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Iniencephalus

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INIENCEPHALUS.*

SAINT-HILAIRE, in his classical and authoritative work on Teratology,[1] defines a monstrosity as a serious deviation from the specific type, complex, apparent on external view, and congenital. His classification of all varieties of anomalies and monstrosities has been followed by teratologists generally since his time. His first division is into single and double monsters. The single monsters may be capable of independent existence for longer or shorter time, or dependent entirely and necessarily

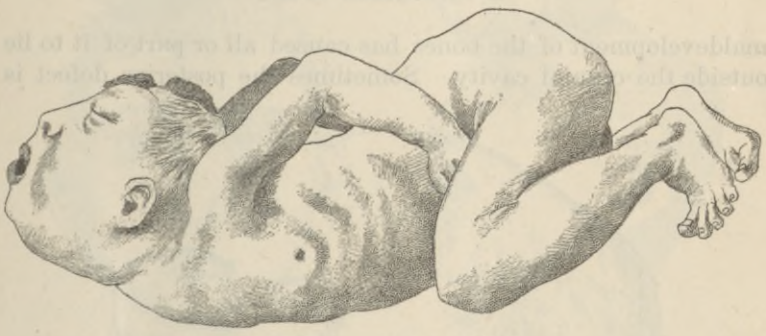

FIG. 1.—Anencephalus. Ahlfeld.

on the maternal tissues. A large family of monsters is characterized by a defect in the closure of the abdomen or thorax, or by defect in the closure of the cranial vertebral laminæ. The latter allows many varieties, according as the defect is in the cranial bones, the spine, both cranial and spinal bones, or parts of each or both. In a large number the brain is entirely wanting or represented only by rudimentary tissue. These are the anencephali (Fig. 1), the most common of all—the so-called frog or owl fetuses, one or more examples of which fall within the

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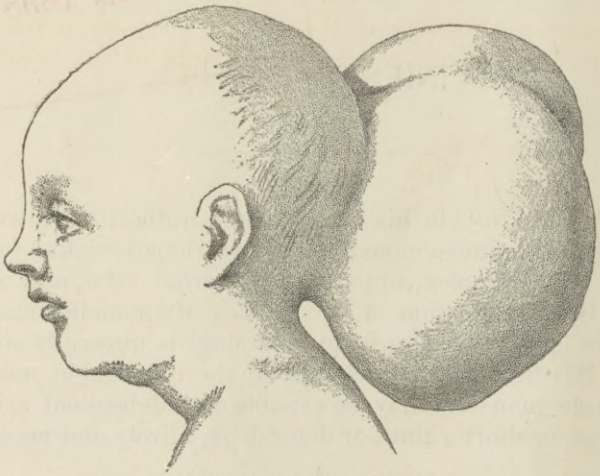
experience of nearly every practitioner. In pseudencephali and derencephali the brain is almost nothing but a mass of vascular tissue. In another class the brain is present, although the



2.—Notencephalus. Ahlfeld.

maldevelopment of the bones has caused all or part of it to lie outside the cranial cavity. Sometimes the posterior defect is

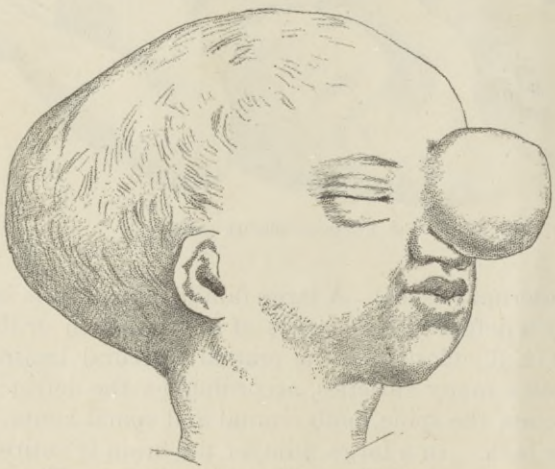


Fig. 3.—Proencephalus. Ahlfeld.

confined entirely or mostly to the vertebral laminæ and we have the spina bifida cases, through the defect in which meningocele usually protrudes. The class in which the brain is

wholly or in part outside the cranium is the exencephalian class. This class is further divided according as there exists or not spinal fissure in addition. Those without spinal fissure are :

1. Notencephalus—where the cranial contents are in large part outside the skull, resting on the back of the neck like a

Fig. 4.—Podencephalus. Ahlfeld.

Fig. 5.—Hyperencephalus. Ahlfeld.

"waterfall," but not adherent. The tumor protrudes through an opening in the occiput (Fig. 2).

2. Proencephalus—the hernial protrusion is anterior (Fig. 3).

3. Podencephalus—the cranial defect is in the vault (Fig. 4).

4. Hyperencephalus—the defect is similar to the last, but of such great extent that the cranial bones are merely rudimentary (Fig. 5).

Those with spinal fissure are :

1. Exencephalus proper—the cranial bones are rudimentary

and the brain lies almost entirely outside on the back. The spinal defect may involve a few or all of the vertebræ (Fig. 6).

2. Iniencephalus—the deformity is similar to that in notencephalus, with the addition of a more or less extensive fissure of the spine from the atlas down. In consequence of the ill development of the cervical vertebræ, there is a lordosis in this region and a compensatory kyphosis in the dorsal and lumbar regions, which extend the head and allow it to sink between the shoulders, thus shortening the length of the back, often to an incredible degree. The cranial bones, indeed, often close the vault above, because the head is so far tilted backward that the margins of the bony defect in the occiput are applied to the back along the margins of the defect in the vertebral laminæ.

Fig. 6.—Exencephalus. Förster.

Thus the portion of the brain that protrudes through the opening in the occiput lies not entirely in the outer world, but mostly upon the spinal cord beneath and almost or entirely covered above by the rudimentary squamous occipital plates and the parietals. This excessive backward tilting of the head exaggerates the shortening of the dorsum. Behind the parietals a meningocele or encephalocele may protrude on to the back.

Saint-Hilaire distinguishes iniencephalus and exencephalus proper thus : The former has the brain situated in great part within the cerebral box, and in part outside it, behind and a little below the cranium, which is open in the occipital portion. The latter has the brain situated mostly outside the cerebral box and behind the cranium, whose superior wall (the frontals and parietals) is more or less rudimentary.

Ballantyne[2] enumerates three cardinal characteristics of iniencephalus : occipital defect, spina bifida, and fetal retroflexion. In different specimens these three characteristics are exhibited in different degrees. As in every classification, there are found some specimens which do not fit well into any category. Especially difficult is it sometimes to distinguish between iniencephali and exencephali proper. The latter may have the retroflexion of iniencephalus, but also will have the most considerable portion of the brain outside in the form of an encephalocele. Some of the cases I shall describe are in this intermediate position and might be classed by different observers with either species. Taruffi[3] groups notencephalus, iniencephalus, and exencephalus proper into one species, which he calls "mero-acrania postero-spinale."

Iniencephalus is a very rare form of monstrosity. Isidore Geoffroy Saint-Hilaire,[1] writing in 1836, says that only three cases had at that time been published. Taruffi[3] states, writing in 1889, that only about twenty-one cases had been reported of mero-acrania postero-spinale. Besides an extensive review of the previous literature, I have collected every case since Saint-Hilaire's time of which I can find reference, including all in the Index Catalogue of the Surgeon-General's Office, U. S. Army, and in the Index Medicus. To these I add one case delivered by myself and two in the Museum of Rush Medical College. These last two I have been permitted to dissect, with the kind assistance of Dr. Louis J. Mitchell, of Chicago, by the courtesy of Dr. Ludvig Hektoen, professor of morbid anatomy in Rush Medical College. Both of these gentlemen have my sincere thanks.

The cases recorded below divide themselves into three general classes. The first includes those without encephalocele, or protrusion of the brain beyond the box formed by the cranial bones and the open vertebræ. This class might be called *iniencephalus clausus*. The second includes those having a small encephalocele, and, with the first, comprises the typical iniencephalus. The third includes those cases where the occipital opening is large and allows the exit of a large encephalocele, but where the other characteristics of the species are so marked that there is no doubt as to the classification. The cases of these two classes might be called *iniencephalus apertus*.

Here follow the cases of the first class, where there is no encephalocele :

Iniencephalus Clausus.—I. Fleck's case[4] (Fig. 7). Ahlfeld[5] gives an abstract and two pictures of this case, which is undoubted iniencephalus of the first variety. The fetus is female. There is tilting backward of the head, causing the face to look forward and foreshortening the back. The head is also turned somewhat toward the left. The anus appears up toward the back. There is no external tumor. There is complete spina bifida as far as the sacrum, fusion of the cervical vertebræ and a wide opening in the occiput, whose margins are attached to

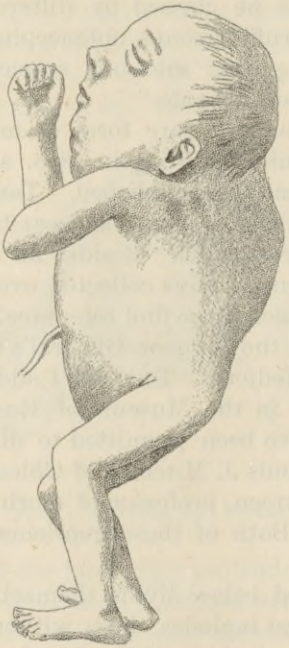


FIG. 7.—Iniencephalus. Ahlfeld.

the transverse processes of the dorsal vertebræ for considerable distance. This case much resembles my own, except that there is less tilting back of the head and shortening of the back. (See p. 12, Fig. 9.)

