seem to have a higher relative incidence of cephalothoracopagus and dicephalus, whereas in other countries thoracopagus is the predominant type.

Key Words: *Conjoined twin, Siamese twin, Symmetry*

INTRODUCTION

Conjoined twins are among the best models in the study of symmetry and development of organ systems (Noonan 1978). Because of this, the analysis of the pattern and degree of fusion of organ systems in conjoined twins is most important.

There are many reports on the anatomical features of conjoined twins. Many systematic analyses of organ systems in conjoined twins have been performed. However, they were mainly concerned with

the cardiovascular system in thoracopagus (Izuka-*wa et al.* 1978). The analysis of various organ systems that is generally applicable to various types of conjoined twins has rarely been studied (Mudaliar 1930; Singer and Rosenberg 1967).

The authors examined 16 pairs of conjoined twins. The autopsy findings were reanalysed with reference to the seven major organ systems and to the external morphological types. The fusion and duplication of the organs were mainly related to the external morphological types but there were significant differences among the different organ systems. The analysis of the cardiovascular system using this series (Seo *et al.* 1985) and some of the cases with their autopsy findings were reported previously (Table 1).

[†] This study was supported in part by research grant of Seoul National University Hospital(1983)

* Author for correspondence

MATERIALS AND METHODS

Sixteen cases of conjoined twins were used for this study. They were chosen from the autopsy file of the Seoul National University Hospital from years between 1967 and 1985. Some cases were sent from other hospitals in Korea, but all autopsies were performed at the Department of Pathology, College of Medicine, Seoul National University.

The external morphological types of these cases are categorized according to the classification of Potter and Craig (1975) (Table 1). Fifteen cases

were symmetrical conjoined twins and one case was heteropagus. Holoacardius, fetus-in-fetu and epignathus were not included in this study. Symmetrical conjoined twins with nearly complete components were five thoracopagi (cases 6,12,13,14 and 16) and a craniopagus (case 8). Symmetrical conjoined twins with fusion in lateral position were divided into four cases of cranial duplication, five cases of caudal duplication and a dicephalus dipygus. Four cases of cranial duplication included two cases of dicephalus dipus dibrachius (cases 4 and 7), a dicephalus dipus tribrachius (case 2) and a

Table 1. Clinico-pathological summary of cases

Case No.	External morphological type	Sex/ Age*	Gestational period (wk)	Body** weight(g)	Maternal age (yr)	Fused** organs	Remarks
1(A67-25)	Cephalothoracopagus	F/ND	32	est. 3,000	31	Sk, GI, Lv	Ahn <i>et al.</i> 1969 twins in triplets
2(A69-30)	Dicephalus dipus tribrachius	M/SB	36	3,500	?	Sk, GI, Lv Ge, Ur	Ahn <i>et al.</i> 1969
3(RCM-335)	Cephalothoracopagus janiceps	M/ND	24	1,120	26	Sk, CNS, GI, Lv	Chi <i>et al.</i> 1981
4(RCM-341)	Dicephalus dipus dibrachius	F/ND	36	3,350	26	Sk, Ht, GI Lv, Ge, Ur	Chi <i>et al.</i> 1981 neuroblastoma
5(A81-45)	Cephalothoracopagus	F/SB	32	est. 2,500	?	Sk, GI, Lv	
6(A81-54)	Thoracopagus	M/SB	35	3,400	29	Sk, Ht, Lv	gastroschisis, cleft lip
7(A81-66)	Dicephalus dipus dibrachius	F/SB	36	3,900	28	Sk, Ht, GI, Lv, Ge, Ur	Shin and Chi 1983 polysplenia
8(A82-41)	Craniopagus	F/6mos	38	4,750	32	Sk, CNS	
9(A82-75)	Cephalothoracopagus	M/ND	30	1,000	23	Sk, GI, Lv	Kim <i>et al.</i> 1984
10(A82-78)	Dicephalus tripus tribrachius	F/SB	28	1,450	28	Sk, Ht, GI Lv, Ur	
11(A83-11)	Heteropagus thoracopagus	M/SB	23	200	?	Sk, Lv	Yu <i>et al.</i> 1983 polysplenia
12(A83-66)	Thoracopagus	F/ND	?	2,500	?	Sk, Ht, Lv GI(?)	
13(A83-120)	Thoracopagus	M/SB	?	2,300	?	Sk, Ht, Lv GI(?)	
14(A83-121)	Thoracopagus	F/SB	?	2,800	?	Sk, Ht, Lv GI(?)	
15(A85-5)	Dicephalus dipus tetrabrachius	M/ND	36	3,800	27	Sk, GI, Lv Ge, Ur	
16(OA85-1)	Thoracopagus	M/SB	32	2,500	26	Sk, Ht, GI,	agensis of left upper extremity

* ND: neonatal death, SB: still birth

** est: estimation

*** Sk: skeletal system, CNS: central nervous system,
Ht: heart, GI: gastro-intestinal system, Lv: liver,
Ge: genital system, Ur: urinary system.

dicephalus dipus tetrabrachius (case 15). The cases of caudal duplication consisted of three cases of cephalothoracopagus syncephalus (cases 1, 5 and 9) and a cephalothoracopagus janiceps (case 3). The dicephalus dipygus was dicephalus tripus tribrachius (case 10). The heteropagus was thoracic union of well formed bodies of different size (case 11).

The autopsy findings were summarized in relation to their external morphologic types and seven major organ systems. The organ systems included skeletal system, central nervous system, cardiovascular system, respiratory system, digestive and hepatobiliary system, genito-urinary system, placenta and umbilical cord.

