

development of the allantoic circulation does not take place under such circumstances, a placenta and cord are not formed.

3. After the establishment of the allantoic or placental circulation. This period produces the forms usually included as acardii in the origin of which defects of the placental circulation play an important rôle. Mention has already been made of the fact established by Schatz, that the placental circulations of monozygotic twins are united by two forms of anastomoses—the constant communication effected by the chorionic villi common to the vessels of both fetuses and the more variable communication effected by the superficial anastomoses, at times limited to the umbilical arteries, at others involving both artery and vein, less frequently only the umbilical vein.

In exceptional cases, which may, however, occur at any time between the completion of the placental circulation and the middle of gestation, acardius may arise in consequence of primary heart-death followed by transfusion of the blood within the umbilical arteries of the stronger fetus into the corresponding vessels of the impaired twin in consequence of the diminished blood pressure within the latter. The blood thus contributed by the stronger fetus flows both to the placenta and to the body of the weaker; should the nourishment be insufficient to maintain both placenta and body, the one or the other suffers unless the meagre supply is equally shared.

Although in these comparatively rare cases the placental anastomoses, following the primary heart-death, directly produce the acardius, such communications in themselves are incapable of originating these malformations, the reversal of the blood stream being dependent upon a cause to which the failure of the heart and the subsequent change in the direction of the circulation are secondary.

The investigations of Schatz render it highly probable that this primary cause lies in obstruction along the course of the umbilical vein, either within or without the body of the fetus, at any point between the heart and the placenta. In consequence of the gradual narrowing of the vessel returning the blood from the placenta, the amount passing to the heart is correspondingly diminished until, sooner or later, this organ is more or less completely deprived of the supply necessary to maintain its nutrition and function. The result of this restriction is increasing impairment of the heart's strength and consequent diminution of the blood pressure within the umbilical arteries. When the resistance offered by the failing powers of the impoverished heart is reduced to a sufficient extent, the blood stream conveyed through the placental anastomosis from the stronger twin overcomes that within the weaker fetus and reversal of the circulation within the latter gradually takes place. In consequence of this reversal the weaker heart, sooner or later, is brought to a standstill and an acardius results. The rapidity with which these changes are effected depends upon the rapidity and the extent with which the venous obstruction develops. Although the superficial placental anastomoses suffice to compensate for inequality in the transfusion taking place between the twins, such communications are powerless to neutralize the effects of the hindrance caused by stenosis of the umbilical vein. On the contrary, they aggravate the untoward transfusion inequality, since the superficial venous anastomosis offers a ready escape for the retarded blood current into the circulation of the unaffected fetus, and the arterial anastomosis provides the channel by which the blood from the vigorous heart passes into the reversed circulation of the acardius.

After the establishment of the acardius, the obstruction within the umbilical vein may remain stationary, or after a time it may decrease, since the returning blood current, which now passes along the vein, is under higher pressure than before the assumption of the circulation by the stronger heart. In those cases in which the narrowing of the umbilical vein is permanent, at a time very shortly after the appearance of the budding limb,

the meagre supply of blood suffices for the imperfect development of the embryo into the globular amorphous type of acardius, in which the retarded return of the blood from the fetus results in edema and failure of development of the internal viscera. When the obstruction appears relatively late, the deficient development may result in producing an oedematous acephalus instead of the more typical amorphous.

It may be emphasized that, in general, the acardii are formed from normal embryos, and, further, that the time of their production may be at any period of gestation, from the early relations of the embryonic areas resulting in the development of a parasitic acardius, on the one hand, to the conversion of a hemiacardius into an acardius near the close of pregnancy on the other.

Although at times the reversed blood stream and the impaired heart are the only defects, in other cases conspicuous malformations are associated with the acardia. These defects may be grouped as *primary* and *secondary*, the former including such as exist prior to the acardia, the latter after its appearance.

The primary defects, again, embrace those which are *intrinsic* and those which are *accidental*. Among the intrinsic primary defects may be mentioned irregularities in the development of the venous system associated with *situs transversus*, absence of the ductus venosus, faulty formation of the liver and its veins, or malformation and stenosis of the umbilical vein. The accidental primary defects are such as have no connection with the condition of acardia, as *spina bifida*.

Regarding the grouping and the fundamental circulatory conditions of the several types of acardiac monsters, the conclusions of Schatz may be followed with advantage.

