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# INIENCEPHALUS.

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## INIENCEPHALUS.

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sity of Vermont College of Medicine.*

Fetal monstrosities always have been and always will be an interesting subject to the scientist. Although it has been the subject of careful study for at least two centuries (before that, the investigations were too vague and entered too much into the supernatural to be of any real value), the cause is by no means definitely settled. The earlier writers ascribed them to some appalling object which the pregnant woman had seen or to the anger of the gods and the appearance of a monster in the family was supposed to portend all kinds of disaster, as if giving birth to a monster was not punishment enough to inflict upon any unfortunate woman. Maternal impressions have not been entirely dismissed from the list of causes. The writer was told quite seriously a few years ago by a medical man that one of his own children had been born with a deformity of one hand which he put down to the fact that the coachman whom he

had employed at the time when his wife was carrying the child had a deformed hand and that his wife could not help gazing at this deformity whenever the man was driving. Again, as late as 1889, the editor of "The Cyclopedia of Obstetrics and Gynecology," in connection with a paper on "The Influence of Maternal Impressions on the Fetus," says that "there can be no question, we think, that there must exist some causal relation, although it is one of those hidden mysteries likely ever to remain beyond human ken." Cannot this causal relation be explained by the shock of seeing some unusual sight causing a contraction, either local or general, of the uterus? It seems to me as though this might be answered in the affirmative, and, if so, pressure upon the ovum would follow with a consequent injurious effect upon its growth. That pressure, change of position, etc., will materially affect the growing ovum has been proved beyond all dispute by Dareste,<sup>1</sup> who, in several thousand experiments, produced all types of monstrosity. In many cases, however, there must be something in the male or female germ itself which has an influence in the production of malformation: how else could those deformities which prevail in certain families, and which are almost characteristic of that family, be explained?

A monstrosity is defined by Geoffrey St. Hilaire<sup>2</sup> as a deviation from a specific type of a complex, very serious, viscious character, congenital and apparent externally. Before

the time of St. Hilaire, there was no scientific classification of monsters, but he divided them primarily into (1) single and (2) composite.

A single autosite monster results when there is either an absence of a part of the elements of a single individual or some serious modification in the connection and disposition of these elements, their complete number being preserved. Single monsters are themselves subdivided into:

(a) Single autosite monsters which are capable of nourishing themselves by the play of their own organs. Here, only a lesser number of organs are affected, the remainder being normal.

(b) Single omphalosite monsters, leading to imperfect life carried on only by their connection with the mother and which ceases when the cord is broken.

(c) Single parasitic monsters, the most imperfectly developed of all, are inert, irregular masses lacking even an umbilical cord. They are implanted directly on the generative organs of the mother.

Composite monsters are those in which one finds united the elements, more or less complete, of two or more individuals. They may be subdivided into (1) double and (2) triple monsters.

(a) Double autosite monsters are composed of two individuals, offering the same degree of development, both contributing to the common life and of which each is analogous to an autosite.

(b) Double parasitic monsters are composed of two very dissimilar and very unequal individuals, the one complete or almost complete, the other not only much smaller but very imperfect and nourishing itself at the expense of the first, of which it is a parasite.

The kind of monstrosity which forms the subject of this paper is included in tribe three of the orders of autosites in Sainte-Hilaire's classification, *i. e.* where the posterior part of the head is gravely modified:

(a) The brain is still existing but is more or less incomplete and is placed, at least in part, outside of the cranial cavity, which is itself more or less imperfect, *i. e.* "Exencephalia."

(b) The brain no longer exists but is replaced by a red tumor mass of vessels, *i. e.* "Pseudencephalia."

(c.) The brain is completely absent, the vault of the skull being deficient, *i. e.* "Anencephalia."

*Exencephalia*.—These are characterized by a deformed brain more or less incomplete and placed, at least in part, outside of the cranial cavity, which is itself very imperfect. This is the first link in a long series of genera which will lead us, by an almost insensible transition to the acephalic monsters. All members of this group present exencephaly more or less well marked, with the following genera:

A. No spinal fissure.

1. The brain is situated in large part outside of the cranial cavity and behind the cran-

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- ium which is open in the occipital region, *i. e.* "Notencephalus."
2. The brain is situated to a large extent outside of the cranial cavity and in front of the cranium which is open in the frontal region, *i. e.* "Proencephalus."
  3. The brain is situated to a large extent outside of the cranial cavity and above the cranium, the upper wall of which is wanting, *i. e.* "Podencephalus."
  4. The brain is situated to a large extent outside of the cranial cavity and in front of the cranium, the upper wall of which is almost absent, *i. e.* "Hyperencephalus."

B. Spinal fissure present.

1. The brain situated to a large extent within the cranial cavity but partly outside of it, behind and a little below the cranium, which is open in the occipital region, *i. e.* "Iniencephaly."
2. The brain situated to a large extent outside of the cranial cavity and behind the cranium, the superior wall of which is lacking to a great extent, *i. e.* "Exencephalus."

