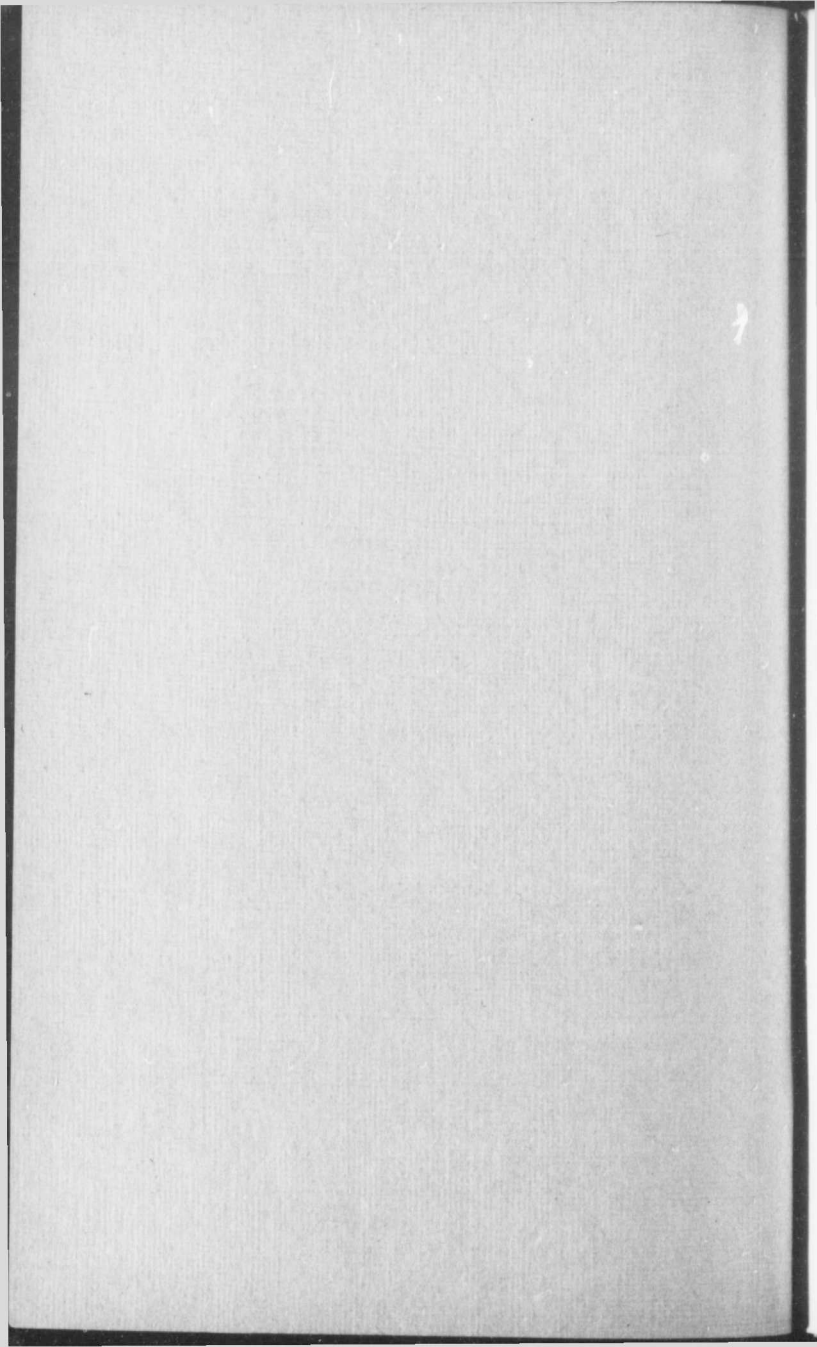


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Cephalothoracopagus Monosymmetros
Cyclops Synotus

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(*From the Medical Museum.*)

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DOUBLE MONSTER OF JANUS TYPE:
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CYCLOPS SYNOTUS.*