RESULTS

1. Skeletal system

The thoracopagus showed longitudinal division of the sternum and fusion of the divided parts of sternum of each co-twin. The xyphoid process and sternal body were always divided in these cases but the manubrium was not divided in case 6. Each of the twins was facing in an oblique direction and the left and right ribs and anterior and posterior sternums were asymmetrical. The thoracic kyphosis was replaced by abnormal lordosis.

The craniopagus (case 8) was of the parieto-occipital type and two cranial cavities were connected through a large opening at the temporo-parieto-occipital junction. The lambdoid and parieto-occipital sutures were widened and ipsilateral parietal bones were hypoplastic.

The dicephalus showed complete duplication of vertebral columns and the vertebral bodies were slightly rotated facing inwards and thoracic kyphosis was decreased (Fig. 1A). Cases 7 and 15 had hemivertebrae at lumbar spine. The inner ribs were short and fused to those of the other. The sternum was seen only at the anterior fusion of ribs. There was a fused scapula at the upper end of the posterior midline in dicephalus dipus dibrachius (cases 4 and 7). The midline upper extremity of the dicephalus dipus tribrachius (case 2) had a complete pair of humerus, radius, ulna and 10 fingers. The two humeri were connected to two midline scapulas. The dicephalus dipus tetrabrachius (case 15) had completely duplicated upper extremities and thoraces. All the dicephalic twins had a common pelvic cavity. Two sacral bones were connected by rudimentary ilia posteriorly, and by complete sets of pelvic bones and pubic symphyses

anteriorly. The lower extremities were a complete single pair. Dicephalus tripus tribrachius (case 10) had three upper and lower extremities. The median leg contained one femur, one tibia, two fibulas and 9 toes. The common pelvis was composed of two sacra and hip bones with three acetabulae. The anterior union of hip bones was the symphysis pubis, but posteriorly, there was cartilagenous union of rudimentary ilio-ischial bones. The middle arm contained a double number of arm and forearm bones, but humeri were fused and there were nine fingers. Three thoracic cavities were seen in the chest.

The cephalothoracopagus syncephalus had a single facial bone, but the temporal bones and ears were duplicated. The calvarium was incompletely duplicated. Two occipital bones were attached by a suture and two foramina magna were seen. The sella turcica was duplicated. Case 9 was an anencephalic cephalothoracopagus (Fig. 1B). The cephalothoracopagus janiceps had a double number of facial, frontal, parietal, temporal and occipital bones in symmetry to the sagittal and mid coronal planes. The inter-facial line was at a right angle to the inter-occipital line. The anterior and posterior sterna were seen in the interfacial line whereas the vertebral columns were in the inter-occipital line. A common pituitary fossa was seen at the center of the skull and the foramina magna were displaced to the center of the occipital bones on the left and right sides. Two complete sets of vertebral columns were seen. Ribs were relatively well developed and were symmetrical. The sternum was completely divided and fused into anterior and posterior sterna.

The heteropagus (case 11) had bony structure similar to that of the thoracopagus.

2. Central nervous system

Thoracopagus and dicephalus in all cases had completely separate brains and spinal cords corresponding to the individual twins. Two spinal cords were in two vertebral columns and separately entered through two foramina magna.

One case of craniopagus (case 8) showed union of the cranial vault at each parieto-occipital area by fusion of each anterior fontanel to form a large canal with no bony septum. The anterior halves of the cerebral gyri of both parieto-occipital lobes were interdigitating and separated by two well demarcated arachnoid membranes. Remaining posterior halves of the hemispheres were directly separated by a dural septum. Each co-twin had her own cerebrum, midbrain and cerebellum and there

was no actual unification of brain parenchyma except for a narrow stalk of white-matter, 0.8 cm in diameter, connecting each pons (Fig. 2). There were two foramina magna and two vertebral columns, in which two normal spinal cords were found.

Of four cephalothoracopagi, a case of janiceps (case 3) showed a relatively large head and two faces which were located exactly at opposite sides to the other. There were two sets of anterior and middle cranial fossae containing each cerebral lobes. Two posterior cranial fossae were seen in left and right sides (Fig. 3) Two falx cerebri were seen at right angles to two tentorium cerebelli. There were four cerebral lobes with poor gyral development. The brainstems and their connection to the spinal cords were not adequately described.

A cephalothoracopagus syncephalus (case 9) was characterized by anencephaly and two spinal cords separately entering the area cerebrovasculosa through two foramina magna, the area cerebrovasculosa was a thick reddish mass containing highly vascular disorganized neuroglial tissue. Two cases of cephalothoracopagus syncephalus (cases 1 and 5) had two posterior cranial fossae, each containing its own cerebellum and brainstem. the cerebrum was single and was located in the anterior cranial cavity. Two spinal cords entered their spinal canals through one (case 1) or two (case 5) foramina magna.

3. Cardiovascular system

The cardiovascular system of the conjoined twins is divided into five distinct groups according to the degree and pattern of fusion (Table 2) (Seo *et al.* 1985).

Five cases of thoracopagus showed atrial (type IIIa: case 12) or atrio-ventricular (type IV: cases

Table 2. Classification* of cardiovascular fusions in conjoined twins

Type	Degree of fusion	No. of cases
I	No significant fusion	5
II	Fusion of large vessels	3
III	Atrial fusion	3
	IIIa: Mirror image right atrial fusion (3)	
	IIIb: Other types of atrial fusion (0)	
IV	Atrio-ventricular fusion	4
V	Single heart in one of the twins	1

* Adopted from Seo *et al.* 1985.