In all the preceding genera (not—, pro—, pod—, and hyperencephalus), whether the vault of the cranium were open at only one point or almost entirely absent, the rhachidian canal still preserves normal disposition, at least in the greater part of its extent; some vertebrae, for instance the first cervical, could very well have been affected by a spina-bifida,

but there was no external spinal fissure. The iniencephalia, on the contrary, as well as the exencephalia, are characterized by the very abnormal condition of their rhachidian canal, which is open either to a very great extent or in its entire length. The exencephalia and iniencephalia have also excellent distinguishing characteristics in the conformation of the cranium, the first resembling the hyperencephalia and the latter notencephalia.

Iniencephaly is very rare. Sainte-Hilaire only knew of one case. H. F. Lewis<sup>3</sup> collected twenty-two cases which he reported in the American Journal of Obstetrics in 1897. To this list, we are able to add six more, making in all twenty-eight. Of these, one very good undissected specimen can be seen in the pathological museum of McGill University, where also the mounted skeleton of the authors' specimen is preserved. In studying this series of cases, various points in common are to be observed. For instance, the three chief characteristics are occipital defect, spina-bifida, and fetal retroflexion. Out of the twenty-eight cases, the sex was mentioned in twenty-three and of these no less than nineteen were females. In six there was a superabundance of liquor amnii, while hydrocephalus was coexistent once. Seven of the fetuses had reached full term, two eight and one-half months, two eight months, one six months, one five and one half months and one five months. The presentation was vertex in two, foot in two, face once and pelvic once. The mon-

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ster was accompanied by a twin twice. In but one case was there any history of any woman having previously given birth to a malformed child and that was in case six, where the monster was the woman's fourth child, her first one having a spina-bifida, while the second and third were normal.

Lewis divides his cases into three classes. First those without any encephalocele, which he terms "Iniencephalus Clausus." Secondly, those having only a small encephalocele, those, with the latter, being true iniencephaly. Lastly, those cases where there is a large encephalocele but where the other characteristics of iniencephaly are so well marked as to leave no doubt as to their character. This class he calls "Iniencephaly Apertus."

#### INIENCEPHALUS CLAUSUS.

1. Ahlfeldt<sup>4</sup> reports the following case of Fleck's. The fetus was a female with tilting backwards of the head. The anus was much higher up the back than usual. There was no external tumor to be seen but there was a complete spina-bifida as far as the sacrum, a fusion of the cervical vertebrae and a wide opening in the occiput, the margins of which were attached to the transverse processes of the dorsal vertebrae.

2. There is one specimen in the museum of the Royal College of Surgeons of England.<sup>5</sup> The fetus is a male with distortion of the vertebral column, defective development of the

occipital bone and but partial closure of the vertebral canal posteriorly. There is a wide rhachischisis of the cervical and first six dorsal vertebrae and the head is tilted backwards until the margins of the occiput and vertebral openings touch and have become united by ligaments. The frontal and parietal bones are very large while the supra-occipitals are rudimentary. There are only five cervical vertebrae and the laminae of these are fused together. There is a second spina-bifida in the lumbo-sacral region and both feet are the subject of talipes. Hydrocephalus co-existed in this case, there being no opening through which the dropsical effusion could escape from the cerebro-spinal cavity, which might cause the coverings of this cavity to stretch. This condition favors the theory of Dareste as to exencephalus being caused by vascular changes, being produced by compression of the defective cephalic covering of the amnion.

3. Coffin's<sup>6</sup> case was that of an eighth month child with a flat chest, scapulae in front of the lateral line and the head tilted back. The posterior fontanelle communicates with an opening in the skin. The spine is small and is curved anteriorly. The coccyx is absent and its usual site is occupied by the anus. The spine consists of three bones, between the first and second of which an opening leads into the cranial cavity. The posterior portion of the occiput is absent and the vertebrae articulate with the basilar portion. The third bone corresponds to the sacrum. The vagina opens

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very far back in the site usually occupied by the anus.



DR. EVANS' CASE, No. 25. (Anterior.)

4. C. A. Yocum<sup>7</sup> reports one case in which the head is retroflexed almost to the sacrum. The rest of the fetus is similar to case No. 9, except that the abdomen is closed.

5. Landucci's<sup>8</sup> fetus is a female. The cranial bones are very movable and the head is tilted backwards, but the chin appears to be fused to the sternum so that the ears are just above the shoulders. The umbilical sac protrudes from the abdomen and contains coils of intestine. The pelvis is tilted backwards and the anus opens on the back a few centimeters below the scalp. The frontal sutures are ossified. The parietals extend to the spinal column except for the intervention of narrow pieces of the occiput which are attached to the spine at the eleventh and twelfth dorsal vertebrae. The spinal canal is open as far down as the last dorsal vertebra but is covered by the overhanging cranium. A deep fissure containing the medulla, extends into the base of the occiput. The cervical vertebrae turn forwards and upwards so that the lower ones lie beneath the lower jaw. In the dorsal region the spinal column turns backwards. The lumbar vertebrae are better developed and each is complete but a small spina-bifida involves the last two lumbar and first sacral vertebrae. The bodies of the first nineteen vertebrae are fused and the ribs are irregular in shape and are partly fused together.