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In this interesting form of double monster we have an excellent example of very early dichotomy giving rise to two parallel primitive streaks which are developed side by side and in turn giving rise to a monster the result of fusional duplication. In this case, therefore, it will be seen that a complete cleavage has occurred very early and a secondary fusion has been brought about between contiguous parts of the body, also at an early period. There are many possible varieties of this type of fusion, for example, head to head, waist to waist, in an axial direction, and so on. At times the directions of growth and interference of tissues with one another lead to the suppression of certain parts of the anatomy. The specimen here reported is perhaps the most interesting and the strangest of this group of monstrosities. As one looks at the monster one observes a broad head and face, two arms, and two legs, and if one walks around it an exactly similar appearance presents itself on the other side, but not of an equal development. In this case the fusion has been so unequal that the two facial surfaces are markedly different and it gives one the impression that the generous development of one side has taken place greatly at the expense of the other or diminutive side. Briefly, the resulting double foetus has a single broad head having two faces, one looking either way, and a single thoracico-abdominal cavity having two ventral aspects, corresponding to the two faces, while the backs, arms and lower portions of the bodies from the umbilicus down, remain independent units, becoming fully and symmetrically developed. Two forms of these so-called Janus monsters exists, according as the union has taken place in a straight line, that is at right angles to the vertical axis of the bodies of the respective foetuses, or obliquely as in this case, so that the fused faces on one side have not been able to develop fully. In the former case (symmetrical type) a Janus having two perfectly formed faces and symmetrical organs, will result. In the latter case, that in which the head ends have come together at an acute angle, one face will be perfectly formed, but the other

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will present the deformity above described, resulting from the inclusion and consequent non-development of the features along the median line (Janus monosymmetros). All degrees of facial asymmetry may exist from a simple narrowing of the space between the eyes and narrowing of the nose and mouth through a condition in which a single median eye (Cyclops) surmounted by a nasal proboscis, and ears united in the median line below a rudimentary chin (synotus), as in the example reported by us here, to a condition in which only a trace of the united ears below an apparently normal occiput remain as external evidence of the obliterated second face. As would naturally be supposed in these asymmetrical forms, the fused thoracic and abdominal viscera share in the asymmetry, i. e., they are correspondingly smaller than the viscera of the perfect side. A glance at the accompanying diagram will explain on simple geometrical principles the development of these two types of Janus monsters.

The specimen here reported was presented to the Pathological Museum of McGill University by Dr. Andrew MacPhail. It is a very fine example of the monosymmetrical form of Janus, the face on the asymmetrical side showing the conditions of Cyclops and synotus referred to above. No clinical notes were obtained, except that the child was delivered at full term. The following is a careful description of the external appearance and the results of dissection of the specimen. Two photographs are appended, No. I showing the perfect, and, No. II the imperfect side of the monster. In referring to these surfaces in the description we will speak of them by these numbers I and II, and the foetus to the right and left of the perfect side will be spoken of as foetus A and foetus B respectively.

DESCRIPTION.

The body is that of two full term female foetuses united ventrally from the umbilicus upward so that face is fused with face, chest with chest, and upper part of abdomen with upper part of abdomen. The resulting double monster has a single broad head bearing two (compound) faces, one looking either way, and two occiputs placed laterally at right angles to the faces, a short broad neck surmounting a trunk which has a double ventral aspect corresponding to the two faces above, four arms, and two backs and spinal columns extending down below the occiputs. A single large umbilical cord is inserted into the fused abdomens, centrally, at their most dependent part. Below its insertion the two foetuses are entirely independent of each other, the lower half of the abdo-

PLATE I.
MODE OF DEVELOPMENT OF JANICEPS MONSTER.