6,13,14, and 16) fusions. The fused heart was in a frontal plane whereas the trunks of the twins were in lateral planes. One case of craniopagus (case 8) showed minor degree of cross circulation (type I). Carotid angiography was performed to confirm the absence of arterial anastomosis. Five cases of dicephalus showed type I (case 15), fusion of large vessels (type II: case 2), type IIIa (cases 4 and 13) and absence of one heart (type V: case 7). Four cases of cephalothoracopagus showed type I (cases 1 and 3) and type II cardiovascular systems (cases 5 and 9). The heteropagus (case 11) showed a type I cardiovascular system.

Eight cases showed abnormal situs. Two cases of dicephalus (cases 4 and 10) showed inverted situs of the right co-twin and the resulting inner sides were right. The heart showed mirror image right atrial fusion (type IIIa). Two cases of thoracopagus (case 12) and dicephalus (case 15) showed bilateral right sidedness and the resulting inner sides were right. Case 12 showed type IIIa fusion but case 15 had two legs and four arms, so the thoraces and hearts were separate and the iliac arteries were fused. Case 2 showed inverted situs of left co-twin and the resulting inner sides were left. The heart was not fused. Bilateral left sidedness of the left co-twin was seen in dicephalus dipus dibrachius (case 7) and throacopagus (case 13). The inner sides were left and the hearts showed a single heart in the right co-twin (case 11) showed bilateral left sidedness in the parasite. The splenic number and location were single left in cases 2,4,12 and 15, double, left and right in case 10 and polysplenia in cases 7 and 11.

4. Respiratory system

Thoracopagus, craniopagus and dicephalus had completely separate respiratory system from nose to lungs. Cephalothoracopagus with single mouth as in the cases of syncephalus (cases 1,5 and 9) had a single nasal cavity, but the pharynx was either single (case 9) or double (case 1). The pharynx was either single (case 9) or double (case 2). The pharynx of the other (case 5) was not examined. In cephalothoracopagus janiceps (case 3) two faces had separate nasal cavities but the pharyngeal cavity was common. The lower respiratory system of all the cases of cephalothoracopagus was duplicated from larynx of lungs. The number of lobes of the lungs was related to the situs of the co-twins, and the size and shape of the lungs were modified by the physical characteristics of the thoracic cavities. The autosite and the parasite in

heteropagus (case 11) had well developed respiratory system.

5. Digestive and hepatobiliary systems

Of five cases of thoracopagus (cases 6,12,13,14 and 16), cases 6 and 14 had two separate gastrointestinal tracts along the entire length and a saddle shaped, midline fused liver with separate gallbladders and separate bile ducts. Case 16 had a fused intestinal segment from the ligament of Treitz to mid-jejunum. The fused segment was dilated due to distal obstruction (Fig. 4A). The livers of cases 12,13 and 14 were fused but the intestinal tracts were not examined due to postmortem autolysis.

Each partner of craniopagus (case 8) had a normal, entirely separate gastrointestinal tracts and hepatobiliary trees.

Four cases of dicephalus revealed duplicated digestive tracts in their upper portions. The separate digestive tracts fused into a single lumen at the duodenum (case 4) or ileum (cases 2,7 and 15) (Fig. 4B). The liver was fused in all cases. The gallbladder and biliary tree were single in case 2, or double in cases 4,7 and 15.

Dicephalus tripus tribrachius (case 10) showed segmental union from ileum to mid colon, and the upper and lower tracts were separate in each co-twin. The liver was fused but the biliary system was separate.

Four cases of cephalothoracopagus showed duplicated digestive tracts in the lower portions. The upper single lumen began to separate into double lumens at the duodenum (case 1), jejunum (case 3 and 5) or ileum (case 9) (Fig. 4C). The pharynx was double in case 1. The liver was fused in case 5. Other cases had two discrete livers with close approximation.

The heteropagus (case 11) had separate digestive tracts and a fused liver with two gallbladders.

6. Genito-urinary system

The conjoined twins with two pairs of lower extremities as in the cases of thoracopagus, craniopagus, cephalothoracopagus and heteropagus showed completely separate genitourinary system.

Dicephalus dipus dibrachius (cases 4 and 7) and dicephalus dipus tribrachius (case 2) had a single pair of genito-urinary organs. Cases 2 and 7 had one additional midline adrenal gland. Dicephalus dipus tetrabrachius (case 15) and dicephalus tripus tribrachius (case 10) had double pairs of kidneys, ureters, bladders and gonads. The urethra showed Y-shaped fusion. The kidneys of the right

co-twin of case 15 were polycystic (Potter type IV) and the urethra was atretic (Fig. 5). Hypospadias was also seen in case 15.

7. Placenta and umbilical cord

Three cases of thoracopagus showed a single placenta and a single umbilical cord. The cords contained 4 arteries and 2 veins (case 6) or 4 arteries and 1 vein (case 14).

The craniopagus (case 8) had a complete pair of placentas and umbilical cords with the normal number of vessels.

Four cases of dicephalus dipus and one dicephalus tripus had a single placenta and umbilical cord. In the cords were 2 arteries and 1 vein (cases 2,4 and 10) and 1 artery and 1 vein (case 15).

Of the four cephalothoracopagi, three cases were examined. Case 1 was conjoined twins among triplets, and the twins had a single placenta and two cords in a common sac. The third baby had her own placenta and cord. Case 3 was janiceps twins and had two placentas. Each umbilical cord were fused at 10 cm distance from the placentas. The fused cord had 4 arteries and 1 vein. Case 9 had a single placenta and single cord with three arteries and two veins.