6. Ballantyne.<sup>9</sup> This was a still-born female fetus. It was the mother's fourth child, the first one having a spina-bifida. It measured 30 cm. in length and 19 cm. from the vertex to the coccyx. The occipito-frontal circumference was 31 cm. and the occipito-mental 34 cm. The head sharply flexed back-

wards on the trunk, so that the occiput came into close relation with the open spinal canal in the lumbar region. There was no indication of a neck. The abdomen was very prominent. A sagittal frozen section showed the spine running upwards and forwards towards the centre of the fetal mass. Several lumbar and dorsal vertebrae were absent and the occiput was wanting in the lower part. There was a slight lateral distortion of the trunk.

7. Ballantyne.<sup>9</sup> The fetus was a female measuring 23 cm. in length, the body alone measuring 14 cm. The head and trunk formed a rounded mass between 12 and 13 cm. in its antero-posterior diameter. The head was so sharply retroflexed that the skin over the occiput was directly continuous with that over the coccyx. The occiput was defective in the lower part. Many vertebrae were absent and the spine had a forward and upward direction so as to reach the centre of the mass formed by the head and trunk. The face looked directly upwards. The mother had hydramnios.

8. Ballantyne<sup>10</sup>. The fetus was born at the seventh month and was a typical example of iniencephaly. The mother was 42 years of age and a tenth para. One child died at the age of five of "trouble in the head," accompanied by starting and screaming at night and great wasting. At the seventh month, fetal movements stopped and the ovum came away entire. Incision of the sac allowed of the escape of about a pint of "pea-soup like fluid."

The fetus was macerated and had an eighteen-inch cord.

9. Lewis'<sup>11</sup> case was one of uniovular twins of which one was a normal five and one-half months child and the other was the monstrosity. Both children presented by the feet. The malformed fetus measured eight and one-half inches in length with an apparent bending backwards of the spine so that the anus was only three-fourths inch from the back of the head. The face looked forwards and upwards and the cranium was turned so far back as to cover most of the spine. No depression existed between the chin and the thorax. Dissection showed that the parietal bones overlapped the upper two-thirds of the spinal column to which they were attached. The spinal canal was open from the upper end to the second lumbar vertebra and the cord could be seen lying at the bottom of the canal. The only abnormal cranial bone was the occiput, the squamous plates of which were represented by an acute triangle on either side of the basilar openings and which articulated with the parietals. The margins of the opening in the occiput were closely attached to the separated laminae of the cervical, dorsal and first lumbar vertebrae.

10. Rush Medical Museum specimen. The fetus was a female at full term.

Measurements:

Length of body . . . . .	12½ inches.
Anus to cranium . . . . .	2⅝ “
Length of leg . . . . .	6¾ “

Length of arm . . . . .	$7\frac{1}{8}$	“
Chin to perineum . . . . .	$7\frac{7}{8}$	“
Bi-temporal diameter . . .	$3\frac{5}{8}$	“
Mento-bregmatic diameter		
	$4\frac{1}{4}$	“
Occipito-frontal diameter		
	$4\frac{3}{8}$	“

There was great tilting backwards of the head, so that the face looks forwards and upwards, leaving almost no sulcus beneath the chin. An opening in the abdominal wall, admitting three finger-tips, is seen at the umbilicus, the attachment of the cord being to the left of this opening which is fringed by folds of peritoneum and through which coils of intestine protrude. An incision down the centre of the back shows a spina-bifida extending to the tip of the coccyx. The cranium extends to the lumbar region and is attached to the lateral processes of the vertebrae by ligaments. No encephalocele is to be detected. On dividing the ligaments and turning the head forwards, a rhachischisis of the cervical and dorsal vertebrae is seen, at the bottom of which lies the spinal cord with the brain on top. There is considerable lordosis of the cervical region of the spine and right scoliosis of the whole column. The openings in the occiput forms a large foramen magnum measuring  $1\frac{5}{8} \times 1\frac{1}{8}$  inches. In the lumbar region, there is a loose piece of cartilage the size of two split peas attached to the spine by ligaments. The anus is pervious and there is slight talipes affecting both feet.

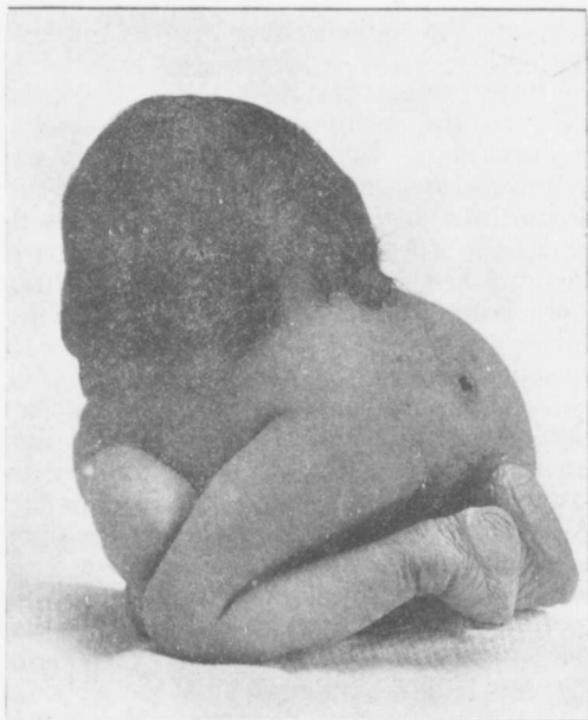
## INIENCEPHALUS APERTA.

