A REPORT OF A CASE OF ISCHIOPAGUS TETRAPUS

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Ischiopagus tetrapus is an unusual type of diplopagus, a conjoined twin in which the components are equal and symmetrical. The connection occurs in the lower pelvic region with the axes of the bodies extending in a straight line in opposite directions. The bodies are fused in the region of the pelvis as far as the level of the common umbilicus, above which each is normally developed. Two legs extend at right angles from each lateral surface, those on the same side being derived one from each component, with urogenital and anal orifices opening between each pair. The two legs on one side may fuse into a composite limb, resulting in the form known as ischiopagus tripus. Most of the reported cases belonged to the latter variety and the vast majority of them were female (Taruffi, 1891; Gemmill, 1902; Schlumberger and Gotwals, 1945).

The mother of the case herein reported was a healthy 29-year-old Chinese multipara who had had three full-time normal spontaneous deliveries 8, 4 and 2 years previously respectively. All three children were girls, two of whom were living and well and one died of high fever at the age of three. The father was apparently well. There was no history of multiple pregnancy on either side. The mother was admitted to the hospital at a maturity of 26 weeks of pregnancy with a history of bleeding per vaginam and abdominal pain for one day. The conjoined twins aborted one hour after admission were dead but not macerated. The head and upper limbs of one twin were delivered first, followed by two lower limbs on one side of the body, then the lower limbs on the other side of the body and finally the upper limbs and head of the other twin. The twins were found in a common amniotic sac with a single umbilical cord which connected the twins to one circular placenta.

The ischiopagus tetrapus weighed 1,134 gm. The twins were equal in size and appeared almost symmetrical, the facial contours strongly resembling each other (Fig. 1). The only external difference was the presence of a big meningocoele in the cervical region of one foetus, which we designate as FA (abnormal foetus), the other apparently normal one as FB. FA and FB had separate head, neck, thorax and upper limbs. Anteriorly, they fused at the abdomen with two lower limbs extending out from each side of the flank. Between the legs on each side were a penis and an empty scrotum. The anterior abdominal wall was not developed but was replaced by a large exomphalos with a single umbilical cord, containing one umbilical vein and two umbilical arteries. The cord was attached to the margin of the exomphalos on the right side of FA (Fig. 1). Posteriorly, the mid-dorsal line of FA and FB terminated at a dimple and the two buttocks diverged at right angles to the longitudinal axis of the twins (Fig. 2). Anal openings, contaminated with meconium, were found one in each gluteal cleft. Talipes equinovarus was found in both feet (one belonging to each of the two twins) on the opposite side of the umbilical attachment.

The specimen was dissected through the anterior body wall. Two complete sets of alimentary tracts and accessory glands were demonstrated. A common loop of ileum, 8 cm. in length, was found immediately above the two caeca. There were four lobulated kidneys, each being covered with a suprarenal gland. The right ureter of FA and the left ureter of FB opened into the urinary bladder on one side and the left ureter of FA and the right ureter of FB into the bladder on the opposite side (Fig. 3 and Fig. 4). Four testicles were situated at the deep inguinal rings.

The diaphragm of FB was complete without any congenital anomaly whilst the left half of that of FA was absent with the stomach and the spleen lying directly underneath the left lung. All the lungs were solid and airless.

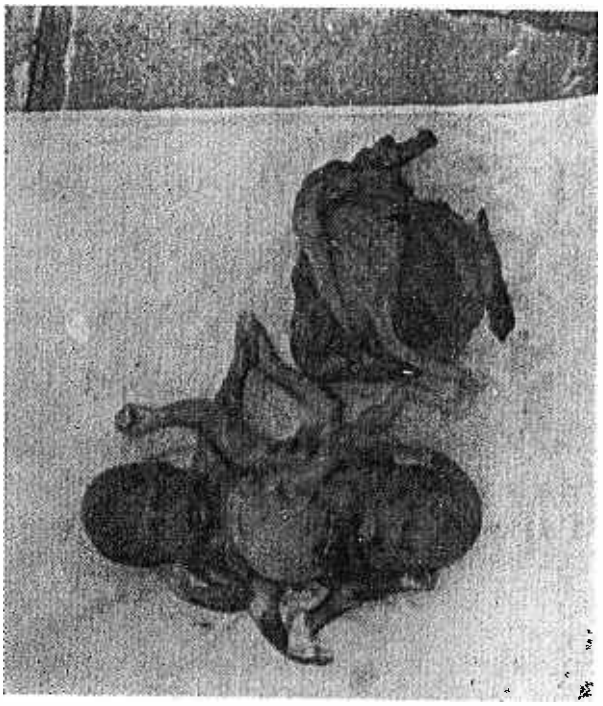

Fig. 1. Anterior view of the conjoined twins.

