ABNORMAL UMBILICAL VESSELS AND SYSTEMIC CIRCULATORY REVERSAL IN THORACOPAGUS TWINS

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A human thoracopagus of slightly unequal twins having a common umbilical cord with exomphalos is described, with special reference to the cardiovascular system. The larger twin had single umbilical artery. The smaller twin had a large left umbilical artery which was in direct continuity with the aorta, a small right umbilical artery with sole connection to the right external and internal iliac arteries (right common iliac artery was absent), missing umbilical vein, a rudimentary nonfunctional heart with atresia of the truncus arteriosus, and a right-sided aortic arch. Exomphalos of both twins is accounted by the possible defective growth of umbilical mesoderm caused by abnormal umbilical vessels. Reversal of systemic circulation in the upper part of the body of smaller twin is discussed. The importance of a careful study of umbilical veins is emphasized.

The higher incidence of single umbilical artery (SUA) in twins than in singletons (Bernirschke and Bourne 1960, Benirschke et al. 1964, Kristoffersen 1969), and uniform occurrence of SUA in acardius monsters (Slipka and Kocova 1970) are suggestive of an intimate relation between the vascular anomaly and twin births. About 90% of the monochorial placentae show some sort of connection between the fetal circulations of the twins (Strong and Corney 1967), and 15-30% of monochorial twins suffer from the transfusion syndrome (arterio-venous shunt in placenta) to a greater or lesser degree (Rausen et al. 1959, Strong and Corney 1967, Benirschke and Driscoll 1967). The arterial connection running between the two cords on the surface of the placenta, producing reversed circulation in SUA of acardiac twin, is blamed for the gross malformation of the acardiac variety (Willis 1958, Slipka and Kocova 1970); although evidence for mechanical compression of the embryo at the embryonic disc stage (Krause and Bejdl 1948), and for primary failure of the parts to develop adequately (Alderman 1973) has also been presented to explain the origin of acardiac monsters. The genesis of SUA (Monie 1970) has been worked out, and the cause-effect relation between SUA and its associated developmental defects (Chaurasia 1974) has been demonstrated.

Thoracopagi are known for extreme variation in their circulatory system (Morison 1970). Fusion of the heart chambers was described by Ysander (1924), Vintemberger (1926-27), Mortimer et al. (1941) and Ojala et al. (1968). A common heart was observed by Trzeciak and Dmitriew (1967) in a human thoracopagus, and by Kanan (1970) in an unequal thoracopagic goat twin in which the aorta supplied the abnormal smaller twin, and the pulmonary artery replaced aorta in the normal larger twin. Vascular connections between the abdominal

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aortae of such twins (Boni and De Camillis 1949) have also been demonstrated. However, abnormalities of the umbilical vein, and a rudimentary heart associated with reversal of the systemic circulation in the upper part of the body have not been reported in this monstrosity. For this reason, the following account of cardiovascular anomalies in a thoracopagic monster is of special interest.

CASE REPORT

A male thoracopagus (Fig. 1) having single umbilical cord with exomphalos was stillborn on 1st August 1974 to a healthy primigravida aged 20 years, at 32 weeks of pregnancy. There was no history of any illness nor any medication during first trimester of pregnancy. The parents are nonconsanguineous and the family history is noncontributory.

One twin was larger (A) than the other one (B). Both of them weighed 582 g. The sitting height of twin A was 15.5 cm and that of twin B, 11.7 cm. The common umbilical cord was short (17.7 cm), irregularly sacculated, and contained one large vein and three arteries of different sizes (Fig. 2). The placental end of the cord was narrow (4 mm) and normally inserted. Placenta weighed 205 g and was 11.0 to 11.5 cm in diameter. Ventral fusion between the two twins extended superiorly to a point 18 mm below the sternal notch of twin A and 5 mm below the sternal notch of twin B, and inferiorly to a point 25 mm above the base of the phallus of twin A and 16 mm above the corresponding point of twin B. The lower part of the fused area was occupied by exomphalos on one side (toward the right side of twin A) and fetal end of the cord on the other side. The exomphalos contained small and large intestines of both twins and a part of the common liver which appeared shaggy and shreded (Fig. 1), possibly due to unavoidable wearing before fixation in 20% formalin. Foetal attachment of the cord covered a wide area measuring 4.3 cm from A to B twins. External examination of twin B for additional malformations revealed a short neck, left scoliosis, curled up left auricle, phocomelia of right upper limb, and varus deformity of both hands with pedunculated thumb on right side and a missing thumb on left side. The larger twin A was comparatively normal but for bilateral club foot.

