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Cross-Sectional Illustration on Major Types of Conjoined Twins

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Conjoined twins show varying degree of conjonining in either facing or side-by-side fashion. Cephalothoracopagus janiceps is a prototype of facing anomaly in which the two bodies demonstrated a cross symmetry to the midline, that is axial symmetry. Interfacial and intersternal lines crossed at a right angle and no abnormality of situs was associated. Dicephalus dipus dibrachius is a case of side-by-side union, in which the bodies facing nearly the same direction were symmetrical to the middle sagittal plane. Abnormal situs of one was always associated. Other types of conjoined twins as thoracopagus lie between the two extremes of facing and side-by-side union. The three dimensional architectures of the organs in each type would be explained using cross sectional figures of skull, thorax and pelvis. Although the facing twins share the internal organs without fusion, the organs in the side-by-side component are fused with modification of the situs. We postulate sixteen pairs of situs and four manners of division for the explanation of the midline organs and the presence of a dominant co-twin. The splenic locations in a given cardiopulmonary situs are evaluated for the appraisal and applicability of these hypotheses.

Key Words: *Conjoined twins, heterotaxy, symmetry, computed tomogram, cephalothoracopagus, dicephalus, thoracopagus*

INTRODUCTION

It is generally accepted that the conjoined twins result not from the conjoining but from the failure in separation of monozygotic twins. But the traditional description of conjoin, union and fusion are widely used. This rare condition of fusion of organs is invaluable in the analysis of symmetry and organ formation (Noonan, 1978). The morphology and the mode of fusion are mutually related. Two bodies can be fused in the symmetrical or asymmetrical alignment. Side-by-side, back-to-back and facing fusions are the possible modes in the symmetrical alignments. Chi et al. (1986) suggested two independent components of the fusion process after analysing 16 conjoined twins in Korea. The conjoined twins were considered as a mixture of facing and side-by-side fusion. The cardiovascular system was the major topic in the analysis of conjoined twins (Izukawa et

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al., 1978; Leachman et al., 1967) and a frequent association of abnormal situs in side-by-side fusion was reported (Seo et al., 1985; Shin and Chi, 1983).

Cross-sectional drawings at the levels of head, thorax and pelvis are made from the morphological anlaysis of organ in three major types conjoined twins, i.e., cephalothoracopagus, dicephalus and thoracopagus. Lungs, hearts, livers and spleens were carefully examined to evaluate the symmetry and chirality of the bodies. In this study we suggest principles of fusion of organ systems in conjoined twins. They are sixteen possible pairs of situs and four probable manners of division which are appraised from a morphological observation on the cardiopulmonary situs and spleen.

MATERIALS AND METHODS

Four cases of cephalothoracopagus, four cases of dicephalus and five cases of thoracopagus were used in this study (Table 1), from the sixteen pairs of conjoined twins in the autopsy files of Seoul National University Hospital from 1967 to 1985 (Chi et al., 1986). All the cases were symmetrical conjoined twins and had typical features of their external morphological types. Dicephalus dipygus, craniopagus and heteropagus were excluded in this study.

The internal organs of the three types of conjonined twins were analyzed with special refrence to the sidedness, symmetry and three dimensional alignment in the body. The lungs, hearts, livers, spleens and body skeleton were carefully examined. The three dimensional architecture and symmetry of internal organs were explained by cross sectional drawings.

Hypothetical lines of division in possible pairs of situs were postulated and the hypotheses are assessed from the autopsy findings.

Terminologies in the description of situs and sidedness should be determined by the standard anatomical position of body. In conjoined twins two types of anatomical positions are defined. One is based on the position of each cotwin and the other, the portion of part of conjoined bodies. The anatomical position of symmetrical conjoined twins is defined as follows. If there is a single common chest that is defined as the anterior. If two common chests are present larger one is defined as anterior one, and

Table 1. Clinico-pathological summary of cases.