I. *Acardii completi*, in which a fairly well-developed trunk and head are present, the limbs being represented by the full complement or by the members remaining after the suppression of one or both upper or lower extremities. The various degrees of development of the head are expressed by the terms *holo-*, *para-*, *oma-*, and *hemicephalic*. Since the different types of acardii depend for their production largely upon the amount and character of the blood supply derived from the stronger twin, it is of interest to consider the probable relations existing in the complete acardii. Bearing in mind the fact that, even under the most favorable conditions, the nutritive requirements of the dependent twin must always remain insufficient, it will be appreciated that in order to secure the degree of development sometimes seen in the complete acardii, the fetus has enjoyed the most advantageous conditions possible under the circumstances. This implies an approximately sufficient nutrition by means of a well-balanced circulation, the blood stream entering by the umbilical arteries being carried away from the fetus by venous channels of adequate capacity to insure the necessary blood pressure, and at the same time to prevent stasis. The general development of the acardius depends upon the sufficiency of the blood supply forced into its various parts by the active heart; the occasional instances in which the affected fetus is relatively well-formed are possible because of the ample and direct anastomosis between the placental circulations.

II. *Acardii acorni*, which possess only the head, with possibly traces of the trunk and limbs, constitute a conspicuous group of monsters depending for their production upon the effects of unusual combinations in their venous channels.

The remarkable acornous acardius represented in Fig. 4647, carefully recorded by Barkow, may serve to illustrate the manner in which, as interpreted by Schatz, such rare malformations originate. The production of an acornus implies a close and intimate primary anastomosis between the vitelline circulation of the fused embryonic anlagen. Although the communication between the two vitelline veins at first consists of numerous vessels, during the subsequent development of the embryos the inoculation is reduced to paths forming a single main channel, the *common vitelline vein*. Under usual

conditions, this vessel becomes impervious and atrophies upon the establishment of the allantoic circulation. In response to the imperative necessities of impaired nutrition, however, it may remain and take an important part in the nourishment of the acardius.

In consequence of early obstructed circulation due to stenosis or arrest of development of the allantoic vein of

sponding vessel of the weaker twin. The reversal of its circulation so induced, following the heart-death, affects not only the blood current within the umbilical arteries, but also within the extended path formed by the common vitelline vein, the typical condition of acardia being now established. Since the vitelline vein affords a ready exit for the blood carried by the superior vena cava, the circulation supplying the cephalic end of the fetus is comparatively well maintained, and hence the conspicuous development of the head and immediately succeeding part of the trunk, and the suppression of those portions of the body not in relation with the superior vena cava.

III. *Acardii acephali* form a group in which the lower or pelvic portion of the trunk constitutes the chief representative of the body of the malformed fetus, to which other parts of the trunk, together with all, some, or none of the limbs, but no head, may be added.

In considering the anatomical factors active in the production of such headless forms, the importance of the collateral venous anastomoses, by which the obstruction in the umbilical vein is partially compensated, becomes evident. The most important collateral path, probably, is that afforded by the enlargement of the veins of Burow, small branches within the abdominal wall connecting the umbilical and deep epigastric veins. Since the most active circulation, and hence most efficient nutrition, is determined by the position of the channels affording the readiest return of the blood, those portions of the acardius affected by the blood current established by means of the collateral vessels will be most favorably

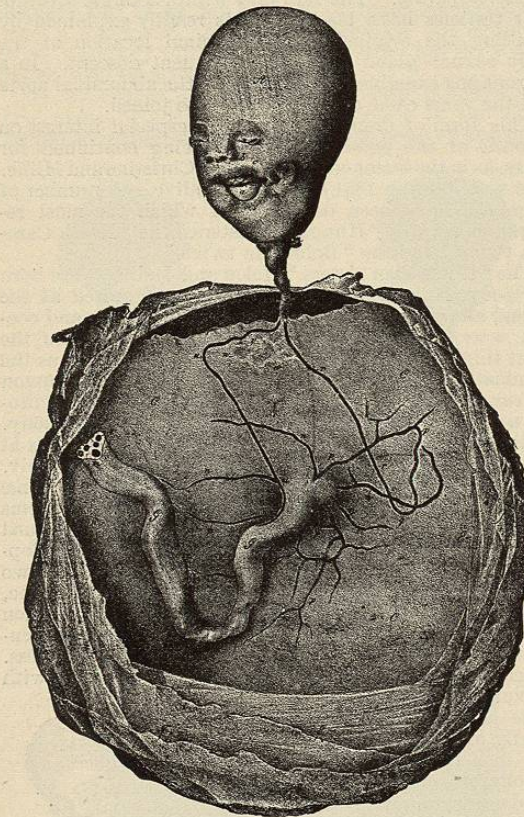


FIG. 4647.—Acardius Acornus. Rudiment of left upper extremity and of digestive tube. (Barkow.)

