

Anencephalous foetus / by Alban Doran, for William Skene.

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25

ANENCEPHALOUS FŒTUS.

By ALBAN DORAN, for Dr. WILLIAM SKENE, of Cardiff.

THIS monster was born on November 22nd, 1888. Dr. Skene writes:—"I never saw the woman, Mrs. E—, till I was called to attend her during her confinement. I found her in strong labour. On examination, a large bag of membranes was protruding, and was ruptured during the first pain she had after my arrival, and the child almost immediately followed. The quantity of liquor amnii was enormous. She had been pregnant, including this, five times during the last seven years. Three children were born alive (males), and are still alive and healthy; the other was an abortion. Mrs. E— tells me that she has experienced nothing unusual during this pregnancy, except that about two months ago she had an attack of hæmorrhage, unattended by pain, which lasted for three days, very considerable at first, then slight, but for which she had no medical attendance; rest sufficed. She had not had any fright, blow, or fall. Her circumstances appear comfortable. Until two years ago she suffered, during a period of six years, from epileptic convulsions." The family history included no evidence relevant to this case. The mother's husband was "a very healthy looking man."

The fœtus weighed one pound thirteen ounces. It measured from the frontal eminence, the highest point in the monster, to the coccyx, six and a half inches; from

the same eminence to the right heel, the leg being drawn straight, ten inches. In general form and the position of the limbs it resembled a frog. The head was strongly extended, the occiput and back of the neck not existing; the space between the chin and sternum was abnormally wide. The face looked straight upwards, the features were large. The vault of the skull was entirely deficient. The integuments joined the dura mater along a line representing the lower limits of the frontal plate of the frontal bone and the lower part of the parietals. The basilar portion of the sphenoid projected upwards; the corresponding part of the occipital was ill developed, and bent downwards from its junction with the sphenoid almost at a right angle. Posteriorly the integumental line ran along each side of the widely opened laminae of the cervical and dorsal vertebræ, and joined in the dorsi-lumbar region. The lumbar vertebræ were similarly deficient, but were covered with cicatricial integument. Projecting from the base of the cranium, so as to rest on the upper part of the back, was a mass which consisted of the cranial hemispheres, small and covered with pia mater.* This mass was partially decomposed when I received the specimen, and I was obliged to remove it. The medulla appeared to be represented by a Y-shaped tract of nerve tissue, each arm, about one fifth of an inch thick, uniting below with its fellow to form a short flattened nerve, gradually lost on the spinal dura mater. In fact, the cord had become flattened out and atrophied.

The thoracic viscera were healthy, the heart was very large. The cæcum and ascending colon appeared to be included in the mesentery. A distinct transverse, descending, and sigmoid meso-colon existed. The uterus,

* In this, and in other respects, the monster resembled a specimen figured in Ahlfeld's 'Missbildungen,' pl. xlviii, fig. 13. In reference to that specimen the author states that, in the comparatively few cases of anencephalus where a distinct trace of the brain is present (as in Dr. Skene's monster), the rudimentary encephalon lies on the back, owing to the characteristic posture of the head.

tubes, ovaries, and vulva were well formed; the anus was distinct and open. The large intestine was distended with meconium. The fingers and toes were perfect. There appeared to be distinct talipes calcaneo-valgus of the right foot, an unusual congenital condition.* On close observation, however, the distortion of the foot was found to be due to extreme flexion of the ankle-joint, increased by the action of alcohol.

Anencephalus is one of the best known of all the forms of monstrosity which occur in the human fœtus. The hideous appearance of the features, the absence of neck, and the open cranial cavity, a well-formed trunk and extremities often co-existing, make an anencephalus very conspicuous. A monster of this kind cannot be overlooked. St. Hilaire has described and figured an Egyptian mummy anencephalus. No doubt the birth of such a monster was once looked upon with superstitious awe. The significance of the general aspect of the head is easy for any anatomist to understand. It is the imperfect development of the upper part of the orbits that gives so ugly an appearance to the face and profile.

Specimens of anencephalus abound in our museums. This form of monstrosity must not be confounded with acardiacus. I fully described the latter variety at the January meeting of the Society this year.† It is developed in one-yolk twin pregnancy; the chief reason why it is often mixed up with anencephalus is because the commonest type of acardiacus is called acephalus. Anencephalus, again, is not identical with perocephalus or simple arrest of development of the head—a condition rarely if ever seen in our species.