Type Case no.	subtype	Sex/ Age 1)	Gestational period (wk)	Body 2) weight (gm)	Maternal age (yr)	Fused organs (3)
Cephalothoracopagus						
1 (A67-25)	syncephalus	F/ND	32	est. 3,000	31	Sk, GI, Lv
2 (RCM-335)	janiceps	M/ND	24	1,120	26	Sk, CNS, GI, Lv
3 (A81-45)	syncephalus	F/SB	32	est. 2,500	?	Sk, GI, Lv
4 (A82-75)	syncephalus	M/ND	30	1,000	23	Sk, GI, Lv
Dicephalus						
5 (A69-30)	dipus tribrachius	M/SB	36	3,500	?	Sk, GI, Lv, Ge, Ur
6 (RCM-341)	dipus dibrachius	F/ND	36	3,350	26	Sk, Ht, GI, Lv, Ge, Ur
7 (A81-66)	dipus dibrachius	F/SB	36	3,900	28	Sk, Ht, GI, Lv, Ge, Ur
8 (A85-5)	dipus tetrabrachius	M/ND	36	3,800	27	Sk, GI, Lv, Ge, Ur
Thoracopagus						
9 (A81-54)		M/SB	35	3,400	29	Sk, Ht, Lv
10 (A83-66)		F/ND	?	2,500	?	Sk, Ht, Lv, GI (?)
11 (A83-120)		M/SB	?	2,300	?	Sk, Ht, Lv, GI (?)
12 (A83-121)		F/SB	?	2,800	?	Sk, Ht, Lv, GI (?)
13 (A85-1)		M/SB	32	2,500	26	Sk, Ht, GI

1) ND: neonatal death, SB: still birth
2) est: estimation
3) Sk: skeletal system, CNS: central nervous system, Ht: heart, GI: gastro-intestinal system, Lv: liver, Ge: Genital system, Ur: urinary system.

smaller one, posterior chest. The left and the right are determined thereafter. Two anatomical positions are same or similar in the cases with conjunction in side-by-side pattern. But they are different or in the right angle to each other in the cases of facing union. The general descriptions such as anterior and posterior, left and right, and midline are used along with specification of the type of anatomical position. However, description related to the anatomical structures such as sagittal, coronal, frontal, etc. are used only in the description of each cotwin.

In two dimensional diagram, there are two types of symmetry. One is line symmetry as a letter "A". Point symmetry is symmetricity to a central point as a letter "S". Some figures such as "O" and "X" are symmetrical both to the vertical and horizontal lines and to the central point. In three dimensional body, plane symmetry is a type of mirror image. Axial symmetry in three dimension is an equivalent of point symmetry in two dimension. These relationships are easily applied in the cross sectional analysis of symmetry in the body.

In the cross sectional diagram of the head, sagittal, coronal, fronto-occipital (F-O), fronto-frontal (F-F), and occipito-occipital (O-O) lines are key lines. the important lines in thoracic cross section are sterno-vertebral (St-V), sterno-sternal (St-St), and intervertebral (V-V) lines. The pelvic cross section shows pubo-sacral (P-Sc), pubo-pubal (P-P), and sacro-sacral (Sc-Sc) lines. These lines could be extrapolated into the planes in three dimensional body.

RESULTS

Cephalothoracopagus has four arms, four legs and single head and trunk. Arms, legs and the trunk are symmetrical to the central longitudinal line. Two hearts reside in the anterior and posterior thoraces with their apices in opposite directions. There are four lungs. No abnormality of situs is associated. The skull is symmetrical to the central antero-posterior plane in cephalothoracopagus syncephalus (cases 1, 3 and 4). They have single face and two occipital bones. Two fronto-occipital (F-O) planes meet an angle about 30-60'. The skull of cephalothoracopagus janiceps (case 2) is symmetrical to both the antero-posterior and the left-right planes. And also the skull is symmetrical to the axis at the sella turcica. The plane through the midlines of the anterior and posterior faces (F-F plane) is in the right angle to the plane through the midlines of the posterior occipital bones (O-O plane). Two F-O planes also cross in the right angle.

The cross sectional figures of cephalothoracopagus syncephalus (fig. 1A) shows a single frontal face, two lateral and two posterior ears and their petrous portions of temporal bones. Single pair of anterior and middle cranial fossae, two posterior cranial fossae with two foramina magna and central sella turcica are also present. The thorax contains two separate sets of heart and lungs. Cardiac apices directed to the opposite sides. Abnormality in the situs is not associated. The thoracic organs are symmetrical to a central axis. Pairs of the lungs and limbs belong to either left or right co-twins, whereas both equally share the hearts, anterior and posterior faces.

Four cases of dicephalus twins have single pair of legs and two heads. The number of arms varies as two (cases 6 and 7), three (case 5), or four (case 4). Two separate skulls were symmetrical to the central antero-posterior plane. The angle between two fronto-occipital (F-O) planes changes with the number of arms; widest in tetrabrachius and almost same direction in dibrachius. The sterno-vertebral (St-V) planes of the conjoined thoraces are similar to that of fronto-occipital (F-O) planes. The cardiopulmonary situs of the cases were inversus of right cotwin (case 6), right isomerism of right (case 8), inversus of left cotwin (case 5), and left isomerism of left (case 7). All cases had same inner sides; right in two cases and left in other two cases.