the subsequent acardiac fetus, the blood pressure of the latter becomes lessened, followed by the passage of the blood from the systemic veins of the stronger twin into the weaker fetus by way of the still present common vitelline vein. Notwithstanding the improved nutrition thus secured, the heart sooner or later becomes enfeebled in consequence of the inadequate blood supply and the greatly increased length of the circuit through which the blood passes. With the development of the umbilical cords, the common umbilical vein is included within both, and hence the blood which is carried to the placenta by the umbilical arteries of the future acardius must pass, in order to return to the latter, through the placental villous anastomoses into the umbilical vein of the normal fetus and thence to the point of entrance of the common vitelline vein; then along the entire length of the umbilical cord back to and across the placenta, into and along the entire length of the cord of the second fetus to the overburdened and under-nourished heart—the blood being propelled by the latter through a course over three times the length of its normal circuit. In consequence of these demands, the insufficiently nourished heart gradually becomes more and more enfeebled, until a period arrives when the reduced blood pressure within the weaker fetus is overcome by the circulation of the more fortunate twin, and the blood of the latter passes, by means of the superficial placental anastomosis, from the umbilical artery of the stronger into the corre-

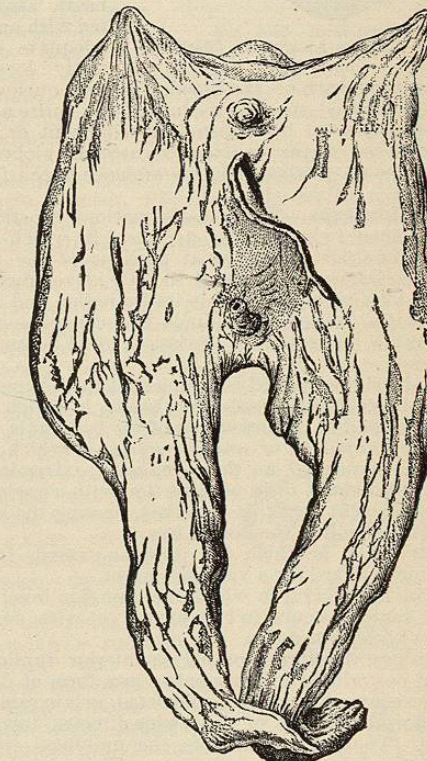


FIG. 4648.—Acardius Acephalus. (Herholdt.)

influenced. These parts will be evidently primarily the lower part of the trunk and the inferior extremities, which are preserved and undergo further development. At times additional veins within the body walls are included within the collateral circulation, the favorable influence of which is thus extended to a larger part of the trunk. The common vitelline vein not being available, either by reason of the late period at which the

acardius was developed, or because the formation of the vein never took place on account of separation of the embryonic areas, the head remains unprovided with an active circulation, and hence fails to develop, the result being the production of the acephalic type of acardius.

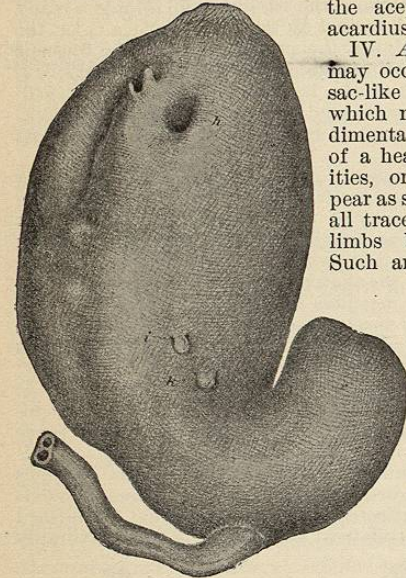


FIG. 4649.—Acardius Amorphus. (Barkow.)

Such amorphous forms probably originate under conditions due to early obstruction of the allantoic veins, followed by insufficient nutrition and subsequent death of the impaired heart, associated with an inadequate centrally located channel for the return of the blood. In consequence of the stasis so induced, marked edema of the entire acardius occurs, followed by arrested development or degeneration of the organs; in extreme cases the globular, skin-covered mass retains little resemblance to the fetal form.