Four arteries and one vein were seen in the umbilical cord of heteropagus (case 11). Only two arterial channels were seen in the parasite.

DISCUSSION

In addition to the 16 cases in this report, nine published cases (Huh 1934; Lee and Park 1934; Chung *et al.* 1965; Kim 1966; Kim *et al.* 1967; Kim 1968; Kim and Kim 1970; Baik and Moon 1973; Hong *et al.* 1980), two unpublished cases and four heteropagi (Noh *et al.* 1980; Chi *et al.* 1984) have been identified in Korea. The external morphologic types of 31 Korean cases and reports of the distribution of those morphologic types are compared in Table 3. The relative frequency of each type of conjoined twins in Korea differs from those of other countries. Reviewing the literature, it is quite impressive that the thoracopagus is the most common variety of conjoined twins in the United States (59.3%) (Edmonds and Layde 1982), West Africa (Mabogunje and Lawrie 1980) and Singapore (Tan *et al.* 1971) in large series. The Korean cases, including the present series, show following comparable incidence; cephalothoracopagus (29.0%), thoracopagus (25.8%) and dicephalus (19.4%). In other words, thoracopagus is less common and

Table 3. The relative frequency of each type of conjoined twins in various countries

	United States 1970-1977 (1)	West Africa 1936-1978 (2)	Singapore 1960-1970 (3)	Korea 1934-1985 (4)
Thoracopagus	48	8	4	8
Pygopagus	2	1		
Craniopagus	5		1	1
Ischiopagus		1		
Monocephalus diprosopus		1		
Dicephalus (dipus)	3	2		6
Cephalothoracopagus	2		1	9
Dicephalus dipygus			1	1
Heteropagus	8	1		5
Others	13			1
Total No. of cases	81	14	7	31

(1) Refer to Edmonds and Layde (1982)

(2) Refer to Mabogunje and Lawrie (1980)

(3) Refer to Tan *et al.* (1971)

(4) Sixteen cases in this reports and 9 other cases in Korea (see text for references).

cephalothoracopagus and dicephalus are relatively more common in Korea than in other countries.

The thoracopagus was characterized by fusion of skeletal, cardiovascular and hepatobiliary systems. The skeletal union was characterized by fusions of anterior and posterior halves of the bifid sternum of each co-twin, but the manubrium was not divided in case 8 (Nichols *et al.* 1967; Singer and Rosenberg 1967). The fusion of the cardiovascular system was types IIIa or IV in this series, but 25 percent of thoracopagus have completely separate hearts (Nichols *et al.* 1967). The livers always fuse in thoracopagus in this series and in other reports. The gastrointestinal tracts are conjoined in 46% of thoracopagus (Nicholas *et al.* 1967) and the united segment is usually from the duodenal papilla to the lower portion of lieum (Zimmerman 1967).

The craniopagus shows fusion of the cranial vault and brain. The cranial cavities were connected by the widened sagittal suture. The cerebral gyri showed interdigitation without direct fusion but the midbrains were connected by a white matter tract.

Dicephalus dipus with two, three or four arms are the cases of cranial duplication with caudal union, and dicephalus tripus with three or four arms are the example of cranial and incomplete caudal duplication (dicephalus dipygus). The vertebral column and spinal cord are compeltely duplicated and the thorax and pelvis are partly

duplicated. The trunk is in a frontal plane and the anterior structures are similar to those of normal individuals. The respiratory system is separate. Abnormal situs in one co-twin is always present and the inner sidedness of both co-twins is either left or right. The cases with situs inversus or asplenia of the right co-twin show right inner sidedness, and those with situs inversus or polysplenia of left co-twin result in left inner sidedness. These findings are closely related to the cardiac morphology (Seo *et al.* 1985). The digestive tract fused into a single lumen from the levels of duodenum or ileum. The fused digestive tract reduplicated at the colon in a case of dicephalus with cranial and incomplete caudal duplication of body. The liver was fused. The urinary system was single or double and that was related to the number of extremities. The middle arm in tribrachius and middle leg in tripus need special comments on their symmetricity and sidedness. The middle arms in cases 2 and 10 were combined left and right arms in a common skin covering. But the single femur in a single socket, and single tibia in case 10 and another report (Mortimer and Kirshbaum 1941); single thumb and great toe in case 10 were neither left nor right ones but middle symmetrical bones.

The cephalothoracopagus showed caudal duplication. The levels of duplication were variable by the organ systems. The central nervous system was completely duplicated as the axial skeleton. The

cardiovascular system was separate in cephalothoracopagus. The two hearts were not left and right but anterior and posterior ones which were equally shared by two co-twins. This finding was previously described and explained by Herring and Rowatt (1981). The digestive system showed caudal duplication from the levels of duodenum, jejunum or ileum. The liver was single or double. The respiratory and genito-urinary systems were double. Janiceps variety of cephalothoracopagus showed symmetry to the interfacial and interspinal planes (Wedberg *et al.* 1979).

One case of the heteropagus was a thoracopagus with different sized co-twins and the internal organization was comparable to that of thoracopagus.