II. Specimen in the museum of the Royal College of Surgeons of England.[6] This is without clinical history or report of dissection. It is the only one except my own which is male. This is marked No. 278 and was presented by Mr. Jonas Hutchinson. The specimen is "the skull and vertebral column, with a portion of the ribs and pelvis, of a hydrocephalic

male fetus, with distortion of the vertebral column, defective development of the occipital bone, and defective closure of the vertebral canal behind." There is a wide rhachischisis of the cervical and first six dorsal vertebræ, and the head is so tilted backward that the margins of a large defect in the occiput, represented by a dilated foramen magnum, rest on the edges of the laminæ of these vertebræ, to which they are ligamentously attached. There are two scale-like bones representing the supraoccipitals and bounding the large opening. The frontal and parietals are very large. There are only five cervical vertebræ, and their laminæ are fused together. There is another spina bifida involving the lumbar and sacral regions, and there is talipes of both feet.

This is apparently a typical case of iniencephalus, but is unique from the coexistence of hydrocephalus. It will be noticed that most of the cases here reported have an opening behind the cranium, so that there is an exit in the posterior tumor for the excess of brain and meningeal fluid. In this case, having no such outlet for any possible dropsical effusion, it is easy to conceive that the hydraulic pressure from within the cranium might stretch the soft ossifying membranes of the calvarium. This is a point in favor of Dareste's [27] theory of the causation of exencephalians by the vascular changes due to the compression of the defective cephalic hood of the amnion. The stasis produced from this cause in the intracranial vessels would naturally engender dropsical conditions. The same defective development of the overlying amnion which would cause the shortening of the dorsum and the tilting back of the head could also cause the vascular changes in the cranium inducing the dropsy. (See p. 39.)

III. Coffin's case.[8] Child about the eighth month, a span and a half long; head small; eyes prominent; nose flat; mouth open and not easily closed by pressure on lower jaw; flattened chest; scapulæ situated forward of lateral line; head tilted back, with face looking almost upward; posterior fontanelle communicates with an aperture in the skin through which brain matter oozed at birth; spine small and curved anteriorly; no coccyx, anus in position normally occupied by coccyx. External oblique attached to clavicle (showing the shortening of the body); cleft in soft palate; heart large, especially the right cavities; lungs small, but have contained air; kidneys large; brain normal except large olfactory lobes. The spine consists of three bones. Between the first and second is an

aperture leading into the cranial vault, and these two vertebræ are fused together. The posterior part of the occiput is wanting, and the vertebræ articulate with the basilar portion. Ribs articulate with these two bones, but in front with their fellows of the opposite side on account of the lack of a sternum. The third vertebra is the sacrum. The vagina opens where normally the anus should.

The next case, reported in the *Hahnemannian Monthly* in 1880, is somewhat vaguely recorded as to scientific facts, and, were it not for a good picture, it would be impossible to tell what sort of a monster it was.

IV. Yocum's case.[9] This was a face presentation in which the forceps failed and delivery was only effected by vigorous traction after version. The mother died one hour after being delivered. Most of the article is devoted to an account of the labor and the condition of the mother, with speculation about possible maternal impressions. The picture, a copy of a photograph, shows a typical example of iniencephalus as far as external appearances. It appears that the fetus is at term. The head turns backward, reaching almost to the sacrum, and the face looks upward and forward. This tilting of the head gives an appearance of disproportionate size of that part. There is no external tumor, and the scalp is entirely covered with hair. Except in age of gestation, the case is similar to my own, and, except that the abdomen is closed, to No. 9. There is no external sign of a neck, and the furrow between chin and sternum is obliterated, probably by edema. No dissection of the monster is recorded, and the author refers to the picture for the description.

V. Landucci's case[10] (Fig. 8). A primipara, 40 years old, pregnancy normal except for hydramnios, was delivered in the hospital of Arezzo of a dead female fetus nearly at term. The head appears large in proportion to the body; the cranial bones are very movable; the head is tilted backward, so that the face looks forward, and yet the chin rests as if adherent to the sternum, thus causing the ears to appear just above the shoulders and the head to sink between the latter; the face is narrow in proportion to the rest of the cranium. The thorax is short and cylindrical, and runs into a smaller but distended abdomen; the fatness of the skin obscures view of the ribs; the belly falls over the pubes; an umbilical sac four and a half centimetres wide protrudes from this and contains coils of intestine; below this hernia is a piece of normal umbilical cord with a ligature;

the thighs are attached high ; the pelvis is tilted backward, thus, with the tilting backward of the head, very much foreshortening the back ; the anus lies upon the back a few centimetres behind the scalp.

The frontal suture is ossified ; the parietals cover a large part of the back of the fetus, and extend to the spinal column except for the intervention of the narrow pieces of the occiput which

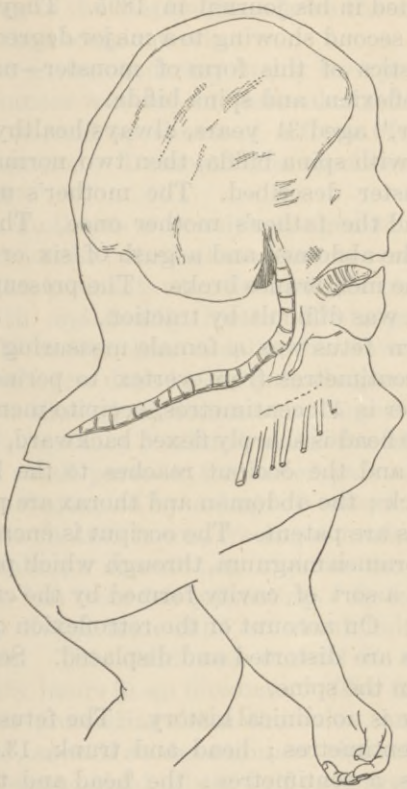


Fig. 8.—Iniencephalus. Landucci's case. Diagrammatic sagittal section.

are attached to the spine at the eleventh and twelfth dorsal vertebræ ; the spinal column is open to the last dorsal vertebra, but covered by the overlying cranium ; a deep sulcus extends into the basilar portion of the occiput and contains the medulla ; the cervical vertebræ turn forward and upward, so that the lower ones are found beneath the lower jaw ; the column turns back in the dorsal region ; the lumbar vertebræ are

better developed and each is complete, but there is a small spina bifida involving the last two lumbar and first sacral vertebræ ; the bodies of the first nineteen vertebræ are fused ; the ribs are irregular in shape, and partly fused owing to the great curvature of the dorsal spine. Except that one kidney is twice as large as its fellow, the rest of the viscera are normal.

The next two cases were shown by Ballantyne, the editor of *Teratologia*, at a meeting of the Edinburgh Obstetrical Society, and reported in his journal in 1895. They are typical iniencephali, the second showing to a major degree the three leading characteristics of this form of monster—namely, occipital defect, fetal reflexion, and spina bifida.

VI. Mother,[11] aged 31 years, always healthy; had her first child affected with spina bifida, then two normal children, and then the monster described. The mother's mother had had twins twice and the father's mother once. There was a great distension of the abdomen and a gush of six or eight quarts of water when the membranes broke. The presentation was foot, and extraction was difficult by traction.

The still-born fetus was a female measuring 30 centimetres long and 19 centimetres from vertex to perineum ; occipito-frontal diameter is 11 centimetres, occipito-mental 13 centimetres ; the whole head is sharply flexed backward, so that the face looks upward and the occiput reaches to the lumbar region. There is no neck ; the abdomen and thorax are prominent ; the vulva and anus are patent. The occiput is encroached upon by a very large foramen magnum, through which part of the brain passes to lie in a sort of cavity formed by the curvature of the spinal column. On account of the retroflexion of the latter the visceral organs are distorted and displaced. Several vertebræ are absent from the spine.

VII.[2] There is no clinical history. The fetus is female ; the length is 28 centimetres ; head and trunk, 13.5 centimetres ; occiput to anus, 5 centimetres ; the head and trunk are fused into practically one mass without semblance of a neck ; the face looks directly upward, and the head is so strongly turned back that the skin over the cranium is directly continuous with that over the coccyx. There are no external signs of rhachischisis.