Radiographically, the right humerus and both radiuses of twin B were absent; the left first metacarpal and skeletons of both thumbs in the same twin were also missing.

Dissection of the umbilical cord showed that twin A had single umbilical artery which was larger than either of the arteries of twin B, one of which was very small. There was only one umbilical vein belonging to twin A (Fig. 2). Separation of twins on one side, so that twin A was on the left side and twin B on the right side of the dissector, demonstrated a common pericardial cavity containing normal heart of twin A and a rudimentary heart of twin B, a common and intact diaphragm, and a common liver (Fig. 3). The umbilical vein was traced within the liver by removing the latter in bits. An arrangement of wide veins (Fig. 3), which could serve both twins, was found. Apart from the malrotation of gut in both twins, the digestive tracts with pancreas and spleen were separate and normal. The left lung of twin B was hypoplastic due to severe scoliosis. In addition, twin B had a horse-shoe kidney. The testes of both twins were intraabdominal.

The heart and great vessels of twin A (Fig. 6) were normal. However, the branching pattern of abdominal aorta deviated from the normal one. Inferiorly, the aorta appeared directly continuous with the single left umbilical artery.

The heart of twin B (Figs. 3, 4, 5 and 7) was represented by a smooth and thin-walled chamber with a right indentation; a rudimentary bifid and apparently muscular structure (primitive atria and ventricles) projected downwards and to the left from the front of the smooth chamber. The muscular structure was notched and shrivelled externally and encroached by a dense reticulum internally, which made it sponge-like; it gave rise from its left side to a thin and delicate cord (atretic truncus arteriosus) which was attached superiorly to the arch of aorta (Fig. 4). The thin-walled chamber communicated inferiorly with the inferior vena cava, on each side with one pulmonary vein, and superiorly with two bilateral and symmetrical channels resembling two horns of a sinus venosus. There was no communication between the rudimentary cardiac chamber and the aorta. Proximally, the arch of aorta, after a short vertical course on the left side of trachea (ductus arteriosus), was continuous with the pulmonary arteries. The arch was right-sided and passed posterior to the trachea. Initially it was narrow, but it gradually became wider with progressive branching. The descending

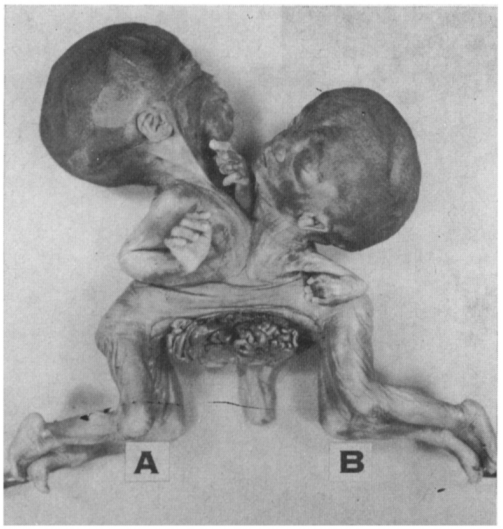

Fig. 1. Thoracopagic male twins having a common umbilical cord and a ruptured exomphalos. Twin B is smaller and malformed.

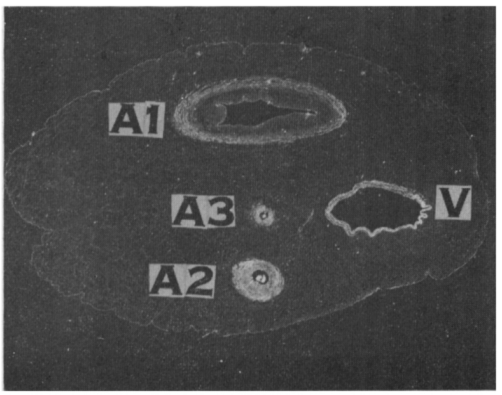

Fig. 2. Cross section of the umbilical cord of the thoracopagus showing one vein (V) and three arteries (A_1, A_2, A_3). A_1 is SUA of the larger twin; A_2 (left) and A_3 (right) belonged to the smaller twin.