Dicephalus (Fig. 1B) is in the extreme end of the side-by-side union, although it is not a complete one. The single frontal thorax is composed of two vertebral columns, ribs and a single sternum. The common trunk and pubic bones are in frontal planes, whereas the heads, vertebrae and sacra rotate slightly inward. Co-twins A and B are summetrical to a central line in the figure and to the central plane in the three dimensional body; abnormal situs is most likely accompanied. Figure 1B shows situs inversus of right co-twin A and right atrial fusion of heart in mirror image.

Five cases of thoracopagus have common features of fusion in the anterior chest in oblique direction. There are two sterna. One was in the anterior chest and the other smaller one was in the posterior chest. Single conjoined heart was in the anterior chest.

Thoracopagus (Fig. 1C) is a mixture of facing and side-by-side union, The fused heart is in the frontal plane and the heads, verterbrae and pelves in the oblique directions. The angles between the St-V

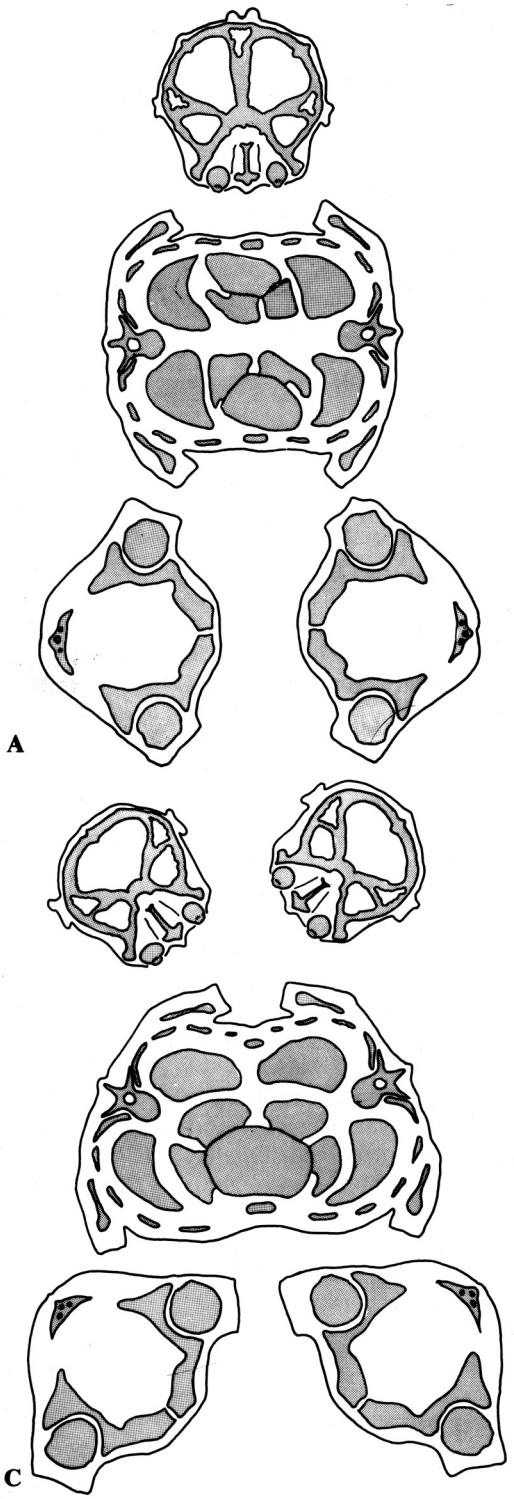

Fig. 1. Cross sectional illustrations at levels of head, thorax and pelvis.
A: Cephalothoracopagus syncephalus.
B: Dicephalus dipus dibrachius.
C: Thoracopagus.

lines through two sterna are different. And the angle between the St-V lines through anterior sternum is narrower than that of cephalothoracopagus, and is wider than that of dicephalus. The wide anterior thorax contains the conjoined heart, whereas the posterior thorax has only two lungs. Abnormalities of situs may or may not be associated.

The splenic location and numbers are studied in five of six cases with abnormal situs (Table 2). Four cases have single left spleen, and one case had left multiple spleen.