**DOUBLE MONSTERS.**—These malformations constitute a class of joined twins (*gemini conjuncti*) in which the union involves a part of the axial portions of the two bodies. The latter may be equal in their development, or one twin may be so retarded in its formation and nutrition that the united individuals are conspicuously unequal, even to the extent of the less favored becoming a parasite.

Equally developed united twins present three types:

1. *Anterior duplicity*, or *posterior union*, in which the junction is limited to the lower end of the body axis.
2. *Posterior duplicity*, or *anterior union*, in which the embryos are connected at their cephalic extremities; either the head alone, or in conjunction with a variable part of the adjacent trunk, is involved, leaving the posterior or inferior end of the bodies free.
3. *Middle union*, in which the junction extends from the umbilicus upward for a variable extent.

**ANTERIOR DUPLICITY**, in which the junction involves the pelvic region, occurs in two forms: (a) with *dorsal union*, (b) with *ventral union*.

(a) *Pygopagus*, the term applied to anterior duplicity with dorsal posterior union, presents a rare form of double monster characterized by a connected pelvic region, with the dorsal surfaces of the twins directed toward each other. The navel is double, the umbilical cords converging toward a common placenta. The axial skeleton of such monsters includes a single coccyx and sacrum common to both individuals, from which spring two spines; rarely the upper part of the sacrum is also double. The innominate bones and the pubic symphyses of the two pelvis are present in normal number. In consequence of the obliquity of the dorsal surfaces of the two bodies, the lower limbs, usually fully formed, are so disposed that one pair, belonging to both individuals, lies ventrally and more closely placed than the other. The digestive tubes remain distinct until near

the termination of the large gut, where the two recta join and open by a common anus located between the more dorsally situated pair of lower limbs. The spinal cords unite at their inferior extremities to form a common conus with filum terminale. The urogenital tracts show slight fusion of the lower segments; thus associated with a single vulva, the urethra, bladder, vagina, and uterus remain separate. The fact that within a single scrotum four testicles have been found, is readily explained by recalling the original intra-abdominal location of the early sexual gland and its subsequent descent. In a pygopagus examined by Marchand, the abdominal aorta and the venæ cavæ of the two fetuses joined.

This form of double monster is of especial interest on account of the possibility of life being continued for years, as in the instances of the sisters Christine and Millie, who were born in 1851 and are still alive. A number of pygopagi have been described, of which the most remarkable are the "Hungarian sisters," the "North Carolina twins," and the "Bohemian twins."

The "Hungarian sisters," Helen and Judith, were born in Szony, Hungary, in 1701. They were united at the second sacral segment, below which the sacrum and coccyx were single. The common anus lay between the right thigh of one and the left of the other, and was the termination of a single rectum. In front was a common vulvar orifice, into which two vaginæ opened; the clitoris, nymphæ, and urethra were distinct for each body. Inclination to evacuate the bowels affected both twins at the same time, but urination was effected separately. The great blood-vessels, the aortæ and inferior venæ cavæ, were fused in the region in which the arterial stems bifurcated. Judith, in consequence of illness, remained permanently backward in strength and general development in comparison with her sister. While the two suffered from smallpox and measles at the same time, they contracted other diseases separately. Menstruation began at sixteen, but subsequently the two sisters menstruated at different times and with varying profuseness. When in their twenty-third year, Judith was seized with convulsions and became unconscious, shortly after Helen was in a condition of collapse, and expired a few minutes before her sister.

The "North Carolina twins," Christine and Millie, were born in 1851 of negro parents. The union extended from the first sacral segment to the coccyx. There are two recta having a common anal opening, defecation occurring at different times. There are two urethræ and two vaginæ, the labia blending at the posterior margins to give the impression of a single vulva. Menstruation takes place at the same time in both. Common sensibility exists to a limited degree, since one sister can recognize and imperfectly locate touch on the lower extremity of her mate; likewise each knows when the other moves her leg without being able to decide which limb it is.

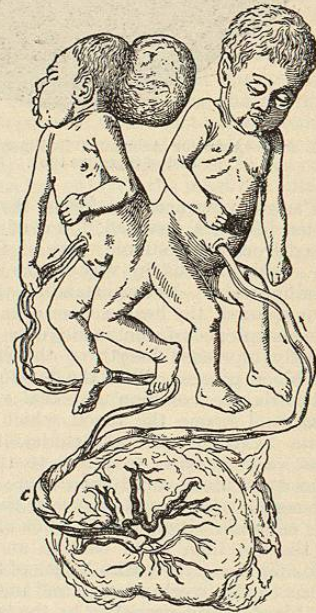


FIG. 4650.—Pygopagus. Sacrum, from second segment, and coccyx single. c, Fused umbilical cords. (Marchand.)