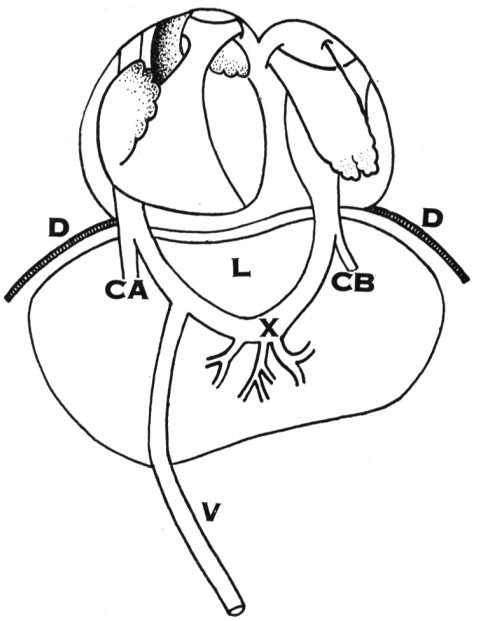

Fig. 3. Schematic representation of the venous anastomosis (X) between the two twins within the common liver (L). CA = inferior vena cava of twin A; CB = inferior vena cava of twin B; D = diaphragm.

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thoracic aorta was also right-sided. However, the abdominal aorta was normally placed, to the left of the inferior vena cava. The caudal end of aorta (Fig. 7) turned to its left side (left common iliac artery), gave rise to a small left external iliac artery and still smaller branches (internal iliac artery) to the pelvic viscera, and finally became continuous with the left umbilical artery, which was equal to the size of aorta. Right common iliac artery was missing; the right external iliac and right pelvic (internal iliac) arteries were continuous with the right umbilical artery. The coeliac, superior mesenteric and inferior mesenteric arteries originated from the upper part of the abdominal aorta close to one another, and emerged in the upper part of the median groove of horse-shoe kidney. Most of the abdominal aorta was overlapped by the fused kidneys which received two arteries in the lower part and one artery in the upper part, from the abdominal aorta.

DISCUSSION

Single umbilical artery of twin A and the left umbilical artery of twin B, which is comparable to a SUA, are both of a low type. Hence, their teratogenic effect on the caudal half of the body is minimum. The high type of SUA replaces abdominal aorta to a varying degree and is associated with gross malformations of the cloaca, the genitourinary, gastrointestinal, and central nervous systems, and of the lower limbs (Chaurasia 1974). The presence of exomphalos

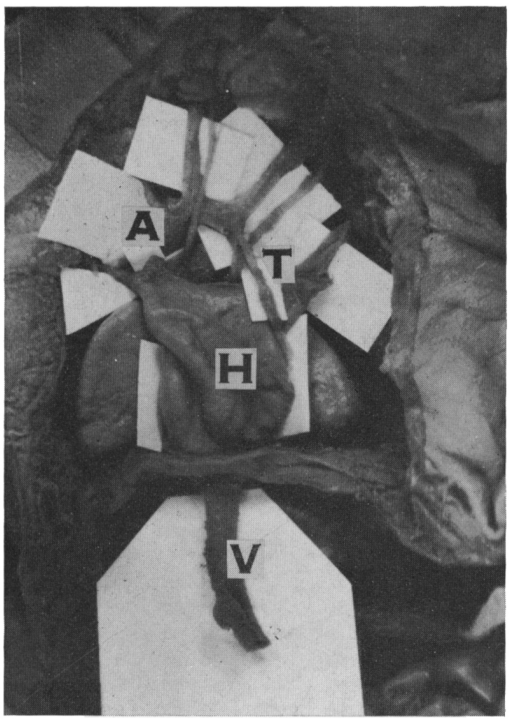

Fig. 4. Dissection of the smaller twin showing rudimentary heart (H), atretic truncus arteriosus (T), rightsided aortic arch (A), and the lungs on each side of the heart. Ductus venosus (V) a part of the venous anastomosis joined inferior vena cava before entering the heart.

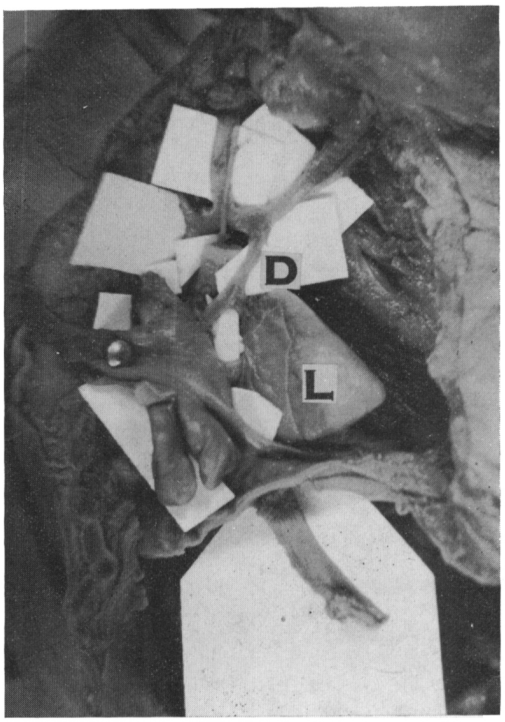

Fig. 5. Same as in Fig. 4 with the heart retracted to the right side to show the left pulmonary vein, and that the arch of aorta is connected proximally to the pulmonary arteries through ductus arteriosus (D). The left lung (L) should not be mistaken for heart.