A medium sagittal frozen section was made. An extraordinary distortion of the vertebral column is evident ; the cervical spine lies nearly in the centre of the mass formed by the head and trunk ; from this point the column passes downward and forward for a short distance, and then turns backward and down-

ward to traverse the body of the fetus and end near the skin just above the anus. The spinal canal is open throughout, but, on account of the extreme retroflexion, a cavity is formed between the occiput and the cervical vertebræ which may be called the occipital spinal cavity. This cavity communicates freely with the interior of the cranium through the large defect in the squamous portion of the occiput. The organs of the thorax and abdomen are pressed downward and forward. A postero-median opening in the diaphragm allows the spleen and intestines to lie in the thoracic cavity.

I next report a case occurring in my own practice in March, 1895. The monster was one of uniovular twins, of which the fellow was a normal specimen of a fetus of five and a half months of gestation. I showed this specimen to the Chicago Medical Society.

VIII. The mother is a primipara, aged 25 years, and married about six months; the last menses occurred October 10th, 1894, and fetal movements were first felt March 1st, 1895. The previous health and family history are negative. She had worked a little harder than usual at her sewing machine and domestic duties a few days before pains began.

Examination showed an abdomen enlarged to about five or six months of pregnancy; a hard mass was felt to the right of the navel and small parts in the lower part of the abdomen; the vagina was occupied by a bag of waters containing feet. This bag broke in two hours, freeing a moderate quantity of fluid and revealing a second bag high in the vagina. Three hands and two feet were felt presenting. Later a head pushed forward and a living male child of about half the size of full term was born. This child was normal in appearance and lived for twenty hours in an incubator. The feet of the second fetus were seen protruding from the vulva, and the fetus was easily removed, as well as the common placenta and the membranes. The second child, about half as large as the first, was dead and somewhat macerated. This was the monstrosity.

The common placenta, with a common chorion, two amnia and two cords, weighed 230 grammes. The first child is 10 inches long; weighs 590 grammes; has finger and toe nails about half-way out to the ends of the digits, and undescended testes. Dissection shows nothing abnormal. The second fetus is 8½ inches long; weighs 190 grammes. Both the children are males, and both amniotic cavities contain the usual amount of fluid.

The monster presents the appearance of great shortening of the back. From the back of the head to the anus is only three-quarters of an inch. The arms and legs seem unduly lengthy and hang parallel. The face looks forward and upward, while the cranium is turned sharply backward, covering most of the dorsum. The neck is edematous and presents no depression between the chin and thorax. The abdomen is protuberant to a marked degree. There is no harelip, cleft palate, club-foot, or external fissure observable (Fig. 9).

Fig. 9.

An incision is made in the median line from the forehead to near the anus, and the skin deflected on either side (Fig. 10). The parietal bones are seen to overlie the upper two-thirds of the spinal column and are closely connected therewith. Cutting these fibrinous attachments on either side and turning the head forward reveals an extensive spina bifida from the upper end to the second lumbar vertebra. The brain is so soft that it is impossible to make out the structures lying directly under the region of the occiput, but the spinal

cord is seen lying at the bottom of the spinal canal and extending into the sacrum as the cauda equina. All the cranial bones appear normal except the occiput. The basilar portion articulates with the atlas, but the bone broadens out so as to leave a large foramen magnum. This opening is continuous with a large defect between the squamous portions, forming with the latter opening the lower circle of a figure 8. The squamous plates of the occiput are represented only by an acute triangle on either side of the opening, articulating anteriorly with the

Fig. 10.

parietals. The margins of the bony defect in the occiput are closely attached to the ends of the widely divergent laminæ of the cervical, dorsal, and first lumbar vertebræ. Thus it will be seen that the spinal canal is covered above by the triangular plates of the occiput and the parietal bones, which form a sort of roof along the dorsum. At the bottom of the spinal canal lies the cord, and over it, contained in the cranium, lies the brain. Thus we have an "exencephalus"; for while the brain does not, as in many cases, escape into the outer world, yet it

does lie partly outside of the cranium and in the spinal canal. The defect is in the occiput or inion, and therefore the variety is "iniencephalus."

The distance across the cleft of the first dorsal vertebra from the extremities of the laminæ is thirteen-sixteenths of an inch; from the upper extremity of the spinal canal to the second lumbar vertebra, where the cleft ends, is seven-eighths of an inch. The cervical spinal column curves downward and forward, making a concavity with the dorsal portion, which turns backward under the cranium. On opening the cranium the cerebral hemispheres are seen to be apparently of normal size. Examination of the abdomen, beyond a considerable protuberance due to the foreshortening of the back, shows nothing abnormal; the organs of this cavity and of the thorax all lie in their proper relations and positions. The umbilical cord is small just as it leaves the belly, but of normal size most of the way to the placenta. Its length is 8¾ inches.

The next case (and also case No. 16) I found in the museum of Rush Medical College, and dissected by permission of Dr. Ludvig Hektoen, and with the assistance of Dr. Louis J. Mitchell. They had not yet been reached in the overhauling which Dr. Hektoen was giving the museum on assuming charge, and were not labelled or dissected. It is not unlikely that search in the museums of medical colleges of the country might find many more specimens of iniencephalus. I believe the twenty-two cases here recorded are all that have ever been reported, at least in literature at all accessible.

IX. Case from Rush Museum (Fig. 11). This is an unmarked specimen without history of any kind. It is a female, apparently of full term. Beclard's centre is present in the femur; the finger nails reach beyond the ends, and the toe nails nearly to the ends, of the digits; the head is full-term size. The following measurements were made: length of body, 12½ inches; length of back (cranium to anus), 2⅝ inches; length of leg, 6¾ inches; length of arm, 7⅛ inches; chin to perineum (anteriorly), 7⅝ inches; bitemporal diameter, 3⅝ inches; mento-bregmatic diameter, 4½ inches; occipito-frontal diameter, 4⅝ inches. It will be seen that there is an immense foreshortening of the back due to the tilting backward of the head. The face looks forward and upward, leaving very little furrow beneath the chin. There is an opening in the abdomen at the navel, admitting three finger tips. The umbilical cord is to the left of this opening, which is fringed by folds of perito-

neum, through which protrude the naked intestines from the jejunum to the sigmoid, and also a small piece of the liver. There is a mesentery common to the small and large intestines. The sternum is cartilaginous. The anterior fontanelle is large and shaped like an acorn set upon a triangle ; length, $2\frac{3}{4}$ inches; breadth at widest part, $2\frac{1}{2}$ inches.

A posterior median incision reveals a complete spina bifida to the tip of the coccyx. The cranium extends as far as the

Fig. 11.

lumbar region, attached ligamentously to the transverse processes and forming a roof over the spinal canal. There is no encephalocele. Cutting the attachments and turning the head forward reveals a wide rhachischisis of the whole cervical and dorsal regions. In the bottom of the depression lies the cord, and on top of the last the brain (macerated and decomposed beyond dissection). There is considerable lordosis of the cervical region of the spine and right scoliosis of the whole column, so that the ribs are irregular in course and deform the

chest cavity. The opening in the occiput consists of a large foramen magnum measuring: length, 1⅝ inches; breadth, 1⅛ inches. The width of the spinal cleft is 1½ inches at the maximum. In the lumbar region is a loose piece of cartilage of the size of two split peas, ligamentously attached to the spine on the right side. The anus is pervious; all the abdominal organs not named above are negative; there is slight talipes varus of both feet.

Except for the gestation age and the celosomia, this case resembles No. 4. The child may perhaps have lived a short time; and, indeed, its deformities are not incompatible with indefinite continuance of life, provided the anterior abdominal opening could have been closed. A successful operation on a much greater celosomia is reported.[53]

Iniencephalus Apertus.—Next follows the second class, having a small encephalocele. The first case is copied in Saint-Hilaire's monumental work on anomalies and monstrosities.[1]

X. Duges' case.[12] The head seemed to be confounded with the thorax, and turned back so that the occiput seemed lost between the shoulders. The anterior part of the neck is flush with the chin and sternum, allowing no cervical furrow. On the back, which seems very short, one sees a fungous tumor of the size of a little walnut, and behind it the thin red membrane by which the tumor is covered extends to the sacral region. Examination of the skeleton shows that the cranium is depressed and prolonged in front of the occipital orifice. The face is oblique and projecting. Several bones are welded together; others, as the parietal, show intervals of cartilaginous formation. The upper occipitals are flattened and run parallel to the base of the cranium. The spine, open posteriorly as far as the sacrum, is remarkable for a torsion so great that its cervical portion is situated under the basilar process and its dorsal portion lies horizontally under the base of the cranium. The cerebral hemispheres remain contained in the cranium; medulla passes by the occipital opening; the rest of the brain is lost in the fungous mass behind the head whence come all the cranial nerves. The spinal cord, adherent above to this mass, but not continuous with it, is complete.

There is also a large diaphragmatic opening, allowing the stomach and left lobe of the liver to occupy the thorax on the left side.