The "Bohemian twins," Rosaline and Josepha Blazek, were born in 1878. The union is latero-posterior and involves sacra and the lower lumbar vertebrae. The two anterior legs are much closer together than the posterior, the former being moved forward with the outer sides of the feet first, to be followed by the posterior pair with the inner sides of the feet directed forward. In this manner the twins are capable of rapid locomotion or even a quick run, although the stronger girl is able to walk naturally forward and draw after her the weaker sister, who then walks backward.

(b) *Ischiopagus*, anterior duplicity with posterior ventral union, is characterized by junction in the pelvic region so related that the right pubic bone of one pelvis

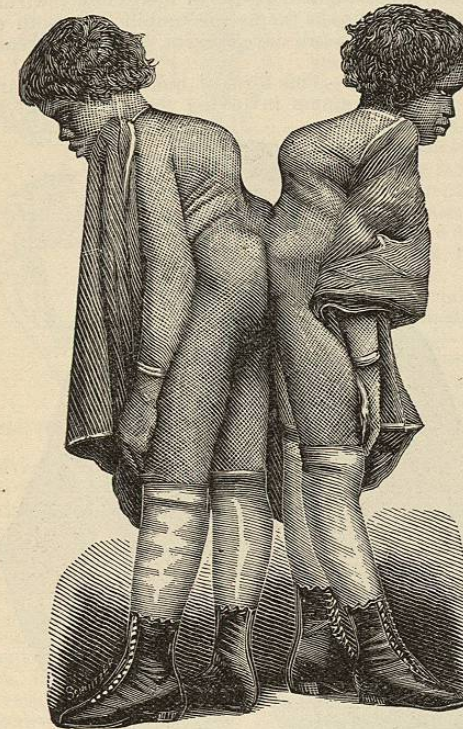


FIG. 4651.—Pygopagus. "Christine and Millie." (Pancoast.)

fuses with the left pubic bone of the other, the ventral surfaces of the two sacra facing each other. The common pelvic ring thus constituted may be subdivided by an osseous bridge formed by the united coccygeal bones. The body axes may correspond in position, thus forming a straight line, or they may be placed so as to include a very obtuse angle. The continuous ventral body surface contains a single navel. The pelvic organs are often fused and the anus may be single or double; likewise the genital organs. When, however, the latter are in duplicate, they are so placed that each is common to the two bodies, the right and left parts belonging not to one individual, but one to each.

The lower limbs may be well developed and, when the body axes correspond, are placed at right angles to the latter. When, however, the axes of the two individuals are placed at an angle, the consequent approximation of the lower limbs on one side may result in more or less complete fusion, or these members may be entirely suppressed, and only two lower extremities be developed. In such cases, one limb belongs to each body. Gemmill<sup>24</sup> has very carefully described the anatomical details of the ischiopagus tripus represented in Fig. 4653. Atresia of the urethra and anus, on one or both sides, is common in these malformations, in consequence of which defects ischiopagi seldom survive for any

length of time. Sometimes conspicuous discrepancy marks the development of the twins, one remaining very rudimentary, the thorax and head being wanting, although the extremities are present. Such forms constitute the *ischiopagus parasiticus* of certain authors.

*Ischio- or ileo-thoracopagus* is the name applied to double monsters in which the ventral or latero-ventral union includes the entire trunk. While the presacral portions of the spines are double, the heads being always separated, the pelvis are so united that the more approximated ilia are rudimentary or are entirely absent, in which case the two sacra are fused. The more ventrally situated iliac bones, together with the corresponding lower limbs, are well formed. The thoraces of the twins are conjoined ventrally by means of a common sternum, while they are continuous on the dorsal aspect. The upper limbs may all be present (*ischio-thoracopagus tetrabrachius*); or those most closely approximated may be fused into a single member, which may be well developed or very rudimentary. In other cases these limbs fail to develop, thus producing the dibrachial type, one of the remaining pair belonging to each body. The lower extremities are subject to the same arrangements, varying from the full complement to a single pair. The external genitals and the anus are single and common to the two bodies, notwithstanding the presence of double uteri; likewise the bladder is single. The digestive tube exists in duplicate until toward its lower end, the two stomachs being arranged with pylori converging. The thoracic viscera remain partially double, although at times the