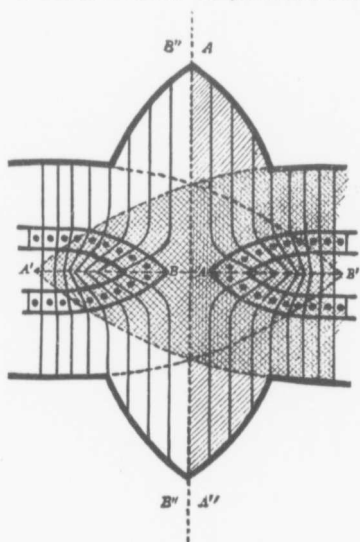


FIG. I. DISYMMETRICAL JANICEPS MONSTER.

The longitudinal axes of the component embryos are in the same line. Fusion has taken place early in front of the growing points A and B respectively; as a result, instead of the cells of the head region derived from A growing to reach A', they are diverted by the opposing pressure of the cells derived from B; each face on either side is thus made up one-half from A (A'') the other from B (B'').

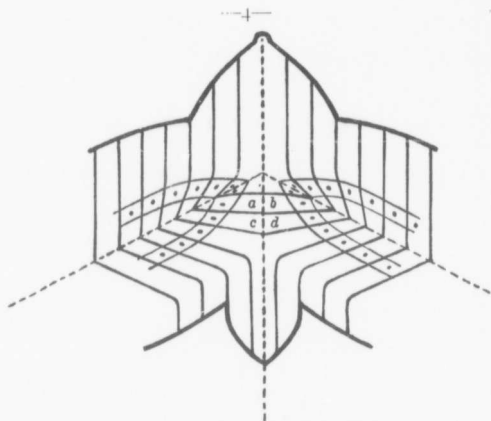
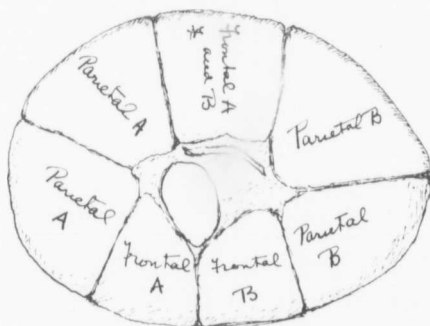


FIG. 2. ASYMMETRICAL JANICEPS MONSTER.

Apicopolar fusion at an angle less (or greater) than 180 degrees, with resultant failure to develop parts of the secondary face on the side of the lesser angle.

From General Pathology. J. G. Adami. Vol. I, p. 231.

II. INCOMPLETE SIDE.



I. COMPLETE SIDE.

FIG. 3.

Diagrammatic sketch of calvarium, showing two frontal bones on complete side (I), and single (fused) frontal on incomplete side (II). A and B refer to the focus to which the various bones belong.



PLATE II. THE SYMMETRICAL SIDE.

Cephalo-thoracopagus monosymmetros cyclops synotus. A double monster of the Janus type in which the heads of the foetuses, and their bodies as far as the umbilicus are fused along their ventral aspect, a single body having four arms and four legs, and a face looking to either side resulting.



PLATE III. THE ASYMMETRICAL SIDE.

Cephalothoracopagus monosymmetros cyclops synotus. On this side of the double foetus fusion has taken place at a somewhat acute angle, so that the two halves of the face (one of which has been derived from either foetus), have been reduced along the median line, a single eye (Cyclops), centrally placed, with the nasal proboscis displaced above it, and the ears approximated to each other beneath the chin. (Synotus, resulting.)

mens, the pelvis with normal external genitalia, and the completely formed lower extremities of each foetus being seen placed laterally to the vertical axis of the composite trunk in a line with the spinal column of each, and at right angles to the double facial and ventral aspects. Thus in this monster the complete side, Figure I, is seen to be made up of the right half of the head, thorax, and upper abdomen with the right arm of foetus A, and of the left half of the head, thorax, and upper abdomen, with the left arm of foetus B, while vice versa the incomplete side, Figure II, is made up of the left arm, left half of the head, face, thorax and upper abdomen of foetus A, and the corresponding right parts of foetus B, the lower part of the abdomen, pelvis and lower extremities of each remaining independent.