Very like this case of Duges' is the following, depicted in Ahlfeld's "Atlas" (Taf. xxxii., Fig. 14), and undoubtedly a typical iniencephalus with small encephalocele:

XI. Potthoff's case.[11] As will be seen by reference to the figure (Fig. 12), there is considerable tilting backward of the head, so as to greatly shorten the back, and, by the depression of the head between the shoulders, an obliteration of the submental furrow. The head and body are almost one mass, from which hang down together the arms and legs. The back is so short that the anus lies high up toward the back of the head as well as a considerable distance from the vulva. A small, spongy tumor lies at the angle formed by the parietals with

FIG. 12.—Iniencephalus celosomus. Ahlfeld.

the dorsum. There is also part of the abdominal contents outside the body, escaping from a cleft in the anterior belly wall. There is complete spina bifida.

XII. Cruveilhier's case.[13, 14] The mother was a multipara, aged 35 years, of ordinary health. The labor was uneventful and the position occiput right posterior. The child is a female, showing the strong tilting back of the head, two tumors behind the cranium, absence of neck furrow, chin continuous with the

chest, nose flattened, and mouth and ear curiously deformed. The mouth looks as if a string or bit were pulled tightly across it. The cheeks are continuous with the palate (Figs. 13 and 14). The first tumor is situated immediately behind the sagittal suture, and is separated by a fibrinous partition from the second, which lies to the right and a little below the first. Both communicate with the cranial cavity and are filled with membranes and serous and bloody fluid. The cranium contains brain, which only partially fills the cavity, floating in a serous fluid.

There is no trace of the squamous portion of the occiput, but

Fig. 13.—Iniencephalus. Ammon.

through a large opening between the parietals the tumors protrude. The parietals extend over the cervical and the first four dorsal vertebræ, which are cleft anteriorly as well as posteriorly, having double bodies. There is no basilar occipital and but two rudiments of lateral occipitals, one on each side behind the temporals, serving as extensions of the cervical vertebræ. The sphenoid is the last bone of the cranium. The inferior borders of the parietals are attached to the transverse processes of the cervical and first four dorsal vertebræ.

The left lobe of the liver and part of the intestines lie in the left thorax through the diaphragm, which is wanting on that

side. The left lung is compressed; the right lung lies partly in a hernia in the neck; the stomach and duodenum lie partly in the posterior mediastinum; there is shortening and invagination of the esophagus.

The next case is reported in 1847. In many particulars it resembles the case of M. Duges, particularly in the extreme retroflexion and the encephalocele.

XIII. S. W. Drew's and J. B. S. Jackson's case [15] (Fig. 15). The mother was a primipara; the labor occurred at eight and

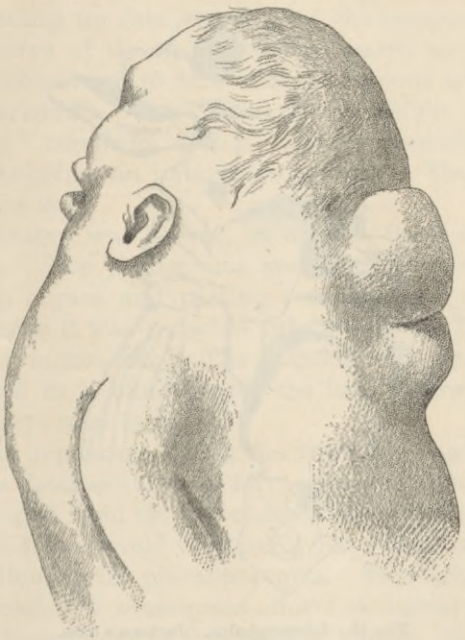


FIG. 14.—Iniencephalus. Ammon.

a half months; the vertex presented. The child, a female, lived half an hour; weight, five pounds; circumference of thorax, fourteen inches; club-feet; there was no neck furrow and the arms extended very low; cleft uvula; lungs irregularly fissured. Brain appeared at the upper part of the spine, looking red and vascular, and of the size of an English walnut. A large mass of brain was found within the cranium, showing distinct convolutions and weighing three and a half ounces. Spina bifida was complete, and two cords existed, one on each side of the spinal canal, communicating above with the medulla.

Examination of the bony preparation preserved in the museum shows the occipital bones united with the spinous processes as far as the first lumbar, except on the left side where there is considerable deficiency. The antero-posterior curve of the spine is excessive; the upper dorsals and middle lumbars form prominent points, and so the pelvis and extremities of the ribs are brought into close contact. The cranial cavity is quite capacious, leaving considerable space between the frontals and parietals and the base of the skull. The basilar and lateral

Fig. 15.—Iniencephalus. Jackson's case.

processes of the occipital bone are well developed. The posterior portion consists of a piece on each side sending a narrow process around behind the parietal to meet its fellow. Besides the marked antero-posterior curve, the spine is somewhat curved laterally. The thorax is very prominent; the anterior half has no connection with the ribs; seven ribs on the right side are partially fused; the twelve left ribs are mostly separate.

XIV. Lawther's case.[16] Primipara 40 years old; labor began in the beginning of the eighth month. In examining in the O. L. A. position the finger fell into a cavity in the cranium oppo-

site the right ilio-pubic junction, the lower margin of which was rounded and the upper indefinable. On account of hemorrhage from a marginal placenta, he pulled down the feet and extracted a macerated female child. A longitudinal section was made into the vertebral column and the halves drawn aside. At the middle of a transverse line drawn between the apices of the shoulders the spinal canal curved forward and downward. In the centre of the column, in an elliptical cavity thirty lines in length, the cord was embedded. The cervical vertebræ were absent; instead is a deep, non-integumentary cavity extending up into the floor of the cranium. Into this cavity the curve of the spinal column partly projected. The occiput was absent except for a narrow, osseous arch in contact with the parietals. There was no cerebellum and only an "amorphous" cerebrum found—"a mere covering of brain substance enclosing an indefinable cavity." The only visible nerve was the optic.

The diaphragm was absent, as well as the mediastina and pericardium. The pleuræ and peritoneum were continuous, investing the organs and passing into the funis as far as the placenta, where it was reflected into a pouch. The cord was less than four inches long. The stomach was absent; the esophagus ended in a blind pouch; the lungs were rudimentary and the kidneys very large.

The finger, in passing during the vaginal examination into a cavity in the fetal skull, must have ruptured an encephalocele which protruded from the defect described in the occiput. The contents of a tumor could very easily have been missed among the blood following the placenta previa. Through this opening artificially made the subsequent efforts at delivery might very easily have forced most of the cerebral matter, leaving the sac enclosing the "indefinable cavity" described. The retroflexion of the spine and the description given of the cranial bones are sufficient evidence for us to class this case with the inienencephali.

XV. Vernier's case.[11] The mother, a primipara 21 years old and six months pregnant, had suffered from extreme distension of the abdomen so great that she could not sit and could hardly breathe. She was seized with strong pains, the cervix dilated, and the fetus followed quickly after an enormous gush of amniotic fluid.

The child had spina bifida, tilting backward of the head, and a hernial pouch behind the cranium containing brain tissue.

The two parts of the occiput did not meet in the middle, but were adherent to the corresponding parts of the upper cervical spine.

There was also cyclops and club-feet. The reporter classes this monster with the acephali, genus rencephalus, but in my opinion it belongs with the class we are considering.

It is not stated how much brain was found inside the cranial cavity, but the turning back of the head, the spina bifida, and

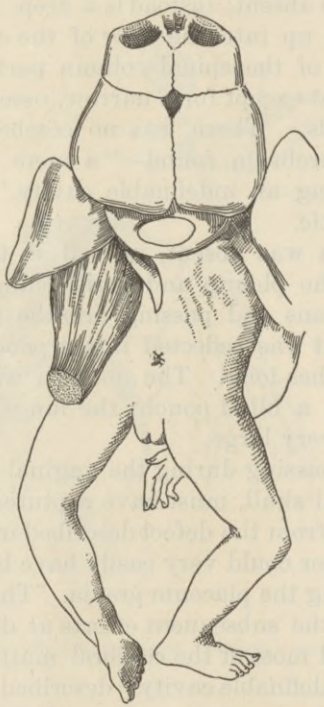


Fig. 16.—Iniencephalus. Remfrey's case.

the attachment of the occipital plates to the margins of the opening in the cervical spine point to iniencephalus.

The next case is an example of the extreme backward flexion which may occur in the bodies of iniencephali. In this the head and pelvis came so near together that the gluteus maximus took an origin from the occiput.