DISCUSSION

The conjoined twins could be categorized into three groups (Zimmermann, 1967). The first group cephalothoracopagus is terata anadidyma in which

the upper portion of its body is single and the lower portion, double (Herring and Rowlatt, 1981). The second group dicephalus is terata catadidyma in which the lower portion is single and the upper, double. The third group thoracopagus is terata anacatadidyma in which the trunk is fused and both the upper and lower, double (Singer and Rosenberg, 1967). The three types were in the majority in our series as in other reports (Chi et al., 1986; Edmonds, 1982).

From the findings, we were able to formulate and propose three rules of fusion. The first rule is that conjoined twins lies in the spectrum between facing union and side-by-side union. Hence they could be listed orderly from the facing extreme to the side-by-side as follows: cephalothoracopagus janiceps, cephalothoracopagus syncephalus, thoracopagus without abnormal situs, thoracopagus with abnormal situs, dicephalus dipus tetrabrachius, dicephalus dipus tribrachius, and dicephalus dipus dibrachius. The second is that the fused organ is in the frontal plane and the rest of the bodies, in the lateral planes. Accordingly, the face in cephalothoracopagus, the fused heart in thoracopagus, and the fused trunk in dicephalus are in their frontal planes. The third is that the same inner sidedness is a feature of a side-by-side union and midline organs are shared by both left and right cotwins. The midline organs are not necessarily fused organs. Anterior and posterior hearts in cephalothoracopagus are midline organs and both are shared by both cotwins, but they are are not fused. In an attempt to understand this sharing phenomenon, an approach is made from a geometrical point of view, whereby the sharing phenomenon might be explained by point symmetry in a two dimensional diagram and line symmetry in a three dimensional body; such principles of symmetry can readily observed at a natural biochemical phenomena, such as the resonance of phenyl group or the racemization of optical isomers.

In addition to the aforementioned principles of fusion, we further suggest possible modes of situs

Table 2. Possible pairs of situs in a conjoined twins.

Situs of right cotwin	Code*	Situs of left cotwin	No. of cases (case no.)
1) Single anomaly and same inner side			
Solitus	(RL — LR)	Inversus	1 (case 5)
Solitus	(RL — LL)	Polysplenia	2 (cases 7, 11)
Inversus	(LR — RL)	Solitus	1 (case 6)
Asplenia	(RR — RL)	Solitus	2 (cases 8, 10)
2) Double anomaly and same inner side			
Inversus	(LR — RR)	Asplenia	0
Polysplenia	(LL — LR)	Inversus	0
Polysplenia	(LL — LL)	Polysplenia	0
Asplenia	(RR — RR)	Asplenia	0
3) No anomaly and different inner side			
Solitus	(RL-RL)	Solitus	7
4) Single anomaly and different inner side			
Polysplenia	(LL-RL)	Solitus	0
Solitus	(RL-RR)	Asplenia	0
5) Double anowaly and different inner side			
Inversus	(LR-LR)	Inversus	0
Polysplenia	(LL-RR)	Asplenia	0
Inversus	(LR-LL)	Polysplenia	0
Asplenia	(RR-LR)	Inversus	0
Asplenia	(RR-LL)	Polysplenia	0

* Situs of each cotwin, RL means situs solitus, LR means situs inversus, RR means right isomerism (asplenia) and LL means left isomerism (polysplenia).