The bodies of both foetuses (A and B) are seen to be well developed, the fingers, toes, and nails of the eight extremities being perfect. There is a single thick composite neck (formed by fusion along the anterior surfaces) which has two backs, two sets of cervical spines forming part of the two perfectly developed spinal columns. The four shoulders are perfect but are pushed somewhat backward in each foetus by the approximation of the two thoraces in the ventral fusion, so that a deep groove remains between them on the dorsal aspect of each foetus. There is also a depression between the shoulders on the ventral aspects of the composite trunk which is most marked on the asymmetrical side, and which is apparently due to lack of space between the shoulder joints and the sternums.

The head is well covered with thick black hair. It is of a peculiar broad flattened shape, its widest diameter being laterally from occiput to occiput, where it measures 10.5 cm. From forehead to forehead it is 7.5 cm. The *symmetrical*, or perfect, side of the head has a broad face, which wears a mournful expression owing to the way in which the eyes and mouth are set, and bears traces on close examination of its double origin. The ears are large and well formed; they are wide apart (6.5 cm.), and are placed somewhat lower on the head than is normal, and in an oblique position, so that their upper ends look outward and their lower ends inwards towards each other. The chin is broad, the mouth small and depressed at either angle, the nose rather broad and flat with a well marked central depression at its cartilaginous tip. The eyes are rather small but they are otherwise normal and are provided with eyelashes. They are symmetrically placed on either side of the flattened bridge of the nose, in a somewhat slanting direction from within outward, so that each droops down-

ward at the outer canthus towards the angle of the jaw, giving a curious expression. The forehead is high and rather broad, but the part free from hair tapers upwards to a central point. The eyebrows are well formed, but droop downward and outward as do the eyes.

The *incomplete* side presents a marked and characteristic deformity. It is much narrower than its fellow, and a single broad eye (Cyclops) covered by four rectangular lids, provided with eyelashes, occupies the centre of the face just below where the bridge of the nose should be. There is no sign of the nose as it is normally seen, but it is represented by a small snout-like protuberance over 1 cm. in length, which projects from the median line just above the single eye. This snout is channelled, and, when held up, resembles a small thick-walled tube. Two eyebrows slant obliquely downward on either side of the single eye. The forehead is narrow and lofty, reverse V-shaped in form, and covered, except in its central portion, with thick black hair. There is no mouth, but a curious small fleshy projection, .5 cm. in length, tapering to a flattened, red, and perforated apex, lies below the eye and suggests the superior maxillary process. Below this nodule the face recedes sharply, and in the depth of this receding angle lies a tiny rudimentary tongue. There is no chin, but the ears lie together in the middle line of the neck just beneath this small organ, occupying a horizontal position with their inferior parts touching each other (*synotus*).

On removal of the scalp the calvarium is exposed and presents the following appearances. It is smooth, oval from side to side, and has a centrally placed, large, diamond-shaped fontanelle, the widest diameter of which is from back to back (5 cm.) and its narrower (3.5 cm.) from face to face of the composite foetus. In it are two sesamoid bones, the larger of which measures 2 x 1.5 cm. On the complete side the calvarium is seen to consist of four bones, namely, one frontal and one parietal of each foetus. The two frontals are centrally placed, the right measuring 3 x 2.5 cm., and the left, which is slightly larger, 3 x 3.5 cm. These bones are definite and distinct from each other, and are held together by thick membranous tissue. The parietals on either side are trapezium-shaped, and each measures 5 x 5.5 cm. diagonally. On the incomplete side the calvarium occupies a smaller area than on the complete, being narrower from side to side. Here it is seen to consist of only three bones, a single large broad frontal, 3 cm. broad by 4 cm. long, which shows when held up to the light a central, thickened, bony deposit, representing the line of union

of the two frontals from which it is formed. To the right and left of this fused frontal bone are two irregularly trapezoid bones measuring diagonally 5 x 6 cm. each, which represent the parietals of B and A respectively. The whole is shown in the accompanying sketch (Fig. 3). The cranium is closed in laterally and below on either side by the occipital bones of the two foetuses.