Fig. 2. Posterior view of the conjoined twins.

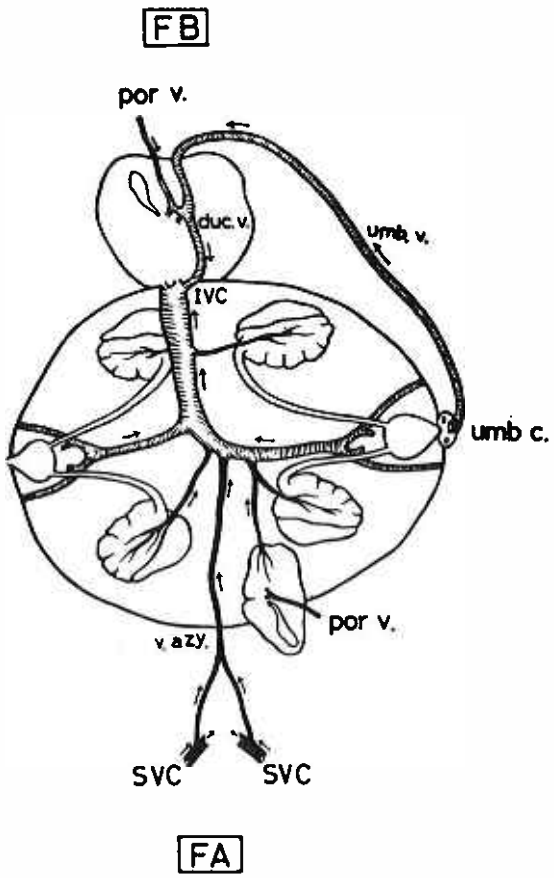

Fig. 3. The Venous System of the Trunk of the Conjoined Twins.

(Veins of FA — solid, veins of FB — cross-striped).
IVC — inferior vena cava, SVC — superior vena cava,
umb. c. — umbilical cord, umb. v. — umbilical vein,
v. azy. — vena azygos, por. v. — portal vein.
duc. v. — ductus venosus.

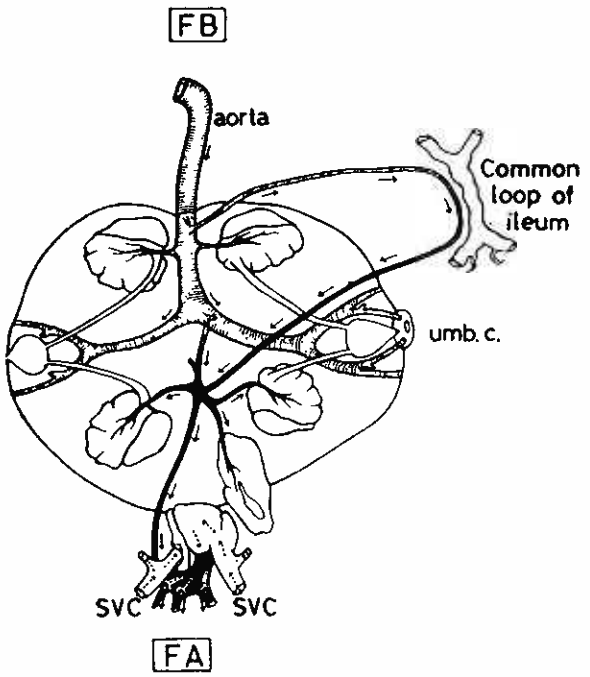

Fig. 4. The Arterial System of the Trunk of the Conjoined Twins.

(Arteries of FA — solid, arteries of FB — cross-striped). Arrows indicate the direction of blood flow, solid arrows (——→) show the blood flow in the arteries of FA and FB, and broken arrows (– – –→) show the venous blood flow in FA. SVC — superior vena cava. umb. c. — umbilical cord.

The heart, lungs, thymus and liver were very much larger in FB than in FA. The heart of FB, 7.1 gm. in weight and 3.2 cm. in width, had normal chambers and blood vessels. The heart of FA, only 2.6 gm. in weight and 2.2 cm. in width, had a tiny left ventricle which led to the pulmonary trunk and communicated with the right ventricle through a large interventricular septal defect. The right ventricle was about normal in size and opened to the aorta. The left atrium was also very small, with 4 pulmonary veins opening into it. The inferior vena cava (IVC) was absent. Two superior venae cavae (SVC) opened into the right atrium, the right SVC being normal in position whilst the left one draining into the much dilated coronary sinus underneath the left atrium.