* The rarity of talipes and deficiency of the fingers and toes in anencephalus, compared with its frequency in acardiacus acephalus, is noteworthy. In the latter no heart exists, and the extremities are ill-nourished by the feeble circulation derived from the brother twin's heart. In anencephalus that organ is generally large and well-formed. It appears to nourish the trunk and limbs in spite of its necessarily imperfect innervation, a mystery which we may leave the physiologist to solve.

† 'Obstet. Soc. Trans.,' vol. xxxi, p. 4.

Some good descriptions of anencephalous monsters are to be found in our Society's 'Transactions.' Dr. Lloyd Roberts's case (vol. x) closely resembled Dr. Skene's which I show to-night. The drawings (ib. figs. 7, 8, pp. 270, 271) might serve for Dr. Skene's specimen, except that ectopia of the abdominal viscera existed in Dr. Roberts's case. Dr. Langston's case, in the same volume, is illustrated by a good woodcut. Mr. Milward's specimen (vol. xiv, p. 140) was an anencephalus; it is described as "acephalus." Dr. Uvedale West (vol. i, p. 107) in describing a case of anencephalus, noted the abundance of liquor amnii, a phenomenon also recorded by Dr. Skene.

Anencephalus has nothing to do with twin gestation. Once it was attributed to adhesion of the amnion to the primordial skull, so that the pressure of the adherent and unyielding amnion interferes with its development. Ahlfeld ('*Missbildungen des Menschen*') has shown that adhesion of the amnion, not a rare condition, produces distinct facial and cranial deformities, as repulsive as those seen in anencephalus, but of a character essentially different.*

Anencephalus or hemicephalia arises most probably from the rupture of a hydrocephalic skull at about the fourth week. Schlegel figures a case where he found such a skull, just about to burst, in a very early embryo (Ahlfeld, loc. cit., pl. xlvi, fig. 1). Rudolphi figures a case (ibid., fig. 2) where the bursting has just occurred, and the remains of the fleshy vault adhere to the base of the skull. The solid cerebral matter rapidly breaks up and disappears, in extreme cases, to be replaced by a mass of cavernous tissue occupying the base of the skull.† A similar

* The adherent amnion prevents the closing of the embryonal clefts in the face. For an extreme case, quoted by Ahlfeld and others, see "A Curious Monster which lived for some time after Birth," by Dr. W. Ross, 'Trans. Obstet. Soc.,' vol. ix, p. 31, and pl. i.

† Thus Ahlfeld's theory is based on fair evidence. Mr. Bland Sutton traces anencephalus to morbid conditions which prevent the development of the encephalon almost from the first. His theory would not account for cases like the present, where a small, ill-formed brain exists.

disintegration of burst structures is seen in the early stages of ectopia vesicæ, which is believed, according to Ahlfeld, to be due to bursting of the allantois. Mr. Shattock, it must here be noted, traces ectopia vesicæ to quite a different origin.

The base of the skull being freed from the pressure of the cranial contents, its own development undergoes modifications easy to understand. Extreme convexity of the middle part (well seen in the present case at the junction of the basi-sphenoid and basi-occipital), great shortening of the upper boundaries of the orbits, and especial arrest of development of the occiput and the cervical vertebræ, are the main features in anencephalus. They account respectively for the shallow base of the skull, the staring eyeballs, and the absence of neck posteriorly. Some well prepared skulls of these monsters are to be seen in the Teratological Series, Museum of the College of Surgeons.

Thus anencephalus is due to changes which originate entirely within the affected fœtus, quite independently of twin gestation. Acephalus, or, more properly, acardiacus acephalus, is due to changes which originate entirely outside the affected fœtus, and invariably in association with twin gestation.

The first of these points is that the twins are not of the same age, which is believed, according to the author, to be due to the fact that the embryo is not a single unit but is composed of two distinct parts, each of which is capable of independent development.

The fact of the skull being formed from the mesencephalon and the brain from the telencephalon and diencephalon is not to be overlooked. The same anomaly of the brain is seen in the present case at the junction of the posterior and anterior horns, and especially at the upper part of the brain. At the base, and especially at the development of the optic and the cerebral vesicles, are the main features in anencephalus. They account respectively for the shallow layer of the skull, the shallow gyrencephaly, and the absence of both posteriorly. The same features of these structures are to be seen in the pathological cases mentioned in the College of Surgeons. Thus anencephalus is due to changes which originate entirely within the affected fetus, quite independently of twin twinning. Anencephalus, or some property, anencephalus, is due to changes which originate entirely outside the affected fetus, and invariably in association with twin twinning.