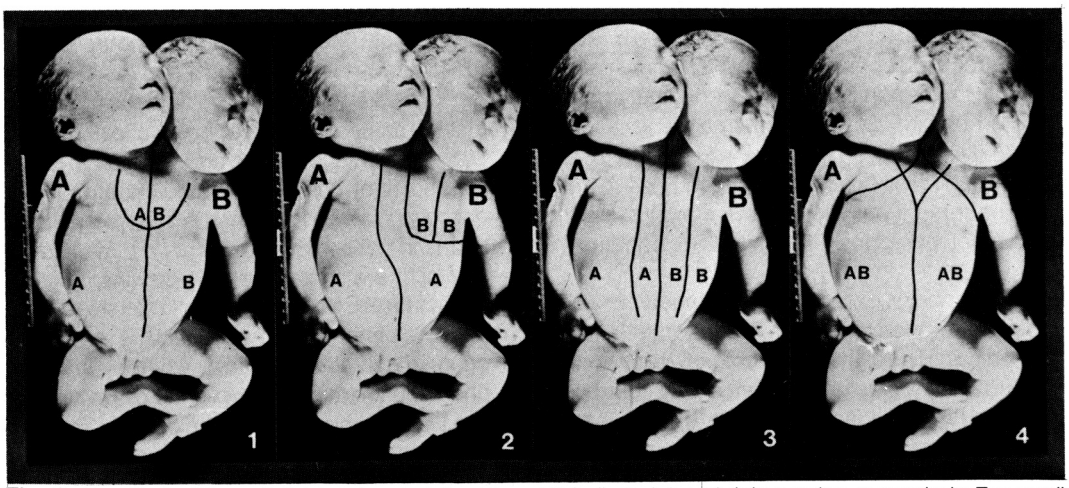

Fig. 2. Hypothetical lines of division. Larger letters A and B indicate left and right cotwins, respectively. Two small A's (or B's) are left and right sides of cotwin A (or B).

and manners of division. In a normal person, there are four types of situs; situs solitus, situs inversus, right isomerism and left isomerism. One pair of conjoined twins can therefore have sixteen possible modes of situs (Table 2). In category one, there are four modes of single anomaly and same inner side. Situs inversus or left isomerism of a left co-twin would make both inner sides left, and situs inversus or right isomerism of right would make both inner sides right. In category two, there are four modes of double anomalies and same inner side and in category three, eight modes of different inner sides including a mode with no anomaly at all. All of the four dicephalic twins and two thoracopagus were in the category one. The other three thoracopagus and all cephalothoracopagus had normal situs in both.

Taking into consideration the above possible modes of situs, we can then draw hypothetical lines of division or union in conjoined twins (Fig. 2). There are four possibilities. The first hypothesis is that the division occurs in the midline and fusion of both sides in the thorax, and fusion of outer sides in the abdomen. The second hypothesis is that the whole lower portion of the body is occupied by the dominant co-twin. According to this hypothesis, symmetrical conjoined twins are considered to be a variant of heteropagus. The third hypothesis is that the division occurs in the midline and the formation of the whole body is contributed by both the inner and outer sides. In the last hypothesis, the thorax and abdomen are postulated to have an equal sharing by both co-twins.

Thus far we have been able to categorize the various modes of situs and formulate the possible lines of division or union into which falls the modes of situs. We now attempt to explain the three groups of conjoined twins in terms of the aforementioned hypothesis. Cephalothoracopagus seems to be best explained by hypothesis four. In side-by-side conjunctions, however, it is difficult to choose a single best explanation. One of the ways to better evaluate such cases of conjunctions is to examine the location and number of the spleens. For example, suppose the right co-twin A is a situs solitus, and the left co-twin B situs inversus. Then, the spleen of the right co-twin would be in the left flank, and the spleen of left co-twin B in the right. According to the first hypothesis, the left abdomen of co-twin A and the right abdomen of co-twin B normally containing the spleens would not be apparent. The spleen would then be absent in this type of conjunction. By the second hypothesis, the single left spleen would be that of the dominant co-twin A. By the third hypothesis, there would be two spleens in the center. Finally, according to the fourth hypothesis, there would be two spleens in the left sides of each of the co-twins.

The hypotheses compatible with the splenic locations are listed in a given cardiopulmonary situs (Table 3). Hypothesis two, dominant cotwin hypothesis, is unanimously approved based on the data of limited observed cases. The dominant cotwin in

Table 3. Cardiopulmonary situs and spleen.

External type	Case	Situs	Spleen	Hypotheses*
Diceph. dipus dibrach.	6	(LR — RL)	Lt, single	2
Diceph. dipus tetrabra.	8	(RR — RL)	Lt, single	1,2,3,4
Thoracopagus	10	(RR — RL)	Lt, single	1,2,3,4
Diceph. dipus tribrach.	5	(RL — LR)	Lt, single	2
Diceph. dipus dibrach.	7	(RL — LL)	Lt, polysplenia	1,2,4
Thoracopagus	11	(RL — LL)	Not examined	?

*Numbers of approved hypotheses on possible line of division.

that hypothesis may be either left or right cotwin and the high rank of approval might be biased by the duality. Further morphological analysis of cases of conjoined twins are needed to properly evaluate the validity of the proposal.

CONCLUSION

Conjoined twins are in the spectrum of disease with varying degrees of facing and side-by-side union. A facing union creates line symmetry, whereas side-by-side union creates plane symmetry. The conjoined portion is shared by both co-twins in line symmetry, and a dominant co-twin may be present in side-by-side union.

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