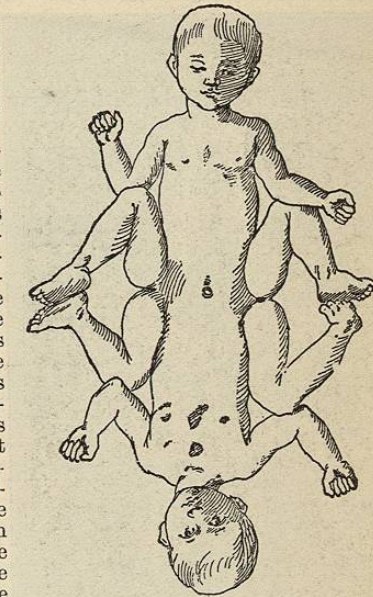


FIG. 4652.—Ischiopagus. (Levy.)

two hearts are enclosed within a common pericardial sac. An interesting case in the Wistar Institute of Anatomy possessed a heart of which the two sides were contributed each by one fetus, an intermediate rudimentary auricle and ventricle being the common possession of both. There were three lungs, the intermediate or middle one possessing five lobes. The kidneys in this case were represented by two large and a small intermediate organ. The ischio-thoracopagi type of duplicity is not incompatible with prolonged life, as conspicuously shown by the Tocci brothers, who have survived since 1877. According to Virchow, who carefully examined these united twins, it is probable the spinal cords

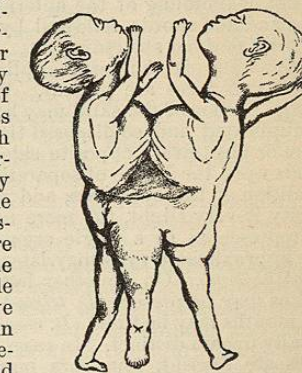


FIG. 4653.—Ischiopagus Tripus. (Gemmill.)

were separate. The functions of the two children were entirely independent, as well as the reflex movements and sensation of the two lower limbs, one of which belonged to each body. Rita-Cristina (1829) and Marie-Rose Drouin (1878) are other noted examples of this form of duplicity; they survived eight and seventeen months respectively.

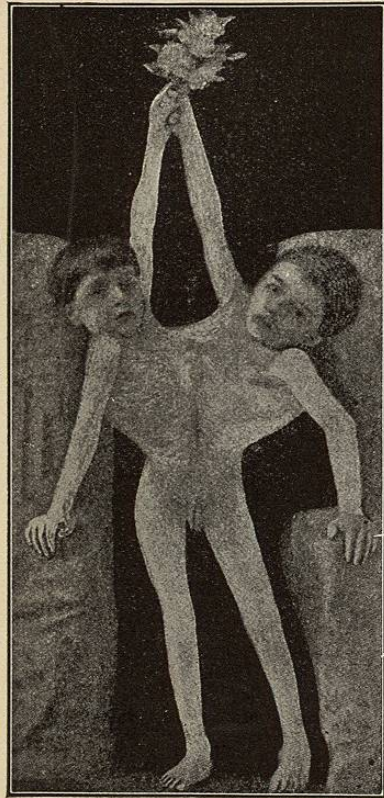


FIG. 4654.—Ischio-thoracopagus. "Tocci Brothers." (Hirst and Piersol.)

as the results of a dichotomous growth involving the cephalic end of a single embryonic anlage, and not, as in the case of the other anterior duplicities, of the posterior union of two distinct embryos.

The malformations in question present varying grades of duplicity from the mere suggestion of doubling to almost complete separation. The most usual condition is that of doubling of the anterior part of the medullary tube and the chorda followed by duplication of the secondarily derived structures.

*Diprosopus* is the term applied to partial duplicity where two faces are present. While almost the entire cranial portion of the skull may be simple, the facial part is duplicated and so disposed that the faces are turned more or less toward opposite sides. The divergence depends upon the degree of separation, as does likewise the condition of the eyes, ears, and mouth. Where the duplicity is very slight, the more nearly related eyes may be represented by a single organ (*D. triophthalmus*), or the approximated eyes may later entirely disappear (*D. diophthalmus*). On the other hand, four distinct visual organs may be present (*D. tetraphthalmus*). More rarely the mouths may be fused (*D. monostomus*), although they usually remain distinct. In cases in which two complete faces are present, four ears may remain distinct; frequently, however, two fuse, or are suppressed. The nervous axis also shows more or less extensive duplication, although parts which normally occur in pairs, as the cerebral peduncles, frequently suffer reduction in number by the formation of a common mesially situated structure in place of two distinct, closely placed members.