Presumably there are three rules of fusion in the symmetrical conjoined twins. The first is: "The twin pairs tend to face each other, and the anterior organs are single whereas the posterior structure of the same organ is partly duplicated." The cephalothoracopagus syncephalus has a single face but the cranial vault and the brain are duplicated. The dicephalus has an anterior sternum but there are two separate vertebral columns with slight torsion facing inwards, and rudimentary ribs posteriorly. The pelvic girdle of dicephalus has a single pubic symphysis but the sacrum and part of the ilium are duplicated. The thoracopagus showed slight variation in the facing angles. the cephalothoracopagus janiceps showed symmetry of the interspinal and interfacial planes and was an exception to the facing. The second rule is: "The fused organ is in the frontal plane whereas the rest of the bodies are in lateral planes." This phenomenon was previously described with the cardiovascular system (Ysander 1924). the single face in cephalothoracopagus and the single trunk in dicephalus are in the frontal plane. In these particular cases the designation of each co-twin should be the right one or the left one instead of arbitrary use of twin A and B. Again, the janiceps variety of cephalothoracopagus is an exception and each co-twin is called A or B randomly. The third rule is: "same inner sidedness in dicephalus and certain cases of thoracopagus." The abnormalities of situs of one or both co-twins made the same inner sides possible. In these cases hearts and livers were fused anteriorly. The trunk and extremities of cephalothoracopagus were more symmetrical both in the plane of fusion and antero-posteriorly. Two hearts and livers were seen anteriorly and posteriorly. So cephalothoracopagus and certain cases of thoracopagus do not exist with

the "same inner sidedness".

The pattern of fusion differs by the organ systems. The central nervous system is always duplicated and the duplicated systems rarely communicate. It is not surprising that the skeletal union corresponds closely to the external morphologic type. The upper digestive system and the nasal cavity seem to be related to the number of faces while the respiratory system from the level of larynx is always duplicated regardless of the complexity of the pharynx and the esophagus. The early development of the brain and the late development of lung buds from foregut would be the explanations.

The union of digestive tract is related to the skeletal union. However, variations exist in the level of union. The common levels of division and union are distal duodenum, Meckel's diverticulum and transverse colon. The digestive tract sometimes repeats duplication and unification in those cases of cephalothoracopagus and dicephalus dipygus. The fusion of the liver was seen in the cases of truncal union except in some cases of thoracopagus.

The genito-urinary system is duplicated in the cases with four legs. Dicephalus dipus with two or three arms has two kidneys, whereas dicephalus dipus with four arms and dicephalus tripus have separate paired genito-urinary systems. There was no case of three kidneys.

The cardiovascular system was the major topic in many of the studies on conjoined twins (Noonan 1978; Marin-Padilla *et al.* 1981; Leachman *et al.* 1967). The type of vascular union could be predicted by the external morphologic type and abnormalities in the situs (Seo *et al.* 1985).

The placenta and umbilical cords are important organs in twin formation. Conjoined twins are presumably monozygotic twins and the fetal membranes are monochorionic and monoamnionic (Wiegenstein and Iozzo 1980). The number of cord vessels were variable by the individual case (Marin-Padilla *et al.* 1981).

ACKNOWLEDGEMENTS

The authors would like to thank Drs. Il Hyang Ko (case 4), Jung Rye Kim (case 9), Byung Doo Lee (case 13), An Hee Lee (case 16) and other physicians for their submission of the cases. We also appreciate the technical assistance of Mr. Dae Bum Song and other members of our laboratory. Also we send our sincere thanks to all the parents of the cases used in this study for their understanding and cooperation.