XVI. Remfrey's case.[18] Full term; female, born dead; pelvic presentation and delivery by traction on feet, during

which cord (two inches long) broke. The face looks upward because of the strong retroflexion of the head, which covers the back so completely that the hairy scalp is flush with the buttocks. On the right side and overhanging the crest of the ilium is a collapsed sac, uncovered by hair, which oozes brain matter. The flexion is chiefly at the occipito-atloid joint; the occiput is very imperfectly developed and lies back so flat on the spine that the head nearly reaches the crests of the ilia, and

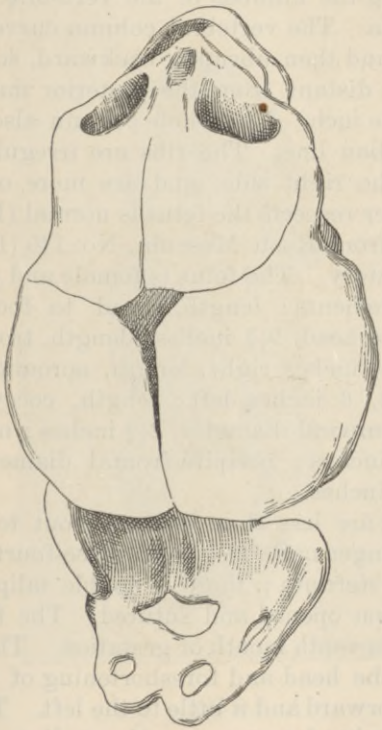


FIG. 17.—Iniencephalus. Remfrey's case.

the gluteus maximus on the left side takes an extra origin from the upper part of the occiput on the same side. There is also some lateral flexion of the body toward that side.

The mento-frontal diameter is $2\frac{3}{4}$ inches, bimastoid diameter $2\frac{3}{4}$ inches; forehead to buttocks $4\frac{3}{4}$ inches, bitrochanteric distance $2\frac{1}{8}$ inches.

The liver is very large and irregular in shape; at its lower edge projects a pedunculated portion, the size of a cherry, into

the amniotic cavity, showing as a prominence on the abdominal wall. The limbs and other viscera are normal.

The tabular portion of the occiput is represented by a crescentic plate on each side attached to the posterior margin of the parietal and separated from its fellow by a wide opening, partly closed below by the displaced spinal column, which thus forms the floor of the third fossa of the skull. The other cranial bones are normal. Irregular centres of ossification prevent counting the number of the vertebræ. There is complete spina bifida. The vertebral column curves forward for a short distance and then abruptly backward, so that the terminal vertebra is distant from the posterior margin of the left parietal only one inch. The whole column also inclines to the left of the median line. The ribs are irregularly developed, especially on the right side, and are more or less fused together. In other respects the fetus is normal (Figs. 16 and 17).

XVII. Case from Rush Museum, No. 140 (Fig. 18). There is no clinical history. The fetus is female and presents the following measurements: length, head to foot, 10⅝ inches; length, body and head, 5$\frac{13}{16}$ inches; length, trochanter to sole, 5⅝ inches left, 5½ inches right; length, acromion to finger tip, 5¾ inches right, 6 inches left; length, coccyx to cranium, 2$\frac{9}{16}$ inches; bitemporal diameter, 2$\frac{3}{16}$ inches; mento-bregmatic diameter, 3$\frac{9}{16}$ inches; occipito-frontal diameter, 3¼ inches; bisacromial, 2⅞ inches.

The toe nails are less than half-way out to the tips of the digits and the finger nails less than three-fourths; the chin is flush with the sternum; there is double talipes varus. The abdomen has been opened and sutured. The fetus appears to be of about the seventh month of gestation. There is the usual tilting back of the head and foreshortening of the back, while the face looks forward and a little to the left. There is no neck furrow, and the head appears as if set directly between the shoulders. In the median line of the back there is a soft tumor covered by a membrane and allowing one to feel the cleft spine underneath. This tumor measures 1¼ by 1$\frac{5}{16}$ inches and occupies most of the dorsal region. Protruding from a bony defect in the skull to the right of the median line and hanging over the right shoulder is another tumor of soft consistence, covered with skin, which bears hair on its anterior third. This tumor, which dissection showed to be an encephalocele, measures 2⅜ by 2¼ inches.

A median dorsal incision is made from forehead to anus and the skin and tissues are turned back. Under the skin the lateral tumor is enclosed in a double sac, the outer part adherent to the skin (dura) and the inner free, allowing, through a small incision, brain matter in a disorganized condition to escape. The cavity of the tumor is continuous with that of the cranium through the bony defect. In dissecting the dorsal tumor nu-

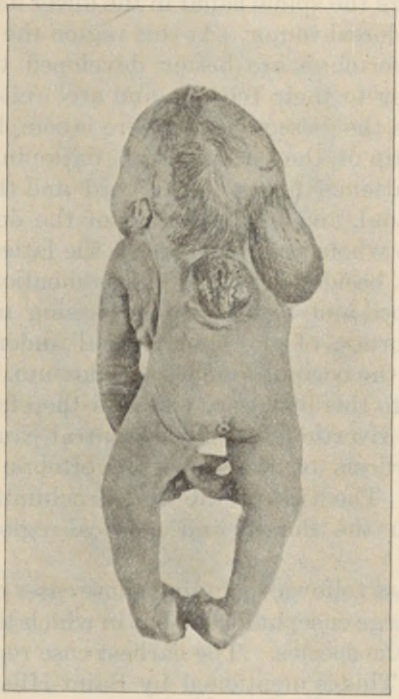


Fig. 18.

merous fibres of the spinal cord and nerves adherent to the under surface are cut.

The parietal bones are of equal size, and an appearance of inequality is due to the fact that the head is twisted toward the right side of the back. The opening in the cranium through which the lateral tumor escapes is between the rudimentary squamous plates of the occiput, which, in the form of acute triangles, are articulated with the parietals at the lambdoid suture and with the basilar part of the occiput at their base below on

each side. The defect in the occiput between these two triangles is a round hole continuous below with the enlarged foramen magnum, thus making a figure 8. Through the upper half of the 8 protrudes the lateral tumor. The margins of the foramen magnum lie flat upon the spinal column and are ligamentously attached to the rudimentary laminæ of the widely open vertebræ. Thus the occiput covers the spinal canal, causing the foreshortening of the back of the fetus. The base of the left lateral occipital plate covers the spinal canal in the lower dorsal region just anterior to the dorsal tumor. At this region the transverse processes of the vertebræ are better developed than elsewhere, approach nearer to their fellows, and are united by a strong ligament (cut in the dissection). There is complete spina bifida as far as the tip of the coccyx, with flattening of the spinal canal. The flattened fibres of the cord and the nerves from it lie in the canal, and in the region of the dorsal tumor are spread over the whole inner surface of the latter. This dorsal tumor contains, besides the fibres above mentioned, a sac filled with clotted blood and with a pedicle passing upward through a constricted portion of the spinal canal under where the left lateral plate of the occiput touches the dorsum. A probe from above passes into this little sac, which is therefore a myelocele, representing a diverticulum of the central canal of the cord. There is a lordosis of the cervical vertebræ and kyphosis of the lumbar. The axis of the spinal column is also turned to the right in the dorsal and cervical regions (right scoliosis).

The third class follows, including those cases of iniencephalus which have a large encephalocele, but in which the other characteristics mark the species. The earliest case reported is that of Hull in 1802. This is mentioned by Saint-Hilaire as a case of iniencephalus. The original I have been unable to see, but I glean the following from abstracts made by J. Z. Lawrence [19] and Cesare Taruffi.[3]

XVIII. Hull's case.[20] The fetus was still-born; the cerebrum, which was defective, almost wholly protruded from the cranium and lay on the back of the neck; there was spina bifida which opened the canal from the neck to the sacrum; the squamous portion of the occipitals was wanting; the frontals and parietals were small and depressed. It will be seen that the case approaches closely the type of exencephalus proper. There also existed club-foot and a union of three ribs.

The following case closely resembles the exencephalus in that a large portion of the brain lies in the posterior encephalocele:

XIX. Gros' case.[21] The mother was a primipara of 40 years of age; the pregnancy was complicated with diarrhea, vomiting, and other digestive disturbances; the labor was not abnormal; there was slight hydramnios; the puerperium was complicated by intense diarrhea, albuminuria, and septicemia.

FIG. 19.—Iniencephalus. Budin's case.

The fetus showed complete absence of the occiput, a notable diminution in the size of the parietals, and an enormous cerebral development behind the cranium. Spina bifida in the cervical region. A band horizontally in the mouth cavity divides it into two portions, in the middle of which is the uvula.

XX. Budin's case[22] (Fig. 19). In January, 1872, entered the Maternité in Paris a primipara 24 years old, pregnant at

term. She was delivered that day of a dead female fetus weighing 750 grammes. There was a large tumor at the posterior part of the cranium, arising below the superior angle of the occiput and descending almost to the sacrum. Hair appeared on the anterior segment of this tumor.