11. In Duge's<sup>12</sup> case, the head seemed confounded with the thorax and turned back so that the occiput appeared lost between the shoulders. The anterior part of the neck is flush with the chin and sternum. Just below the occiput, on the back, is a small protuberance the size of a walnut, covered by a thin reddish membrane which continued down to the sacrum. The cranium is depressed and projects in front of the occipital opening. The face is oblique and projecting. Some of the cranial bones are welded together while others have cartilagenous areas. The upper occipitals are flattened and run parallel to the base of the skull. The spine is open posteriorly as far as the sacrum. Its cervical portion lies under the basilar process and its dorsal portion under the base of the cranium. The cerebral hemispheres lie within the skull, the medulla being in the occipital opening while the rest of the brain is in the fungous mass behind the head, from which come all the cranial nerves. Hernia of the stomach and the left lobe of the liver had taken place into the thorax through the diaphragm.

12. Potthof's<sup>13</sup> case shows considerable tilting backwards of the head with obliteration of the submental furrow. The head and body form almost one mass, to which are attached two long arms and two legs. The anus lies near the back of the head and quite a distance from the vagina. A small spongy tumor lies

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at the angle formed by the parietal bones and the dorsum. A complete spina-bifida exists and an opening in the anterior abdominal wall allows the escape of part of the abdominal contents.



DR. EVANS' CASE. (Posterior.)

13. Cruveillier's<sup>14</sup> case was that of a female fetus with its head strongly retroflexed upon the spine. There were two tumors behind

the cranium, the chin was continuous with the chest, the nose was flattened and the ear deformed. The mouth looked as if a string were pulled tightly across it and the cheek was continuous with the palate. The first tumor was just behind the sagittal suture and was divided from the second, which was to the right and below it, by a fibrous partition. Both of these communicate with the cranial cavity and are filled with membranes and a serosanguineous fluid. No trace of the squamous part of the occipital bone exists but the tumors protrude through the large opening between the parietals. These bones extend over the parietals and first four dorsal vertebrae which are cleft both anteriorly and posteriorly, thus forming double bodies. No basilar part of the occipital bone can be seen and there are only two rudimentary lateral occipitals, one on each side behind the temporals, forming extensions of the cervical vertebrae. 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 dorsal vertebrae. The diaphragm is absent on the left side, allowing part of the intestines and the left lobe of the liver to enter the thorax. The left lung is compressed and the right lies partly in the neck. The stomach and duodenum are partly in the posterior mediastinum and the esophagus shortened and somewhat invaginated.

14. S. W. Drew and J. B. Jackson<sup>15</sup> report a case of a female infant which lived half an

hour. It weighed five pounds and measured fourteen inches around the thorax. Club feet were present. There was no neck furrow and the arms were very long. The uvula was cleft and the lungs were irregularly fissured. The brain appeared at the upper part of the spine, looking red and vascular, and the size of an English walnut. A large mass of brain had remained within the cranial cavity. The spinabifida was complete and there was a separate cord on each side of the spinal canal communicating with the medulla. The occipital bones were joined to the spinous processes of the vertebrae as far down as the first lumbar except on the left side where there was a considerable deficiency. The antero-posterior spinal curve was excessive, the upper dorsals and middle lumbar forming prominent points, the pelvis and ends of the ribs being thus brought into close proximity with one another. The cranial cavity was quite capacious, leaving considerable space between the frontals and the parietals and the base of the skull. The basilar and lateral processes of the occipital bone were well developed. The posterior portion consists of one piece on each side, sending a narrow process around behind the parietals to meet its fellow of the opposite side. The spine is curved antero-posteriorly and laterally and the thorax is very prominent. The anterior half has no connection with the ribs. On the left side, seven of the ribs are partially fused, while the twelve on the right side are chiefly separate.

15. Lawther<sup>16</sup> reports a case in which the fetus was a macerated female. A longitudinal section was made into the vertebral column and the two halves were drawn to one side. At the middle of the transverse line joining the apices of the shoulders, the spine curved downwards and forwards, the cord embedded in the centre. The cervical vertebrae were only represented by a cavity extending up into the floor of the cranium and into this cavity the curve of the spinal column projected. The occiput was only represented by a narrow osseous arch connected with the parietals. There was no cerebellum and only an imperfect cerebrum in the form of a thin covering of brain substance enclosing an indefinite cavity. The only visible nerve was the optic. The diaphragm, mediastina and pericardium were absent. The pleura was continuous with the peritoneum and, after investing the organs, passed into the funis, forming a pouch at the placenta. The cord measured less than four inches. The esophagus ended in a blind pouch, the stomach being absent. The lungs were rudimentary but the kidneys were large.