REFERENCES

- Ahn GH, Chi JG, Lee JH, Lee SK, Kim HT, Chun KH. Conjoined twins-two autopsy cases report. Korean J. Pathol. 1969, 3:51-57
- Baik SH, Moon H. A case of conjoined twin (Dicephalus dipus tribrachius). J. Nat. Med. Center 1973, 8:93-95
- Chi JG, Lee YS, Ko IH. Conjoined twins (two autopsy cases) Environment. Mutag. Carcinog. 1981, 1:11-20
- Chi JG, Lee YS, Park YS, Chang KY. Fetus-in-fetu, report of a case. Am. J. Clin. Pathol. 1984, 82:115-119
- Chung KS, Park MY, Yoon JH, Kim KH, Lee HC. Thoracopagus. Korean J. Obstet. Gynecol. 1965, 8:47, cited by Kim 1970
- Edmonds LD, Layde PM. Conjoined twins in the United States, 1970-1977. Teratology 1982, 25:301-308
- Golladay ES, Doyne Williams G, Seibert JJ, Dungan WT, Shenefelt R. Dicephalus dipus conjoined twins: A surgical separation and review of previously reported cases. J. Pediat. Surg. 1982, 17:256-264
- Herring SW, Rowatt UF. Anatomy and embryology of cephalothoracopagus twins. Teratology 1981, 23:159-173
- Hong SH, Cho MH, Oh CH, Park YH, Chi JG. A case of thoracoventropagus. Eulji Med. J. 1980, 3:49-53
- Huh S. A case of conjoined twins (cephalothoracopagus). J. Chosen Med. Assoc. 1934, 24:228-235
- Izukawa T, Langford BS, Kidd L, Fred Moes CA, Tyrrell MJ, Ives EJ, Simpson JS, Shandling B. Assessment of cardiovascular system in conjoined thoracopagus twins. Am. J. Dis. Child. 1978, 132:19-24
- Kim CS. A case of conjoined thoracoventropagus twins. J. Chonnam Univ. Med. Coll. 1968, 5:135-137
- Kim IK. Anatomical observation of the cephalothoracopagus. J. Pusan Med. Coll. 1966, 6:231-235
- Kim JK, Kim JJ. Anatomical observation of conjoined twin. A case dicephalus tetrabrachius bispinalis dipus. J. Pusan Med. Coll. 1970, 10:71-78
- Kim JJ, Kang TH, Kim JH. A case of cephalothoracopagus. Chong Hap Med. J. 1967, 12:339-342
- Kim JR, Lee S, Kang JS, Lee HK, Shin SS, Chi JG. Cephalothoracopagus syncephalus-an autopsy case report. Korean J. Pathol. 1984, 18:89-93
- Leachman RD, Latson JR, Kohler CM, McNamara DG. Cardiovascular evaluation of conjoined twins. In Bergsma D (Ed) Conjoined twins, Birth defect original article series, Vol. 3. National Foundation-March of Dimes, Alan R. Liss, New York, 1967: pp. 52-62
- Lee KM, Park DK. Cephalothoracopagus monosymmetros synatus. Obstet. Gynecol. (in Japanese) 1934, cited by Kim 1970
- Mabogunje OA, Lawrie JH. Conjoined twins in West Africa. Arch. Dis. Child. 1980, 55:626-630
- Marin-Padilla M, Chin AJ, Marin-Padilla TM. Cardiovascular abnormalities in thoracopagus twins. Teratology 1981, 23:101-113
- Mudaliar AL. Double monsters. A study of the circulatory system and some other anatomical abnormalities and their complication in labor. J. Obstet. Gynecol. Br. Empire 1930, 37:753-768
- Nichols bL, Blattner RJ, Rudolph AJ. General clinical management of thoracopagus twins. In Bergsma D (Ed) Conjoined twins, Birth defect original article series, Vol. 3. National Foundation-March of Dimes, Alan R. Liss, New York, 1967: pp. 38-51
- Noh HI, Band CH, Lee KY, Lee HP, Jang JJ, Chi JG. Epignathus (pharyngeal teratoma)-An autopsy case report associated with renal agenesis. Korean J. Obstet. Gynecol. 1980, 23:989-994
- Noonan JA. Twins, conjoined twins and cardiac defects. Am. J. Dis. Child. 1978, 132:17-18
- Potter EL, Craig JM. Conjoined twins. In Pathology of the fetus and the infant, 3rd ed. Year Book Med. Publisher, Chicago, 1975: pp. 220-237
- Seo JW, Shin SS, Chi JG. Cardiovascular system in conjoined twins. An analysis of 14 Korean cases. Teratology 1985, 32:151-161
- Shin SS, Chi JG. Polysplenia in one of the conjoined twins. Seoul J. Med. 1983, 24:147-151
- Singer DB, Rosenberg HS. Pathologic studies of thoracopagus conjoined twins. In Bergsma D (Ed) Conjoined twins, Birth defect original article series, Vol. 3 National Foundation-March of Dimes, Alan R. Liss, New York, 1967: pp. 97-105
- Tan KL, Goon SM, Salmon Y, Wee JH. Conjoined twins. Acta Obstet. Gynecol. Scand. 1971, 50:373-380
- Wedberg RC, Kaplan G, Leopold G, Porreco R, Resnik R, Benirschke K. Cephalothoracopagus (Janiceps) twinning. Obstet. Gynecol. 1979, 54:392-396
- Wiegenestein L, Iozzo RV. Unusual findings in conjoined (Siamese) twin placenta. Am. J. Obstet. Gynecol. 1980, 137:744-745
- Ysander F. Studies on the morphology and morphogenesis of human thoracopagus monsters with special reference to the malformation of the heart. Almquist & Wiksells Boktryckeri, A.B., Uppsala, 1924, cited by Singer and Rosenberg 1967
- Yu ES, Shin SS, Ahn GH, Chi JG. Heteropagus (an autopsy case). Seoul J. Med. 1983, 24: 337-340
- Zimmermann AA. Embryologic and anatomic consideration of conjoined twins. In Bergsma D (Ed) Conjoined twins, Birth defect original article series, Vol 3, National Foundation-March of Diems, Alan R. Liss, New York, 1967: pp. 18-27

LEGENDS FOR FIGURES

- Fig. 1. Skeletal system of the conjoined twins.
A: Roentgenograph of dicephalus dipus dibrachius (case 7) shows duplications of skull and spine, and rudimentary midline structures, e.g., midline scapula and pelvic bones. The radiolucency of the skull (right one) and multiple fractures are due to the intrapartum events.
B: Roentgenograph of anecephalic cephalothoracopagus (case 9) shows single set of facial bones and duplication of the spine, arms and legs.
- Fig. 2. Union of the brain in craniopagus (case 8). Intermesencephalic connecting stalk (arrow) is seen.
- Fig. 3. The skull base of cephalothoracopagus janiceps (case 3). Interfrontal and interoccipital lines are seen in right angle, sella turcica being the center of symmetry. (arrow heads: foramina magna, F: frontal bones, T: temporal bones, O: occipital bones)
- Fig. 4. Digestive tracts of conjoined twins.
A: Thoracopagus (case 16) having a fused and dilated midgut.
B: Dicephalus (case 4) revealing two separate upper digestive tracts fused into single lumen at duodenum.
C: Cephalothoracopagus (case 9) exhibits upper single lumen beginning to duplicate at the ileum. (sto; stomach, du; duodenum, jej; jejunum, ile; ileum, M; Meckel's diverticulum, GB; gallbladder, P; pancreas, sp; spleen)
- Fig. 5. Posterior view of the genitourinary system of dicephalus dipus tetrabrachius (case 15). The case had four legs and had double numbers of kidneys, ureters, bladders and gonads. The urethra shows Y-shaped fusion. The kidneys of the right co-twin (asterisks) are polycystic and the urethra is atretic.