*Dicephalus* implies a more extensive and complete du-

plication of the head, and possibly also of the upper part of the vertebral column. When the spine is involved to a considerable extent, the double-headed monster properly belongs rather to the results of extensive posterior union than to partial anterior duplicity, the differential test being the single or doubled condition of the pelvic end of the spine and the pelvic organs. Frequent examples of dicephalic monsters are seen in the doubled-headed calves of the popular museums, in which the duplication involves, in addition to the head, only the cervical region. In man a true dicephalus is comparatively rare, since the apparent examples of this malformation are really cases of extensive posterior union.

POSTERIOR DUPLICITY with anterior union occurs in two forms: (a) *dorsal union*, limited to the head; (b) *ventral union*, involving to a greater or less extent the trunks of the two embryos as far as the umbilicus. Both embryos may undergo full development, or one may remain stunted.

(a) *Craniopagus*.—This form of monstrosity is characterized by dorsal union involving the heads alone, the

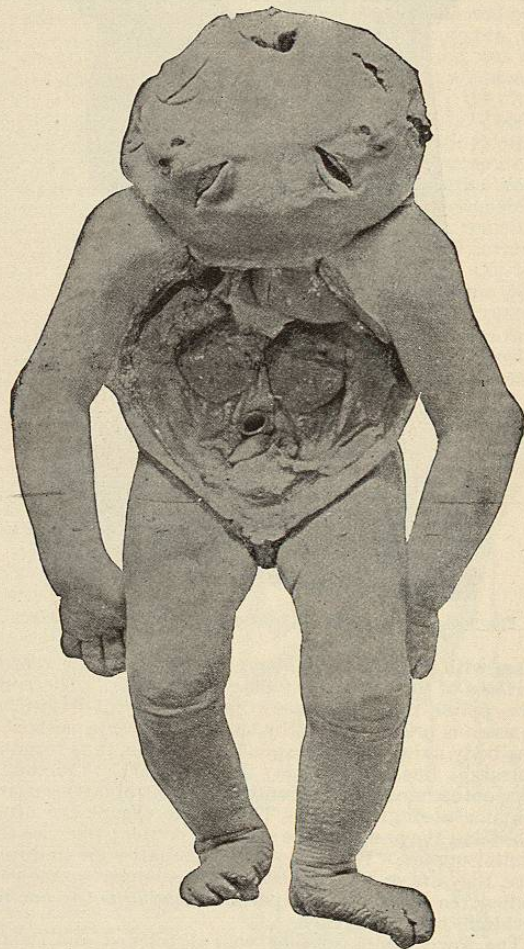


FIG. 4655.—Diprosopus. (Hirst and Piersol.)

axes of the two attached embryos either forming a straight line, or lying at varying angles with each other. The junction may correspond in position with the frontal, parietal, or occipital regions, and usually includes only the integument and cranial vault, the two brains remaining separated by their membranes within a common cranial cavity. It is of interest to note that identical parts of the united skulls are by no means always opposed. This is conspicuous in those rare forms

in which partial rotation of the body axes occurs, in consequence of which the faces are turned toward opposite sides. Craniopagic monstrosities are rare and usually short-lived; an exceptional case of craniopagus occipitales, however, is reported as having survived ten years.



FIG. 4656.—Diprosopus. (Ziegler.)

The development of the united embryos may vary so that marked difference is presented in their general growth. This lack of correspondence may be so considerable that the one fetus becomes an appendage to the more favored, giving rise to a very rare form of head-joined twins, known as *craniopagus parasiticus* (Fig. 4659). One individual is finally represented by a head, alone or in connection with part of the trunk, fused to the autosite, as the better developed fetus may be termed.