Dissection showed considerable serous fluid between the skin and the dura and in the subdural and arachnoid spaces. The tumor also contained cerebral hemispheres. The cerebellum lay in the right occipital fossa within the cranium, and beneath

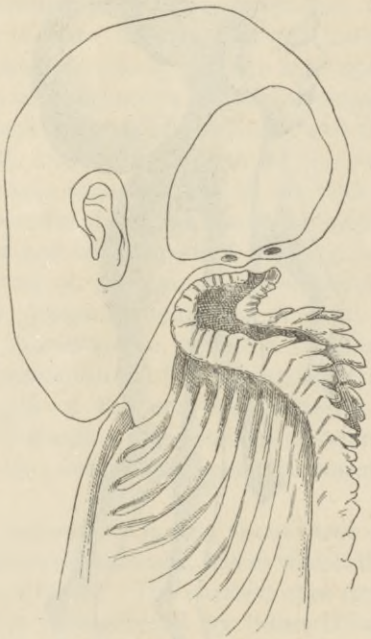


Fig. 20.—Iniencephalus. Budin.

it the medulla and part of the spinal cord. The bony opening in the cranium was bounded by the occiput, the cervical and the upper six dorsal vertebræ (Fig. 20). An anterior curve of the spine in the cervical region formed a deep concavity. The occiput is merely rudimentary as to the squamous portion, which consisted of plates at the sides and above the cranial opening, but the basilar process and the rest of the cranial bones were normal. The thorax is pigeon-shaped because of the sharp anterior curve of the spine. The head is tilted

considerably backward and the dorsum is correspondingly foreshortened.

XXI. Rogers' case[23] (Fig. 21). The mother was 22 years old and a quadripara. At term the abdomen was immensely distended, and a very copious flow of waters occurred after rupturing the membranes. A soft tumor was felt preceding the head, and the delivery of the body was delayed by a hard tumor in the anterior part of the fetal pelvic region. The child gasped and died immediately. It weighed eight and a half pounds.

At the basis cranii is a soft tumor as large as the head ; the back of the child is soft and no vertebræ are to be felt. The first cervical vertebra articulates with the sphenoid or the detached basilar portion of the occiput; thence the spine descends

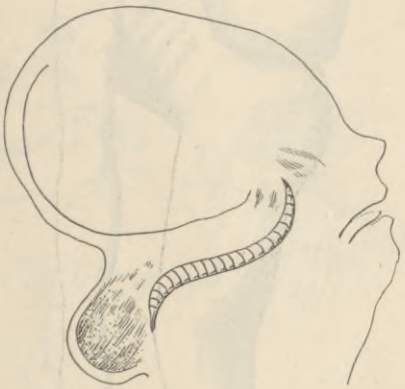


FIG. 21.—Iniencephalus. Diagrammatic sagittal section. Rogers' case.

to a level with the manubrium of the sternum, whence it turns backward and upward and terminates with a short downward curve between the spines of the scapula ; there is no bony column below this level until the pelvis is reached. The occipital bone is entirely free in its inferior portion and does not articulate with any other bone, but, curving upward into the cranial cavity, it leaves an opening into the brain box, whence came the fluid filling the external sac above mentioned ; there are no articular condyles on the occiput.

A hard, tense growth occupies the lower abdomen, extending down to and continuous with the labia majora. To the left of this is attached the umbilical cord. The abdominal tumor is filled with a brown, gelatinous fluid and contains folds of intestine. The anus is closed.

XXII. Bonnaire's case[24] (Fig. 22). A secundipara, without pathological personal or family history, entered the hospital about two weeks before term, showing great distension of the abdomen, but otherwise normal. Biovular twins were born having two placentas, the first fetus presenting by the vertex and the second by the breech. The first was normal in appearance, but the second had an amnion very dropsical. It made

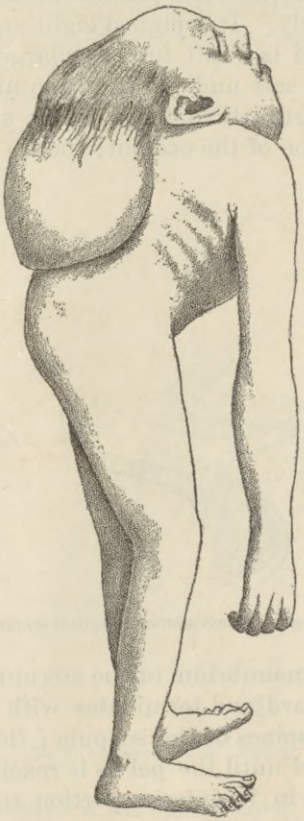


FIG. 22.—Iniencephalus. Bonnaire's case.

a few efforts at respiration and died. The head, which was mostly face, is turned upward and set deep between the shoulders, so that the lobules of the ears touch the ends of the acromia. The chin and sternum are continuous, so that there is no neck. The spinal axis is so curved that the characteristic appearance of a much shortened body and disproportionately long limbs is presented. Behind the head a soft tumor, the

encephalocele, extends as a hernial pouch from the cranium and covers the back as far as the lumbar region. The hairy scalp covers the anterior third of the tumor ; the rest is covered with membrane. Below the tumor is seen a complete spina bifida forming a gutter by the lamellæ and containing the flattened cord.

The cranial cavity and the parietal and frontal bones over it are rudimentary. Most of the brain is in the external tumor. There is a large opening in the occiput (Fig. 23) ; the basilar portion is in two lateral pieces. The most interesting feature is a complete splitting of all the cervical and upper ten dorsal vertebræ, making a Y with the rest of the spinal column (Fig. 24). In the cervical region the branches of the Y curve sharply

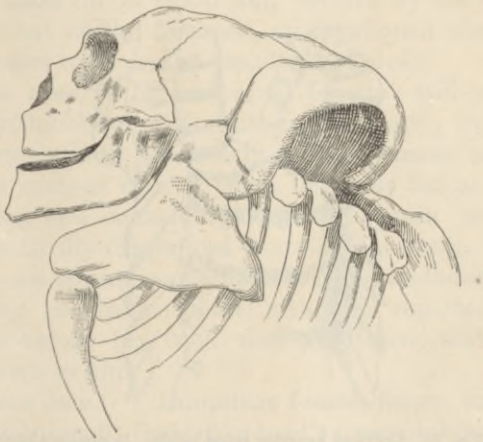


Fig. 23.—Skull. Bonnaire's case.

backward, so as almost to touch the atrophied dorsal. Below the spinal cord a hernial pouch protrudes through the anterior fissure of the upper part of the spinal column, and this pouch contains part of the esophagus.

Dareste says that anterior fissure of the spine is not very rare. I have found only two cases where it existed in connection with iniencephalus.

The distinction between iniencephali and exencephali proper, while, as in most classifications, obvious at the extremes, becomes indefinite at the boundary line. I have, following Saint-Hilaire and Ballantyne, confined the former to those cases where the whole brain was practically outside the cranium. A few cases might by different authorities be classed in either

category. Thus I find one case, entitled "Description d'un Cyclope Iniencephali," reported by Poelman.[25] There is one compound eye in the middle of the face below a proboscis; the cranium consists of rudimentary bones in the vault, with most of the occiput lacking, so that the rudimentary brain escapes and lies on the back over a cervical and dorsal spina bifida; there seems to be no brain within the cerebral box, and the picture confirms the text in describing an exencephalus synophthalmus. I found almost the counterpart of this specimen in the Rush College Museum.

Taruffi,[3] in the section on mero-acrania postero-spinale, states that he has collected ten cases where the cranial defect was in the occiput, and this class he considers synonymous with the

Fig. 24.—Cervical anterior spina bifida. Bonnaire's case.

iniencephalus of Saint-Hilaire. Taruffi's point of view differs much from that of other teratologists in regard to classification. Were we to consider all cases of occipital exencephalian in one class, there would be no division into exencephalus proper and iniencephalus. From a careful study of these ten cases I am constrained to believe that only one (that of Hull, p. 26) is truly an iniencephalus.

A. Reisel's case.[26] This is reported in 1683. A tom-cat was placed in sport about the neck of a pregnant woman. This so affected her that she gave birth, a few months later, to a living and robust boy who had a tumor on the back of his neck like a scrotum, adherent to the occiput and covered with hair of the same color as that of the cat. This tumor swelled with the

efforts of defecation and urination. It was lanced and several ounces of serous fluid evacuated. The child promptly died in convulsions at the age of about seven weeks. There is no mention of rhachischisis, and probably the case was one of notencephalus, that form of exencephalian which seems most compatible with continued life.

B. Hull's case (see p. 36).

C. Detharding's case.[27] The vault of the skull is flattened by reason of the rudimentary condition of the bones; the occiput is wanting in the condyloid portion, and the foramen magnum is enlarged into an irregular hole, out of which hangs a membranous substance; the cavity is only one-sixth the normal size and contains no trace of brain, in place of which is a soft, pulpy mass full of blood and covered by the membrane which hangs out of the foramen, as mentioned above. This specimen appears to be one of pseudencephalus.

D. Marye's case.[28] Hydramnios; female; still-born; head tilted back, with obliquity of cranium and face; frontals and parietals shortened and depressed; a membranous sac extending from the occiput over a spina bifida as far as the tenth dorsal vertebra and containing softened brain matter; the cranial cavity hardly exists and contains no brain. The case seems to be one of anencephalus. It is not included by Saint-Hilaire among the iniencephali, although reported in Paris several years before his book was published, and therefore doubtless known to him.

E. Scavone's case.[30, 3] Immature female fetus; sac hanging from the back of head; face like a toad's; parietals and frontals flattened and without fontanelles; no indication of cerebrum, cerebellum, or spinal cord; occiput wanting in the squamous portion. Thus the specimen has all the characteristics of anencephalus.