16. Vernier's Case.<sup>17</sup> The mother had hydramnios and the child was born at the sixth month. There were spina-bifida, a tilting backwards of the head and a pouch, containing brain matter, behind the cranium. The two halves of the occipital bone did not meet in the middle line but were attached to the lateral parts of the upper cervical vertebrae. Cyclops and club feet were also present.

17. In Remfrey's<sup>18</sup> case the fetus is a dead-born female. During delivery (which was a footling) the cord, which only measured two inches in length, broke. The face looks upward and the head is tilted so far backwards that scalp and buttocks are flush with one another. On the right side, and projecting over the crest of the ilium, is a sac which is collapsed and from which exudes brain matter. The chief flexion is at the occipito-atloid joint. The occipital bone is poorly developed and covers such an extent of the spine that it nearly touches the iliac crests and the right gluteus maximus arises from it. The body has a flexion towards the right side.

The diameters are:

Fronto-mental . . . . .	2 $\frac{3}{4}$ in.
Bi-mastoid . . . . .	2 $\frac{3}{4}$ "
Forehead to buttocks . . . . .	4 $\frac{3}{4}$ "
Bi-trochanteric . . . . .	2 $\frac{7}{8}$ "

The liver is large and irregular in shape, having a pedunculated mass the size of a cherry projecting from the lower edge. The tabular part of the occiput is represented by a crescentic plate on each side, attached to the posterior margin of the parietal bone and separated from its fellow of the opposite side by a wide opening, the floor of which is partly formed by the spine. The other cranial bones are normal. The spine curves first forwards and then abruptly backwards so that the last vertebra is only one inch from the posterior margin of the left parietal bone. It also

curves towards the left. The ribs are irregularly developed, being more or less fused together.

18. Case from Rush Medical Museum. The fetus is a female measuring:

Head to foot	.....	10 $\frac{3}{8}$ in.
Head and body	.....	5 13-16 "
Trochanter to sole	....	Left 5 $\frac{3}{8}$ "
		Right 5 $\frac{1}{2}$ "
Acromion to finger-tip.	Left 6	"
	Right 5 $\frac{7}{8}$	"
Coccyx to cranium	.....	2 9-16 "
Bi-temporal	.....	2 13-16 "
Mento-bregmatic	.....	3 9-16 "
Occipito-frontal	.....	3 $\frac{1}{4}$ "
Bisacromial	.....	2 $\frac{7}{8}$ "

The toe-nails were less than half way out to the tips of the toes and the finger-nails less than three-quarters. The chin was flush with the sternum and there was double talipes. The usual tilting backwards of the head and foreshortening of the back were to be observed, while the face looked forwards and a little to the left. In the median line of the back there is a soft tumor covered by membrane through which can be felt a cleft in the spine. The tumor measures  $1\frac{1}{4} \times 15-16$  in. and occupies most of the dorsal region. Protruding from the bony defect in the skull to the right of the median line and hanging over the right shoulder is another soft tumor covered by skin from the anterior third of which grow hairs. This is an encephalocele and measures  $2\frac{3}{8} \times 2\frac{1}{4}$  inch.

Under the skin, the lateral tumor is enclosed in a double sac, the outer part adhering to the skin (dura) and the inner being free. Through an incision in the latter, disorganized brain-matter escapes. The cavity is continuous with that of the cranium through the bony defect. In dissecting the dorsal tumor, numerous fibers of the spinal cord and nerves, these being adherent to its under surface, are cut. The parietal bones are of equal size, their apparent inequality being due to the twisting of the head to 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, articulate with parietals at the lambdoidal sutures and with the basilar part of the occiput at the base below on each side. The margins of the foramen magnum lie flat upon the spinal column and are ligamentously attached to the rudimentary laminae of the widely open vertebrae. Thus the occiput covers the spinal canal, causing 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 vertebrae are better developed and approximate each other more closely than elsewhere and are connected by strong ligaments. The spina bifida is complete to the tip of the coccyx and the canal is flattened. The compressed fibers of the cord and the nerves from it lie

in the canal, and, in the region of the dorsal tumor, are spread out over the inner surface of the latter. This tumor also contains a sac filled with clotted blood and, with its pedicle, passes up through the constricted portion of the spinal canal under where the left lateral occipital plate touches the dorsum. This sac is a myelocoele. Lordosis of the cervical and kyphosis of the lumbar vertebrae are present, and the axis of the spine is turned to the right in the dorsal and cervical regions.

19. Hull's<sup>19</sup> case. A defective brain lay almost entirely outside of the cranial cavity on the back of the neck. There was a spina bifida from the neck to the sacrum. The squamous portion of the occiput was absent and the frontals and parietals were small and depressed. Three ribs were fused and talipes was present.

20. Gros'<sup>20</sup> case. The mother was a primipera and the labor was complicated by intestinal disturbances. Slight hydramnios was seen. There was no occiput and the parietals were small while a large cerebral tumor lay behind the cranium. A spina bifida was present in the cervical region. The mouth was divided into two parts by a horizontal band.