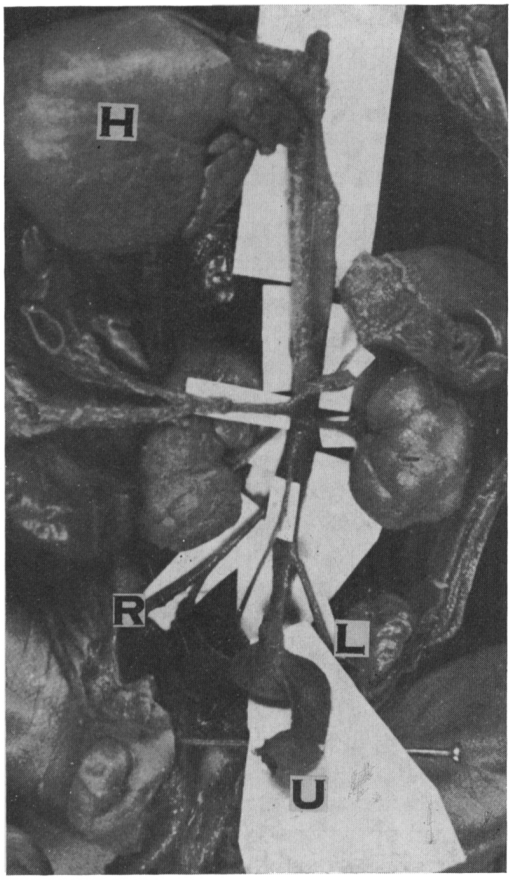

Fig. 6. Dissection of the normal aorta of the larger twin. Inferiorly, aorta is directly continuous with the SUA (U). Right (R) and left (L) external iliac arteries have been labelled. The origin of coeliac, superior mesenteric, inferior mesenteric and renal arteries is crowded in the upper part of the abdominal aorta. A well developed normal heart (H) is clearly visible.

Fig. 7. Dissection of the right-sided aorta in the smaller twin. Proximally, the aorta is narrow and continuous with the pulmonary arteries; distally, it is directly continuous with the left umbilical artery (LU). Right umbilical artery (RU) was continuous with the external and internal iliac arteries of that side. The coeliac (C), superior mesenteric (M), inferior mesenteric (I), renal (R), and left external iliac (L) arteries have been labelled. The insert in the right upper corner shows anterior view of the rudimentary heart with atretic truncus arteriosus in the right upper corner.

in the specimen presented here is explicable by a probable defective growth of umbilical mesoderm as a result of abnormal umbilical arteries, because the longitudinally disposed abdominal muscles may arise directly from the mesenchyme of the umbilical cord (Wyburn 1937). However, it is difficult to relate the cardiovascular abnormalities of twin B with its umbilical anomaly. Presence of right umbilical artery in twin B, in absence of any communication with the aorta is a curious finding. It may be that the flow of blood in this small channel was in reverse direction due to a possible anostomosis with any of the umbilical arteries either in

placenta or in the cord. One could assume a normal flow in this artery provided a veno-arterial shunt in the caudal half of the body of twin B was admitted, which appears less likely.

The absence of umbilical vein in twin B (Fig. 3) has more than one implication. First, that circulation of twin B is dependent on the umbilical vein of twin A for its supply of the oxygenated blood; second, that any inadequacy in this input quota would reduce twin B either to a mere parasite or a teratoma attached to the chest of twin A; third, that the left umbilical artery of twin B is bound to carry blood to the placenta — a reversed flow in this channel cannot be presumed. This emphasizes the importance of study of the umbilical veins as well. A negligence to this effect might lead to erroneous interpretations and wrong conclusions. Monie (1971) has drawn attention to certain abnormalities of the umbilical vein which have clinical significance.

The rudimentary nonfunctional heart of twin B is apparently a result of compression by the normal heart of twin A (Fig. 6) within the limited space of the common pericardial sac. It is feasible that pulsations of the normal heart of twin A provided the driving force for maintenance of the circulation in twin B. The efficiency of transmitted pulsations was possibly accentuated by the mirror-image pattern of the arch and descending thoracic aortae.