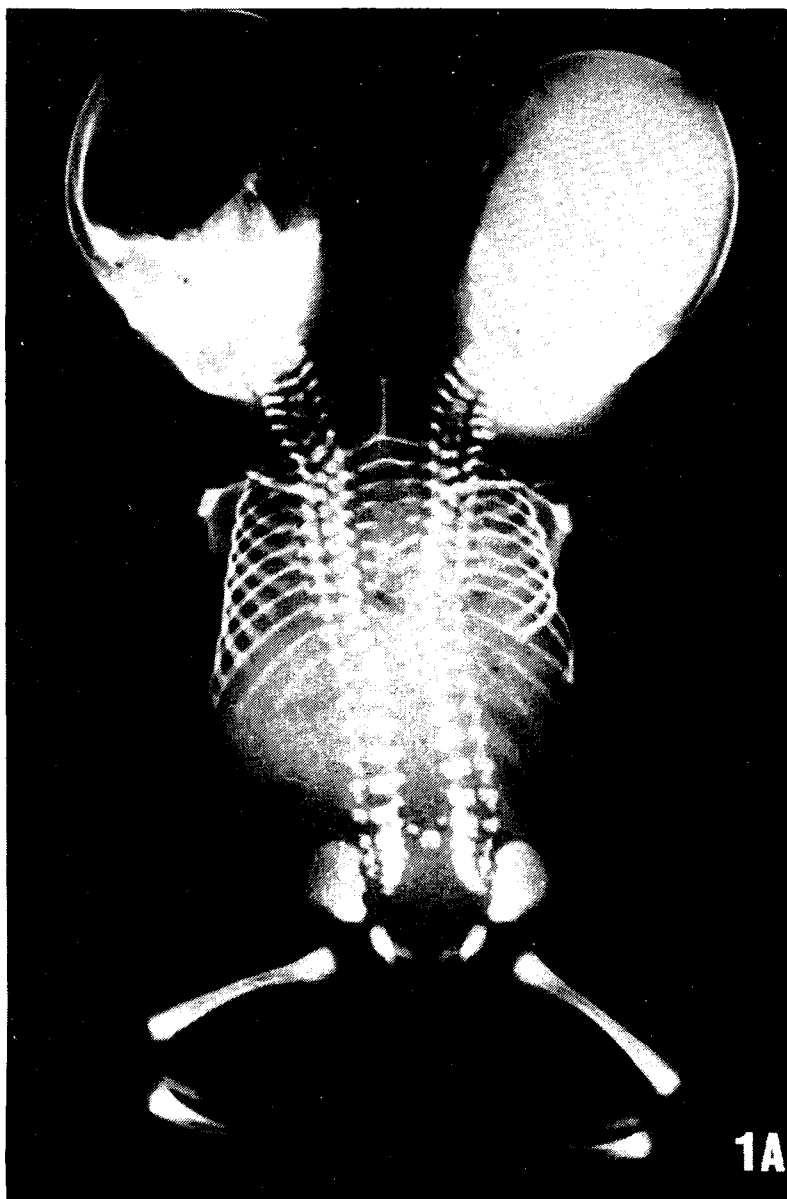
= 국문초록 =

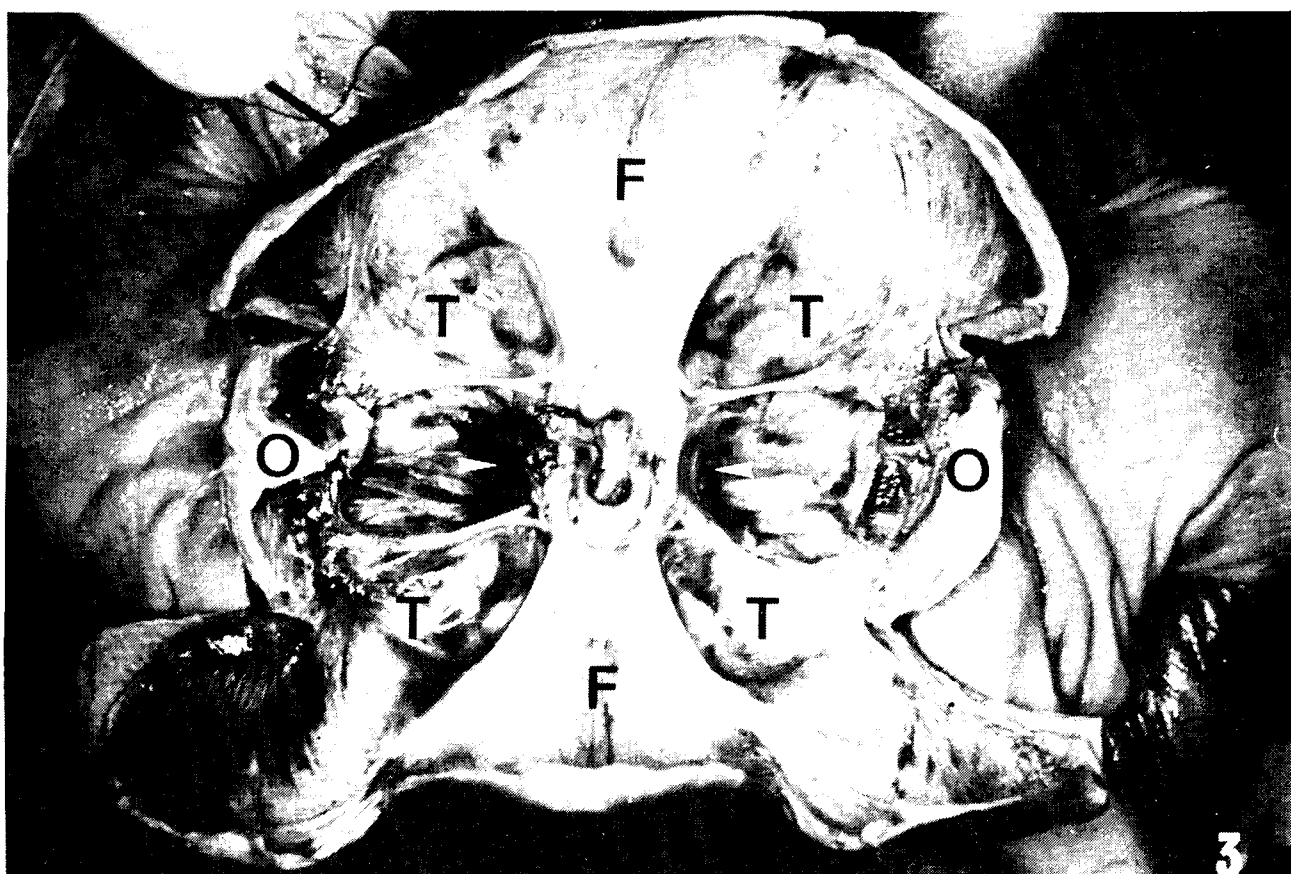
한국인 유합쌍태아의 형태학적 분석

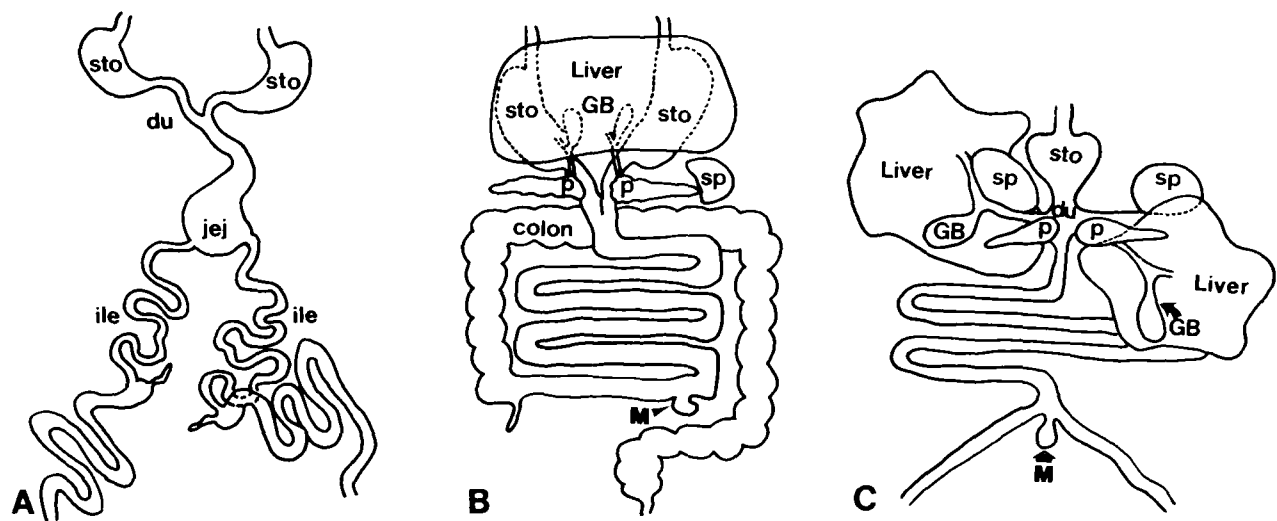
서울대학교 의과대학 병리학교실

지제근 · 서정옥 · 이해경 · 안공환 · 이윤성 · 신성식 · 유은실 · 김상윤 · 조경자

1967년부터 1985년까지 서울대학교 병리학교실에서 부검한 16예의 유합쌍태아를 형태학적으로 검색하고 이를 각장기와 외형적 유형별로 분석하였다. 외형적 유형은 흉부융합(5예), 쌍두형(4예), 두흉부융합(4예), 두부융합(1예), 쌍두쌍둔형(1예), 그리고 이종결합체(1예)로 분류되었다. 골격계의 형태는 외형적 유형과 합치되었으나 척수를 포함한 중추신경계는 전예에서 중복되었다. 심맥관계는 융합정도에 따라 5가지로 분류되었다. 소화관은 외형적 유형과 관련있었으나 융합과 분리의 위치는 증례에 따라 차이가 있었다. 흉복부가 융합된 쌍태아의 간은 두흉부융합 쌍태아를 제외하고는 융합되어 있었다. 비뇨생식기의 수는 다리의 수와 관련이 있었다. 제대혈관은 흉복부의 수와 관련있었으나 증례에 따라 차이가 있었다. 한편 저자들은 유합쌍태아의 융합과정에는 몇가지 기본원칙이 있음을 알수 있었다. 즉 유합쌍태아는 각도는 다르지만 서로 마주보는 형태로 융합하고, 융합된 장기는 전방을 향하는데 반하여 융합되지 않은 장기는 서로 마주보고 있으며, 마주보는 정도가 작은 쌍두형 유합쌍태아의 경우에는 쌍태아중 하나에서 장기의 위치이상이 동반되었다.







4