21. Budin's<sup>21</sup> Case. The fetus was a female which weighed 750 grams. A large tumor was present at the posterior part of the cranium, arising below the superior angle of the occiput and descending almost to the sacrum. There was hair on the anterior surface of this tumor. Between the skin and

dura, there was quite a quantity of fluid, as also in the sub-dural and arachnoid spaces. The tumor also contained the cerebral-hemispheres. The cerebellum was in the right occipital fossa within the skull and on top of the medulla and part of the spinal cord. There was a bony opening in the cranium bounded by the occiput and the cervical and upper six dorsal vertebrae. The cervical vertebrae formed a deep curve with its concavity directed posteriorly. The squamous part of the occiput was rudimentary, consisting of plates at the sides of and above the cranial opening. The basilar and rest of the cranial bones were normal. The thorax was pigeon-shaped on account of the anterior curve of the spine. The head was tilted back with a corresponding foreshortening of the dorsum.

22. Roger's<sup>22</sup> Case. An excessive amount of liquor amnii was present and the child, a female, weighed  $8\frac{1}{2}$  lbs. At the basis cranii was a soft tumor as large as the head. The back of the child was soft and no vertebrae could be felt at the lower part. The first cervical vertebra articulated with the sphenoid or detached basilar portion of the occiput; thence the spine descends to the level of the manubrium sterni, where it turns backwards and upwards and terminates with a short downward curve between the spines of the scapulae. There is no bony column between this and the sacrum. The inferior part of the occiput does not articulate with any other bone but curves upwards into the cranium, leaving

an opening into it. There are no articular condyles on the occiput. A hard tense swelling, containing intestines and brown gelatinous fluid, occupies the lower abdomen down to and continuous with the labia majora. The anus was imperforate.

23. Bonnair's<sup>23</sup> case. This fetus was one of twins, its fellow being normal. The amnion of the monster was very dropsical. Its face, composing the chief part of the head, looked upwards and was set so deeply between the shoulders that the ears touched the acromion. The chin and sternum were continuous and the spine was so curved that it gave the body the appearance of being greatly shortened and the limbs an apparent elongation. Behind the head, an encephalocele extends as far back as the lumbar region, having its anterior third covered with hair, the rest with membrane. The spina bifida is complete and contains the flattened cord. The frontal and parietal bones are rudimentary. The occiput contains a large opening and its basilar portion is in two lateral pieces. There is complete splitting of all the cervical and upper ten dorsal vertebrae, forming a Y with the rest of the spinal column. The cervical parts of the Y curve sharply backwards so as to almost touch the atrophied dorsal vertebrae. Below the cord, a hernial pouch, containing part of the esophagus, projects through the fissure in the vertebrae. Lewis has found two cases where this splitting of the upper part of the spinal column complicates iniencephaly.

24. Ballantyne (Edin: Obstetrical Soc: Trans: Vol: 24) showed at the Edinboro Obstetrical Society in 1899 frozen sections and photographs of an iniencephalic monster. The fetus was a small male. The head was sharply flexed backwards upon the trunk; the basis cranii looked directly backwards; the cervical spine showed marked lordosis with compensating kyphosis in the dorso-lumbar region together with some torsion. The cranial vault was defective in the region of the posterior fontanelle, allowing the cerebrum to project into a sac which lay on the back of the fetus. The cartilagenous part of the occipital bone was directed downwards, leaving in front of it a very large foramen magnum, in which lay the cerebellum and spinal cord. The bladder contained colorless urine and in the dorsal sac was blood clot and cerebral substance, the sac probably having ruptured during labor.

25. Evans' Case. This specimen is in the pathological museum of McGill University, Montreal. The fetus is a small female, showing marked retroflexion of the head, so that the occiput merges into the back at a point 4.25 cm. above the anus. The face looks almost directly upwards, its longest diameter running from the occiput to the upper lip. The fetus measured 39 cm. in length. The other measurements were:

Anus to base of nose.....	14.25 cm.
Occipito-frontal .....	9 cm.
Occipital-mental .....	9 cm.

Sub-occipito bregmatic . . . . .	8 cm.
Bi-parietal . . . . .	9 cm.
Bi-temporal . . . . .	8.5 cm.
Fronto-mental . . . . .	6.25 cm.
Circumference of head at level of O. F. diameter . . . . .	28.5 cm.

The face is well formed and the head well covered with hair, which is dark and extends very low on the forehead but not abnormally low at the sides. Behind, this hair stops abruptly at the junction of the occiput with the back. The anterior fold of the neck is obliterated by the retroflexion of the head. Both ears are abnormal in form. The extremities are flexed closely upon themselves and there is a left talipes varus. In the middle line of the back just below the occiput is a round, bluish-red elastic nodule the size of a bean, indicating a spina bifida. No skiagraph could be obtained owing to the saturation of the specimen by the mercurial solution in which it had been kept.