The rudimentary heart of twin B received oxygenated blood from the umbilical vein of twin A through venous connections in the common liver (Fig. 3). Since the heart had no connection with the aorta, the veins of head, neck and upper limbs, and the pulmonary veins, were the only alternative outlets. Deoxygenated blood from upper part of the body and from the lungs presumably returned to the arch of aorta through its branches and through pulmonary arteries. The thin-walled nature of the aorta and its gradual increase in size distally, provide evidence in favour of a reversed (veno-arterial) circulation in the upper half of the body of twin B. This reversal differs substantially from the popular one described in the SUA of acardiac monsters. It may be recalled that umbilical vessels correspond to the pulmonary vessels, and any circulatory reversal in these channels would therefore correspond to a reversal of pulmonary or lesser circulation. Reversal of the flow in SUA of an acardius all the same ensures distribution of blood to the body tissues through the usual arterial channels via aorta. In the present case (twin B), however, the reversal is in the systemic vessels rather than in the umbilical ones.

The position in the lower part of body of twin B appears normal. Blood collected in aorta from head, neck, upper limbs, and lungs, partly got distributed to the lower part of body which returned to heart through inferior vena cava, and partly passed on to the placenta through the left umbilical artery.

From the present observations and interpretations it may be concluded that abnormalities of the umbilical vein may assume equal or even greater importance than those of umbilical arteries, and that the scope of cardiovascular abnormalities in thoracopagic twins seems to be far greater than what is presently known. In general, it may be remarked that the primary role of the vascular abnormalities in producing developmental defects of other organ systems cannot be underestimated. Determination of the precise relation between the vascular anomalies and particular congenital defects calls for further studies on a wide malformed material combined with a comparative study of phylogenetic succession.

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RIASSUNTO

Vasi Ombelicali Anomali e Inversione Circolatoria in Gemelli Toracopagi

Viene descritto un caso di gemelli toracopagi con cordone ombelicale unico, lievemente diversi con particolare riguardo al sistema vascolare. Il gemello piú grosso presentava arteria ombelicale singola, mentre il piú piccolo presentava una grossa arteria ombelicale sinistra in diretta continuità con l'aorta, una piccola arteria ombelicale destra connessa soltanto alle arterie iliache esterna ed interna di destra (l'arteria iliaca comune destra era assente), ed in piú l'assenza della vena ombelicale, un cuore rudimentale non funzionale con atresia del tronco arterioso, e un arco aortico piegato a destra. La prominenza ombelicale può essere spiegata da un difettoso accrescimento del mesoderma ombelicale a causa dell'alterata vascolarizzazione. Viene inoltre discussa l'inversione circolatoria nel gemello piú piccolo e viene sottolineata l'importanza di un attento studio delle vene ombelicali.

RÉSUMÉ

Vaisseaux Ombilicaux Abnormaux et Inversion Circulatoire chez des Jumeaux Thoracopages

Description d'un cas de jumeaux thoracopages légèrement différents, avec cordon ombilical unique. Le plus gros des jumeaux présentait une artère ombilicale simple, alors que le plus petit présentait une grosse artère ombilicale gauche en continuité avec l'aorte, une petite artère ombilicale droite en connexion avec les artères iliaques externe et interne droites (l'artère iliaque commune droite étant absente) et en plus, l'absence de la veine ombilicale, un coeur rudimentaire non-fonctionnel avec atrésie du tronc artérieux, et un arc aortique vers la droite. La proéminence ombilicale peut être expliquée par un défaut de développement mésodermique ombilical à cause de la vascularisation altérée. L'inversion circulatoire dans le plus petit des jumeaux est discutée et l'importance d'un étude des veines ombilicales est soulignée.

ZUSAMMENFASSUNG

Anomale Nabelgefässe und Kreislaufinversion bei einem Thorakopagus

Beschreibung eines Thorakopagus mit nur einer Nabelschnur und leichten, vor allem das Gefässystem betreffenden Unterschieden zwischen den beiden Zwillingen. Der grössere Zwilling hatte nur eine einzige Nabelarterie. Bei dem kleineren hingegen war die linke Nabelarterie gross und ging direkt in die Aorta über, die rechte war klein und nur mit den rechten AA. iliacae (externa und interna) verbunden, während die gemeinsame rechte A. iliaca fehlte. Es fehlte auch die Nabelvene, das Herz war rudimentär und nicht funktionell mit Atresie des Arterienstammes (Truncus arter.), der Aortenbogen nach rechts verbogen. Das Hervorstehen des Nabels erklärt sich vielleicht daraus, dass das Nabelkeimblatt aufgrund der veränderten Gefässverhältnisse nicht regulär gewachsen ist.

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