The venous system could be traced clearly by the presence of the coloured latex which was injected through the umbilical vein. The sole umbilical vein circumscribed one-quarter of the margin of the exomphalos and opened into the portal vein, ductus venosus and IVC of FB. In the middle of the abdominal cavity, the IVC of FB received two big tributaries, each receiving blood from the pelvis and lower limbs of its own side. It also received drainage from all the kidneys and suprarenals, from the hepatic veins of both FA and FB and from the venae azygos and hemiazygos of FA, which connected the IVC of FB with the two SVC of FA (Fig. 3).

The descending abdominal aorta of FB bifurcated in the centre of the abdominal cavity to supply the pelvis and lower limbs of both sides. The two umbilical arteries arose from the internal iliac arteries on the same side of the umbilical cord. The aortic arch of FA, after giving off the three usual branches to the head, neck and upper limbs, abruptly tapered into a narrow descending aorta, the abdominal portion of which soon broke up into its terminal branches. The abdominal aortae of FA and FB supplied branches to their respective alimentary tract, urogenital organs and suprarenal glands. Two anastomotic channels were demonstrated between the two aortae: one through the superior mesenteric arteries supplying the common loop of ileum and the other through a more direct branch connecting the two inferior mesenteric arteries (Fig. 4).

A hypothesis for the possible pattern of blood circulation was deduced from the findings of the cardiovascular system, the assumption and reasonings being as follows: -

1. Since the sole umbilical vein was connected to the IVC and the heart of FB, the oxygenated blood could only reach FA by passing through the 2 arterial anastomoses between FB and FA. The direction of blood flow must have been reversed in the descending aorta and the aortic arch of FA as shown in Fig. 4.
2. All the veins of FA below the diaphragm drained directly into the IVC of FB. The blood from the head, neck and upper limbs of FA drained into both SVC and then partly into the right atrium and partly into the IVC of FB via the venae azygos and hemiazygos as shown in Fig. 3.
3. Judging from the difference in size and weight of the hearts of the two apparently equal-sized twins, it seemed plausible to assume that the oxygenated blood was sent to all parts of both FA and FB by the ventricles of FB. The ventricles of FA, the right one especially, must be functioning. However, the pressure in its ascending aorta must be considerably lower than that in its arch, which was maintained by the contraction of the heart of FB; and hence, the branches of the aortic arch of FA must have received blood from both hearts as shown in Fig. 4.

The posterior abdominal wall was exposed. Twelve thoracic vertebrae were found to be connected to 12 pairs of ribs for each foetus. Below the first lumbar vertebra, the vertebral columns of the two foetuses became confluent at a small median knob. Dorsal to the first lumbar vertebrae and the median knob was a large osteo-cartilaginous mass which probably represented the merged and undifferentiated second to fifth lumbar vertebrae. Only one sacrum was found in the pelvis on the same side as the attachment of the umbilical cord. The two vertebral canals were opened and found to communicate with each other on the umbilical cord side (or the sacral side) of the median knob. The lower ends of the two spinal cords in the vertebral canals met and

fused with each other. The cauda equina formed by the sacral nerves of both spinal cords occupied the sacral canal. Pairs of spinal nerves emerged from the spinal cords through the intervertebral foramina in the usual manner with the exception of the lower lumbar nerves which emerged together through the lumbar vertebral masses just above the sacrum. Four sacral plexuses were formed one on each side of the two pelvic cavities with each plexus supplying its appropriate lower limb.

COMMENTS

Symmetrical conjoined twins are rare. According to Potter (1957), only one such specimen has been delivered at the Chicago Lying-in Hospital since 1931, in the course of over 60,000 deliveries. Ischiopagus tetrapus is even rarer, the case reported here being the only one encountered at the Obstetrical and Gynaecological Unit, University of Hong Kong, since 1946, during which more than 100,000 deliveries have been undertaken.