26. This specimen was presented to McGill Pathological Museum by Dr. Dewar of Ottawa. The mother was the subject of rickets and cerebral syphilis. She gave birth to a full term female fetus of large size, measuring 33.5 cm. from the heel to the middle of the anterior fontanelle. The head is retroflexed, the occiput, just below the posterior fontanelle, becoming continuous with the back in the intra-scapular region 8.25 cm. above the anus. There is also slight lateral flexion of the trunk to the right. The arms are in extension and

the lower extremities are flexed upon themselves. Through the retroflexion of the head, the fold of the neck anteriorly is completely obliterated. The face looks obliquely upwards, the anterior fontanelle upwards and backwards. The facial expression is natural and the head is covered with thick black hair, which grows far down laterally below the ears to the shoulders, running particularly low on the left side where it reaches the level of the scapula. On this side, a strip of scalp about 3 cm. wide on a level with the forehead is quite bare of hair. The ears are placed low down near the neck, and the left one is slightly anomalous in form. The diameters of the head are:

Occipito-frontal . . . . .	10.25 cm.
Occipito-mental . . . . .	11.00 cm.
Sub-occipito bregmatic . . . . .	9.00 cm.
Bi-temporal . . . . .	8.00 cm.

The trunk is very well nourished. In the middle line of the back, about 2.0 cm. below the occiput is a small tuft of hair; about 2.5 cm. below this is a dimple and at a similar distance lower down, just a little way above the coccyx, is another depression, slightly deeper than the first. The anus is perforate. The lower part of the thorax and the abdomen are prominent and rounded. There is an umbilical hernia about the size of a walnut. It is a soft reddish-brown mass of tissue and is elastic as though it contained fluid. The cord arises from this mass. The upper extremities are normal and the nails are well

formed. There is a marked talipes equinovarus of the left foot and a slight one of the right foot. On the toes, the nails are rudimentary. In the skiagraph, the basis cranii is seen to be high up and the occiput to be adherent to the bodies of the vertebrae in the lower dorsal region. There is a lordosis of the lumbar and lower dorsal regions.

27. The author's specimen was given him by Dr. A. R. Griffith, Montreal, in whose practice the case occurred. The mother is a healthy 2-para, 27 years of age. Her last child was only 11 months old when this monster was born and it had been nursed for nine months. Both of the previous children were healthy and well formed and there was no history of any malformation having occurred among any of the parents' family connections. Nothing unusual had been noted in connection with the pregnancy and the presentation was normal. Delivery was natural and easy. There was a very large amount of liquor amnii but the placenta was rather small. The measurements are:

Occipito-frontal . . . . .	9.25 cm.
Occipito-mental . . . . .	10.00 "
Bi-temporal . . . . .	6.5 "
Bi-temporal . . . . .	6.5 "
Vertex to heel . . . . .	29.00 "
Head and trunk:	
Length . . . . .	16.00 "
Circumference around	
shoulders . . . . .	34.00 "
Occiput to anus . . . . .	5.00 "

Cord to base of penis.....	2.50	“
Shoulder to finger-tip.....	16.00	“
Fold of thigh to knee.....	7.00	“
Fold of thigh to external maleolus.....	10.00	“

A condition of double talipes exists. Inspection of the cord shows no abnormality until a transverse section is made when the artery is seen to be accompanied by only one vein. The child is retroflexed upon itself and rotated to the left upon its vertical axis so that the face looks slightly upwards and to the left. The eyes, mouth, and nose are well formed and the ears are deepset but symmetrical. The whole head is covered with dark hair which has the usual distribution, except that it extends abnormally low on each side posteriorly. The anterior neck sulcus is obliterated except just in front where there is a small submental hollow. Posteriorly, a globular swelling, cystic in character, extends from the anterior angle of the posterior fontanelle to the middle of the back and on either side as far as the middle line of the neck. This mass gives the head an ovoid shape, the long diameter of which runs from left to right with an inclination slightly backwards and downwards, owing to the left lateral flexion of the spinal column. No sign of an abdominal fissure can be observed anteriorly, but posteriorly, immediately below the cystic swelling, is a spina bifida which measures 3 cm. in length and .12 cm. in its broadest part, which is above. This fissure is covered by a thin whit-

ish membrane which extends further out on the right side than on the left. The spina



Author's Case, No. 27.  
(Anterior)

bifida extends as low down as the 2nd lumbar vertebra, while from the 11th dorsal up it

forms the floor of the cranial cavity. On the left side, the occiput is attached by ligaments to the transverse processes of all of the cervical and upper six dorsal vertebrae, while on the right side the attachment extends down as low as the 10th dorsal.

On opening the abdomen, the liver is seen to be larger than normal and to be displaced downwards. It is attached to the anterior abdominal wall by a thin velamentous membrane, the other hepatic ligaments being normal. The left lobe is decidedly the larger of the two. The heart contained a patent foramen-ovale but was otherwise normal. The kidneys were represented by a horse-shoe shaped kidney, lying across the spine, with the two ureters running from it to the bladder.