(b) *Cephalo-thoracopagus*, or *syncephalus*, applies to posterior duplicities where the union is ventral or latero-ventral and involves the head and the thorax, the pelvic ends of the joined embryos remaining free. The early united twins pass through their development in common, each contributing its quota to the formation of the composite individual. Two faces are formed, each of which belongs half to each embryo. When they are exactly opposite and equally developed, the condition is known as *Janus symmetros*, one face looking ventrally, the other dorsally. More usually there is some lateral misplacement and unequal development as seen in *Janus asymmetros*, in which one face alone is well formed, the other presenting a pair of closely approximated ears. This discrepancy may result in cyclopia, synotia, or obliteration of the oral opening, as well as in suppression or fusion of various parts of the skull. The internal organs of syncephalic monsters present various degrees of union, corresponding in a measure with the extent of the exter-

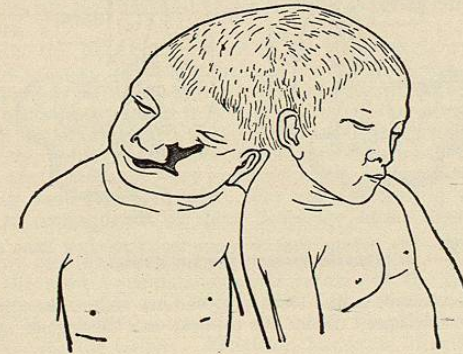


FIG. 4657.—Craniopagus. (Barkow.)

nal fusion. In general, however, the viscera retain their independent arrangement. The cerebral divisions may be fully represented, or the adjacent hemispheres may be

fused, although the cerebelli, pontes, and medullae are separate. The two hearts are often so disposed that one lies in relation to the common ventral, the other to the dorsal body surface.

MIDDLE UNION.—The ventral or latero-ventral attachment of the embryos forming such malformations extends for a variable distance from the umbilicus upward, at times as far as the head; the pelvis, however, always remains uninvolved. The resulting anterior and posterior duplicity, with intermediate union, suggested the term *duplicitas parallela* applied to these monsters. The umbilicus is single, except in rare cases in which the cord is formed by short converging limbs from the two individuals.

THORACOPAGUS embraces two subdivisions of middle union involving the thoracic region: (a) *Xiphopagus* and (b) *Sternopagus*. The former presents an attachment at the xiphoid processes, the latter along the sterna of the two embryos.

*Xiphopagus* is characterized by a more or less extensive epigastric bridge which connects the opposed ventral or ventro-lateral surfaces of the two bodies. The inferior limit of the attachment is the common umbilicus, the upper being marked by the united, usually elongated, xiphoid processes. The thoracic cavities are entirely separate, and divided from the peritoneal sacs by intervening diaphragms. The two livers are usually connected by a bridge of hepatic tissue, although they may remain ununited. In the former case the peritoneal cavities may communicate. The digestive tubes may also open into each other in the vicinity of the stomach or small intestine; in other cases the canals remain entirely distinct.

In a number of xiphopagic twins life has continued for many years. The most noted of these, the Siamese Twins, Eng and Chang, attained the ripe age of nearly sixty-three years before their death in 1874. They were of Siamo-Chinese parentage, being born near Bangkok in 1811. The mother, thirty-five years of age, presented a remarkable predisposition toward multiple births, of the fourteen children which she bore ten being twins. Four daughters preceded the birth of Eng and Chang. The twins were united by a bond extending from the common umbilicus to the xiphoid cartilage, measuring three and one-quarter inches vertically. The connection was of greater length above (one and one-third inches) than along its lower margin where it measured almost three inches. The circumference was nine inches, the bridge being broader above than below. Eng, the stronger and better developed of the two, was upon the right, Chang upon the left. Their bodies lacked symmetry, since the inclined and opposed parts were generally less well developed than those turned away. The opposed thoracic surfaces were lacking in rotundity, the outer exhibiting unusual fulness. Chang's chest was deflected to the left owing to marked lateral spinal curvature. The outer arms and legs were stronger than the opposed members.

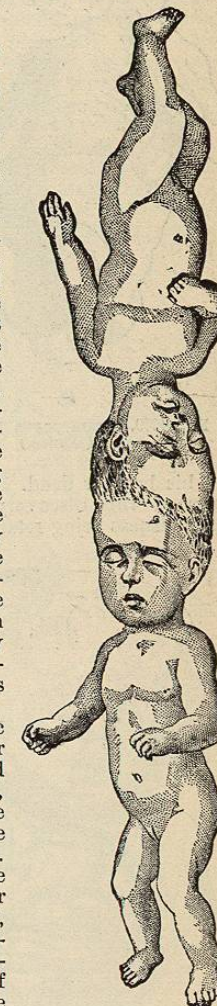


FIG. 4658.—Craniopagus. (Sannie.)