Corner (1955) described three critical stages at which twins might have developed from a single ovum: (1) separation of the early blastomeres resulting in dichorionic twins, (2) duplication of the inner cell mass resulting in monochorionic diamniotic twins and (3) duplication of the embryonic rudiment of the germ disc resulting in monochorionic monoamniotic twins. Coulton et al (1947) and Salerno (1959) suggested that if the splitting of the germ disc occurred slightly later still, after the appearance of the primitive streak so that the single axial arrangement of the germ disc had been determined, then conjoined twins or monstrosities would be produced (usually after 13 days). Schlumberger and Gotwals (1945) observed that the symmetric diaxial conjoined twins always had a union of like parts, e.g. head to head, never head to buttock, and concluded that the developing embryos must have had parallel axes. They assumed that a duplication of the developing ovum by fission occurred sometime between the two-cell stage and the formation of the primitive streak. The subsequent union of the two individuals probably occurred during the first three weeks of development. Willis (1962), however, contended that from the sites and degrees of union exhibited, it was clear that most, probably all, conjoined twins and double monsters had the same essential mode of origin, namely, a coalescence of the closely contiguous twin embryonic areas overlapping each other within a single blastocyst. He took pains to emphasize that the coalescence of twins was not a secondary union of formerly quite separate organisms because of their close juxtaposition, but a primary union by overlapping of two embryonic fields. He postulated that two separate but closely adjacent axes (primitive streaks and Hensen's nodes) developed in the blastoderm forming the floor of a single amniotic cavity and that the two embryonic areas so destined overlapped to a varying extent. The site and extent of coalescence of the resulting twins depended on the orientation of the two axes with respect to one another and to the extent of the overlap of their fields

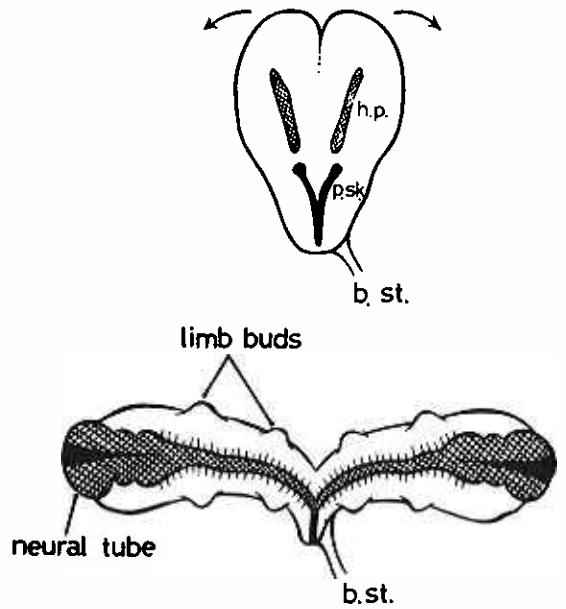

Fig. 5. Diagramatic presentation of the formation of the present ischiopagus tetrapus from incomplete fission of the embryonic disc.
Top — Early third week, splitting of the cranial portion of the primitive streak.
Bottom — Fourth week, formation of two neural tubes with common lower end.
b. st. — body stalk,
h. p. — head process.
p. sk. — primitive streak.

of influence. For the present case of ischiopagus tetrapus, basing on the finding of a common lower end of the vertebral columns and spinal cords, it would seem highly probable that the twins were formed as a result of incomplete fission of the embryonic disc after the appearance of the primitive streak. The

longer cephalic segment of the central axis split and turned away from each other until they were 180 degrees apart, whilst the shorter caudal segment remained unsplit and served as a common portion for both foetuses (Fig. 5).

The causes of abnormal development are either hereditary, environmental or combinations of these (Hamilton et al, 1952). In the present case, the conjoined twins shared identical hereditary and environmental conditions as they came from the same ovum and grew in the same amniotic cavity, but FB was apparently normal, while FA bore all the developmental abnormalities — cervical meningocoele, absence of left half of diaphragm and cardiovascular anomalies. The regions showing these malformations were all supplied by branches of the ascending aorta and the arch of the aorta of FA, which contained a mixture of venous blood from its own right ventricle with some oxygenated blood from FB. It would seem reasonable to suggest, therefore, that the hypoxia present in the tissues supplied by those arteries was responsible for the teratogenesis in FA. Ingalls et al (1952) were able to induce congenital malformations experimentally by hypoxia in mice. The talipes equinovarus present in the two feet on the same side of the conjoined twins (one belonging to each foetus) may be accountered for by the mechanical theory which rests on the supposition that the uterine wall comes in contact with and exerts pressure on the foetus (Gordon, 1961). Severe talipes equinovarus may be seen in association with congenital absence of sacrum (Franklin, 1963). In this case, the talipes was found on the side where the sacrum was absent.

SUMMARY

A case of ischiopagus tetrapus is presented and described in some detail. The incidence and mode of origin of conjoined twins and the aetiology of the congenital abnormalities which are found in this case are then briefly discussed.

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