On removing the calvarium the inner aspect of the cranial cavity is seen. It presents a smooth, normally adherent dura, and is irregularly divided from side to side into two main divisions by a thick process of dura mater. The smaller division is on the incomplete side of the foetus. It measures 6.5 cm. at its widest part and about 4.5 cm. in altitude from the division as marked on the fontanelle to the base of the skull. The larger division is on the complete side. Each division contains two cerebral hemispheres, those in the smaller division (incomplete side) being small, devoid of sulci or convolutions, and having a finely granular aspect, while the hemispheres in the larger division (complete side) are much larger, and show well-marked sulci and convolutions which, except for their irregular outline have the appearance of normal brain. The cerebral hemispheres on the incomplete side are less than one quarter the size of those on the complete side, and are imperfectly divided from each other by a single fissure. A similar fissure completely divides the hemispheres on the complete side. There is a well formed cerebellum for each foetus situated posteriorly (laterally in the specimen) above the neck of each. The olfactory nerves are absent on the incomplete side. With this exception the cranial nerves are all present and paired. The optic nerves running to the complete side are well apart, those on the incomplete side are placed well together, are smaller than their fellows, and run forwards to the single eye.

There is definitely a fusion of only the two forebrains, the mid and hind brains being perfect on each side. The peduncles, corpora mamillaria, optic thalami on the incomplete side are somewhat smaller than those on the complete, that is, the half of each cerebellum touching on the incomplete side is less developed than the half touching on the complete side. A peduncle runs from each cerebellum to the incomplete half and one from each to the complete half. There is apparently a fusion of the pituitary bodies on the incomplete side.

DISSECTION OF THE BODY.

Of the two sternums present that on the incomplete side is smaller and softer than the other, presenting irregular centres of ossification. Two perfectly developed hearts are placed ventrally

one on the left side of either surface; that on the incomplete side being smaller in all respects than that on the complete. That each of these hearts was derived from the fusion of the right chambers of one foetus with the left chambers of the other, was proved by their relationship to the lungs. Each heart gives off a large branch to a large common aorta which descends vertically. There are four lungs present. The pair on the complete side are formed by the right lung of foetus A, and the left lung of foetus B, and they derive their blood supply from the larger heart which lies on the left aspect of this surface. The lungs of the incomplete side are formed by the right lung of A, and the left lung of B, and are connected with the smaller heart of their surface. There is a single trachea present and a large common oesophagus which runs down to the left of the incomplete side to reach a large balloon-shaped common stomach. There are two livers formed, and two gall bladders which open by separate ducts into a common duodenum. The diaphragm is perforated by two inferior cavae, one going to either heart. The intestines are common until the place of insertion of the omphalomesenteric duct. Here the small intestine widens and divides into the two parts in which it is continued. All the structures below this point, i. e., the remainder of the small intestine, the caecum, appendix, colon, rectum, as also the internal genitalia, are all of course double.

The following are briefly the most interesting points of the above report:—

I. We have here a typical example of early embryonic cleavage with secondary fusional duplication.

II. The line of fusion has been oblique resulting in an acute angular fusion on one side, and an obtuse angular fusion on the other.

III. The resulting monstrosity is a monosymmetrical Janiceps: where fusion was on the acute angle side we have the asymmetrical, or poorly developed, side of the monster, and where the fusion was on the obtuse angle side we have the symmetrical, or fully developed, side of the monster.

IV. The fusion was along the ventral aspects from head down to omphalomesenteric duct, consisting of fusion of heads, necks, thoraces, and upper half of the abdomen and their contents, while below the omphalomesenteric ducts all parts were normally developed and tends further to support the equality of the originally cleaved halves, and that the resulting monstrosity of the upper part of the body and head was due to a bad angular fusion.