F. Calori's case.[31, 3] Female; great part of occiput lacking; complete spina bifida; marked cervical kyphosis; various anomalies of the intestines and of the umbilical and intestinal vessels. The original articles reporting this case and the preceding I have been unable to see, but refer to Taruffi's abstracts thereof.

G. Vrolik's cases.[32] In the table of Vrolik's atlas, referred to by Taruffi, are two specimens depicted, neither of which is an iniencephalus. The first shows the vault of the cranium almost flat, with the occiput rising slightly higher than the other bones of the vault; a wide defect in the occiput, out of

which issues a membranous sac containing rudiments of brain in two lobes; hardly any cranial cavity; spina bifida to the sacrum; cervical and dorsal kyphosis so marked as to throw the head forward on to the sternum; the brain lies along the cervical spine and upon the basilar portion of the occiput, but entirely uncovered by bony vault—in other words, exencephalic (Figs. 25 and 26).

The other specimen has a "waterfall" tumor issuing from the occiput and hanging upon the neck, but without spina bifida or tilting back of the head—a true notencephalus (Fig. 27).

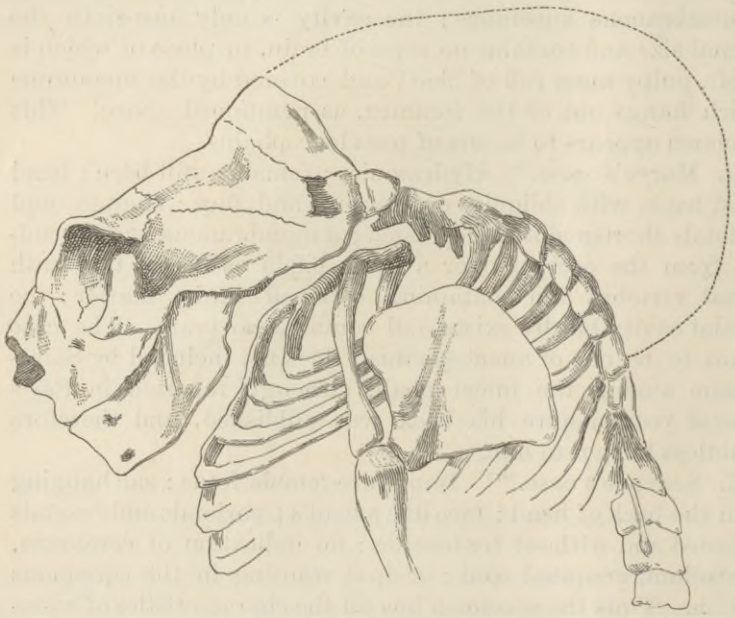


FIG. 25.—Exencephalus. Vrolik.

H. Castellani's case.[33] Female; premature about two months; head has appearance of that of a frog; neck furrow obliterated; a tumor of three lobes is attached to the posterior part of the head, resting upon the open spine; no cerebral cavity; cervical vertebræ wanting, right lung absent, and other anomalies. The case has every appearance of exencephalus proper.

I. Gallez's case.[34] Female with fungoid tumor behind the head, divided into three lobes by membranous septa and corresponding to the cerebral and cerebellar lobes; spina bifida, but

absence of cord ; face turned to left and head set well down into the chest. The whole brain seems to be in the outside tumor, and therefore, in spite of the author's classification of iniencephalus, it must be considered a case of exencephalus proper.

J. Stein's case.[35] This case is reported mainly to show a growth between and connecting the head and placenta. In addition the specimen, which is illustrated by a picture, shows the characteristics of derencephaly. From the posterior half and to the right of the head are given off two tumors from

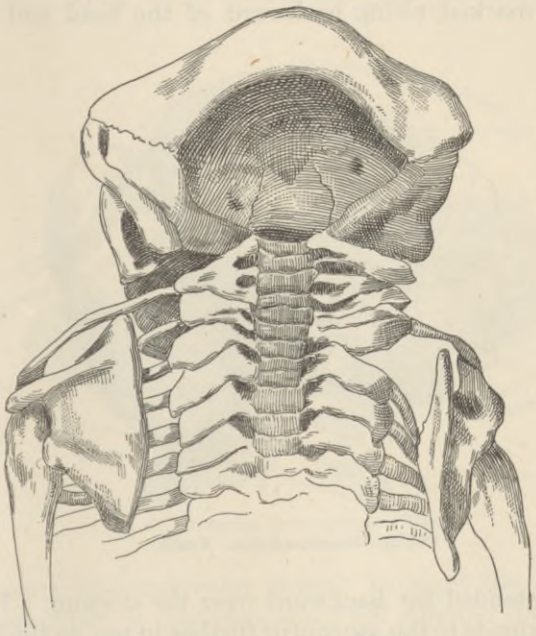


Fig. 26.—Exencephalus. Vrolik.

defects in the frontal, parietal, and occipital bones, the anterior one connected by a piece of placental tissue to the main placenta, which lies three inches from the head. The posterior tumor is composed of rudiments of brain tissue. No brain seems to be within the cranial cavity, which is itself very limited.

Besides these ten cases collected by Taruffi, only one of which is undoubtedly an iniencephalus, I find reference to two more whose originals I have not yet been able to obtain. One is a specimen depicted in Ahlfeld's "Atlas" [5] (Tafel xliii., Fig. 6), and

merely shows a skull with enlarged foramen magnum.[52] This may be either iniencephalus or notencephalus. Vrolik also reports a case (Table lii., Fig. 5) of dilated foramen magnum with diminished calvarium, but does not say how much brain was in the cranial box, nor does the picture allow an inference (Fig. 28). Saint-Hilaire states that one of the three cases of iniencephalus reported up to his time was by Burkhardt,[46] but I can find neither original nor abstract of this article thus far.

A résumé of the twenty-two cases collected by me shows several facts of interest.

It will be seen that, in all cases where mention is made, there has been a marked tilting backward of the head and the cra-

Fig. 27.—Notencephalus. Vrolik.

nium has extended far backward over the dorsum. The rhachischisis extends to the sacrum or further in ten, to the last dorsal or further in two, to the middle of the dorsal region in four, through the cervical region in three, and not stated in the remainder. There occurred anterior spina bifida in two cases, in one of these as far as the eleventh dorsal vertebra. Diaphragmatic hernia exists in four cases, is absent in three, and is not mentioned in the rest. Four children were born living but died soon after, seven were still-born, and the rest not stated. In fourteen the sex is stated; in only two was it masculine. Seven were born of primiparæ, four of multiparæ, and the rest not stated. Six were at full term, one in the ninth month, two in the eighth month, one in the seventh month, two in the

sixth month, and the rest not stated. In all cases where the fact was stated the spine was strongly curved antero-posteriorly and in three also laterally. In six cases there was hydramnios, in one a normal amount of amniotic fluid, and in the rest not stated. Club-foot existed in three, cyclopia in one, hydrocephalus in one, umbilical hernia in one, celosomia in two; other deformities or displacements of the alimentary tract were noted in eight; myelocele in one; in one the external oblique took an origin from the clavicle, and in one the gluteus maximus had an extra origin from the occiput. In nine there is no serious malformation mentioned besides the iniencephalus and spina bifida. Only twice is it stated that the pregnancy was

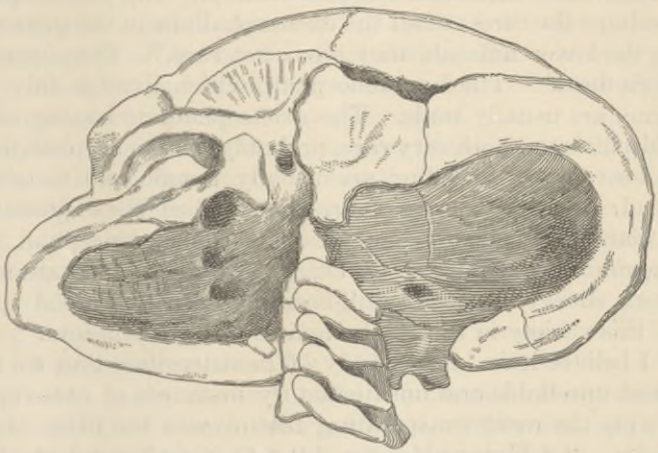


FIG. 28.—Occipital exencephalus. Vrolik.

double, once with uniovular twins. In eight the clinical history is not given, so that one cannot tell whether there was one or more fetuses. In the remainder the language would imply that the pregnancies were single.

The group of monsters having the main fault in the cerebro-spinal axis is a numerous one, if we include all forms from the small lumbar or sacral spina bifida to the rarest forms of exencephalus. The essential of the monstrosity is an abnormal openness of more or less of the cerebro-spinal bony canal. Spina bifida alone is tolerably frequent and, to a considerable extent, compatible with continued existence, especially when it can be relieved, as sometimes, by an operation. The anencephali are also relatively common. Puech[37] estimates that

they occur 14 times in 100,000 births—that is, 1 to 7,143. There is here a possibility of extrauterine life, although they are usually born dead. Some have lived several days,[35] usually dying in convulsions. They are more often female. Otto, in a collection of 600 monsters, reports 65 anencephali, of which 33 were female, 19 male, and 13 of unknown sex.[39] He described 23 exencephalians, among which is no iniencephalus. The exencephalian is not very infrequent[3] and averages nearly equal as to sex. It is said to occur 6 times in 100,000, or 1 to 16,666 births.[37] They are usually born before term and die very soon. Of the exencephalians the least rare is the notencephalus. This is not necessarily fatal; indeed, an adult notencephalus with intellectual faculties has been observed.[40] The proencephali are rather more rare than the last variety. The podencephali are perhaps the rarest of all the exencephalians in the *genus homo;* in the lower animals they seem not rare.[39] They usually are born dead.[40] The hyperencephali, which live for only a short time, are usually male. The exencephalians having also rhachischisis are both very rare, probably the iniencephali the most. As we have seen, they are usually female and usually born dead. To this variety Duges has applied the expressive term "diacrania occipito-rhachideana." The exencephali proper, the cranio-rhachideana of Duges, are also very rare and live but a short time or are still-born.[40] There is a good specimen of this variety in the Rush Medical College Museum.

I believe it is in the study of monstrosities that we find the most unreliable and unsatisfactory instances of case-reporting. To me the most exasperating feature was the titles of the articles. "A Unique Monster," "A Curious Teratological Case," "A Remarkable Monstrosity," and such lucid titles compelled the overhauling of hundreds of articles before finding one bearing on our subject. Few men seemed to have sought to classify their monster, and many seemed afraid to dissect it, obviously for fear of spoiling the esthetic appearance of the specimen. This last fear is groundless, for a fetus can be very extensively dissected and afterward completely restored to its former beauty by a little sewing and cotton-stuffing. In many, however, the greatest interest consists in the appearance of the skeleton. A good photograph of the monster and a good dried skeletal preparation, with the report of a complete and careful dissection, is of far more value to teratology than hundreds of uncut fetuses hidden away in jars and thousands of pages dealing mostly with the course of the labor and laboriously speculating on the possible "maternal impressions."

Teratology requires more nowadays than, as Prof. Cleland says, to classify your monster and then put a sesquipedalian name to it. We must needs speculate on the etiology. Heredity and atavism, while doubtless playing some sort of rôle in malformations in general, can hardly be thought to have much influence on exencephalic monsters. Number 6 is the only one here where heredity is reported as having the least influence, and even there it is more likely that some influence was at work peculiar to that mother.

In this presence I should not dare to mention maternal impressions, and I refer those interested in that form of mysticism to Foerster's unanswerable arguments against it.[41] "The cause of malformations is little known, but, from experiments on birds, is found to be physical and mechanical and pathological, and not mysterious" (Lancereau[42]). The term "lusus naturæ" accounts not at all for teratological cases. Nature never errs and never has freaks, although her course is sometimes modified by circumstances accidental or experimental.

There have been two sets of views as to the origin of malformations and monstrosities. The pathological theory explains the anomaly by disease of the fetus, recognizing it as an entity like an adult. If the pathological influence acts very early in the history of the organism it influences the general harmony of the whole, especially modifying the nervous system, disarranging the relations, and producing serious vices of conformation.[43] Coming later in fetal life, disease can only influence the form of parts, producing only local malformations or anomalies. The embryological theory, which is held now by the majority of teratologists, explains monstrosities and malformations by arrest of development, usually by pressure at a more or less early period of embryonic life. Cruveilhier[13] attributes great importance to contraction of the uterus, Panum to an arrest of development of the germinal spot or area pellucida, or in the fetal membranes, like the amnion. He admits the value of compression of fluid, as in hydrocephalus, and of contraction of cicatrices in the skin in compressing organs. Dareste[44] considers the cause of simple autosite monsters to be almost always due to arrest of development of the amnion and compression of the latter on the tissues growing beneath it. He experimented extensively on the eggs of birds, particularly of hens. He found that nutrition of the amnion came from the albuminous twists at each end of the yolk. These best convey the albumen from the white when the egg lies in certain positions and when

the egg is turned over daily. Swammerdam, as early as the eighteenth century, had found that the embryo could be modified in form by modifying the chrysalis. Dareste produced monsters by four means: the vertical position of the egg; applying an impenetrable covering to its shell; raising or lowering the temperature; unequal heating of different parts of the egg. The last method was used most. In an egg the germinating spot lies floating on top of the yolk, and in the usual situation this is the part which gets the most warmth. The eggs in the experiments were placed under a coil of warm water tubes in such a way that the heat was not equally distributed to the germinative area. Thus the parts getting the most warmth developed best, and other parts were correspondingly arrested. Different arrangement of the eggs produced different anomalies in the development of the germinating area, and the arrangement could even be so as to produce different anomalies at will. The arrest of development of the amnion, consequent on his treatment of the eggs, modified the evolution of the different parts of the embryo by the compression it exercised on them. This explains the association of several anomalies in the same individual monster. The slight differences in the date of the beginning of the compression cause the differences in the variety of the monstrosity, such as cyclopia, anencephalus, exencephalian, etc. The last, and iniencephalus as a variety of exencephalian, usually arose from an arrest of development of the cephalic hood of the amnion which is applied to the upper aspect of the head and compresses it. The cerebral vesicles are shortened and flattened. The rest of the cranium which has escaped compression goes on to ossify, and the variations of the places of the compression correspond to the different types of the exencephalic class.[7] In iniencephalus there must be in addition a compression from the part of the amnion lying behind the cephalic hood to account for the great foreshortening of the back due to the anterior curving of the spine and the tilting backward of the head.

On the other hand, noted authorities on teratology combat this theory of the causation of single monsters, and even Dareste himself seeks not thus to explain double fetuses. He even states that in all his experiments he never produced a double monster, although he has often studied them in process of formation.[45] Taruffi[3] objects that the amnion in these cases is often found normal in appearance. Cleland[46] believes that every vertebrate animal has at an early period a latent capabil-

ity of splitting itself up indefinitely. He would account for monstrosities as the results of over-stimulation of cell growth in the more specifical parts, and depending for particulars on the precise date or extent of the over-stimulation. When occurring soon after impregnation it leads to fissiparous division and two embryos; a little later to partial division of the central axis and a double monster partly united; later still it may cause imperfect closure of the neural canal and the production of a monster by defect, such as anencephalus or exencephalus. In favor of Dareste's theory has been urged the fact that monsters of the single variety so much more often occur in twin pregnancies, where compression could be easily exercised upon the parts of one of the embryos. However, in iniencephali it will be noticed that twins have seldom occurred. Again, hydramnios is a frequent accompaniment of monstrous birth, pointing to the probability of vascular disturbances and consequent dropsy of the amniotic membrane.

Syphilis is mentioned[46] as a possible cause of some of the changes in the embryo or its surroundings which lead to monstrosity. It is not difficult to conceive of such a cause acting upon the very early embryo itself or upon the maternal side through the placenta and fetal envelopes.

The real study of teratology is quite modern. There are no records of monsters found among the Greek, Roman, or Arabic physicians. In early times doubtless the occurrence of a monstrous birth was considered evidence of the wrath of the gods and therefore kept concealed, because it is hard to believe that monsters did not occur in all periods of human history. They were hardly mentioned, however, until after the revival of letters, and even then the reports are full of mysticism and abound in accounts of fabulous monsters. Ambrose Paré, in his work on surgery, published in the sixteenth century, describes many monsters, mostly from hearsay, among them a winged, griffin-footed and horned specimen, a bird-headed dog, a human-faced horse, mermaids and mermen. These, and many other possible and impossible forms, are depicted in old-fashioned woodcuts.[47] During the next two centuries similar works appeared by Wallreich,[48] Kalinkius,[49] Stengelius,[50] and Mauriceau.[51] The popular theory at these times was that monsters were the products of sexual union of widely differing species—after all, a more rational etiology than maternal impressions. Taruffi[3] quotes the case of the sexual union of an Arcadian shepherd with a she-goat, which resulted in the birth

of a kid with a human head. Even Bartholin[7] mentions, as something not remarkable, the burning at the stake of a young girl who gave birth to an anencephalus which was supposed by the teratologists of the day to look like a cat. This occurred in Copenhagen in 1683, "ob lasciviorem cum fele jocum." Buffon in 1800 made some attempt at scientific classification, but it was left for the Saint-Hilaires, especially the younger, to elaborate a scheme of classification which fills the needs of to-day. Modern teratologists have devoted themselves largely to the etiology of the subject, and although the advance is relatively small and theories differ much within certain limits, still much has been done and is being done. Teratology is the pathology of embryology, and the advance of the last must contribute to the advance of the first. Experiment will in the future, as now, do much to clear up these obscure problems, but we will still continue to be largely dependent upon careful dissections clearly reported.

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