Dissection of the scalp showed the anterior and posterior bellies of the occipito-frontalis to be well developed and to be united by fascia over the vertex while the posterior portions lost themselves over the coverings of the nontencephalocele. This latter formed a bi-lobed swelling, the larger of the two (the right) being about the size of a tangerine orange and the smaller that of a large walnut. The sac was covered by a thin, tough, fibrous membrane directly continuous with the periosteum covering the bones of the cranium and but slightly adherent to the duramater beneath. This latter structure formed the wall of the sac which protruded from the cranial cavity. The sac contained thin chocolate colored fluid (two ounces) in which was apparently the

cerebrum, the convolutions of which were fairly well marked although flatter than usual. On opening up the cerebrum, the ventricles were seen to be widely dilated by an internal hydrocephalus, the brain-substance remaining about  $\frac{1}{4}$  inch thick around these cavities. Removal of the cerebrum showed the foramen in the occipital bone to be bridged across by another fold of duramater which bulged outwards and which enclosed a mass of nerve tissue about the size of a walnut, quite smooth and without any folds whatever. Section of this showed that it contained a large cavity. This mass appeared to be the cerebellum. Contained within the cranium itself, was a large mass of soft gray nerve tissue which was apparently continuous with the medulla. The defect in the occiput is about 2 cm. below the posterior fontanelle, and is apparently an enlarged foramen magnum. All of the embryonic parts of the occiput can be made out along its boundaries.

The spinal canal is open in its whole extent, except at the twelfth dorsal and first lumbar vertebrae, the arches of which are present and form a bony roof to the greatly enlarged vertebral canal, which is thus closed in just before it widens out to form the floor of the altered cranio-rhachidian cavity. The occiput is adherent to the left side of the twelfth dorsal vertebra. Owing to the flexion of the fetus on its lateral axis to the left and also to its retroflexion upon itself, a peculiar deformity of the thorax is produced. The ribs on

the right side are widely separated from each other, while those on the left are closely jammed together. They are also deflected downwards so that a defect in the bony framework of the anterior wall of the chest results. The defect is much larger on the right side and is bridged across by the first and second ribs, its upper boundary being formed by the clavicle, its lower by the fourth rib. The defect on the left is smaller and is bounded above by the clavicle, below by the upper four ribs which are bound closely together, resembling at first sight one structure. The clavicles are large and strong, having the acromial ends bent sharply down to meet the scapulae and approaching each other closely (within 1 cm.) above the sternum with which they articulate along its upper border.

On the right side, the ribs are as follows: The first arises from the middle of the shaft of the second; it is a small cartilagenous process running forwards and upwards to be inserted in the second segment of the sternum about 2 cm. below its upper margin. The second arises from the second dorsal vertebra; the third as a bony process in common with that of the opposite side in front of the body of the third dorsal vertebra. These second and third ribs unite about the center of their shafts to form a single broad flat bone, from which a single costal cartilage, becoming narrower as it proceeds, passes to be inserted into the sternum just below the attachment of the first. These three ribs form a bridge across the up-

per third of the defect on this side. The fourth rib arises, with that of the opposite side, as



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(Posterior.)

a bony bridge in front and to the right of the body of the fourth dorsal vertebra. It is

narrow and curves sharply downwards and forwards, forming the lower boundary of the defect. The fifth is broad and heavy, as are also the remaining ribs, which except for a sharp curve at their origin, run straight forwards. The intercostal spaces are very broad, and there is a thirteenth (supernumerary) rib which is joined at its tip to the twelfth, which latter is very long and broad.

On the left side, the first and second arise from the corresponding dorsal vertebrae; the third and fourth as a bony bridge in common with those of the opposite side, in front of the bodies of the third and fourth dorsals. All four are bound together, forming the lower boundary of the defect in the left thorax. The lower eight ribs lie close together, are narrow, especially at their origin, and the lower four have grown together, forming a broad, flat, bony process. No supernumerary rib is present on the left side.

28. Lop and Pujol<sup>24</sup> report a case of fetal malformation which is, as far as one can tell from the abstract of their paper which was at my disposal, a case of iniencephaly. The fetus came from the Maternity of Marseilles, from the service of Queirel and Benet. It was a female measuring 23 cm. in length and was delivered in Sept. 1895, in an advanced state of decomposition, the mother being at the fifth month of pregnancy. On each side of the head there was a cutaneous fold, very large, which extended from the vertex to the



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Dissected, showing opening in occipital bone, interior of skull, lateral rotation of skull to the left, and attachment of the left side of the occipital bone to the lateral processes of the cervical and 12th dorsal vertebrae.

shoulders and then became lost in the inferior third of the trunk. This skin had great mobility. While the neck was well formed in front and very distinct in spite of the two folds which masked the lateral parts, in the rear it was entirely wanting, and the occiput was directly continuous with the back. The skin, lax and too abundant, passed from one to the other, forming numerous folds. The head was in extension and it was impossible to bend it without producing at the same time a flexion of the vertebral column at the middle of the back. The two lateral cutaneous folds, which have already been mentioned, were formed solely by the doubled skin of an aponeurotic leaf, a continuation of the anterior and posterior aponeuroses and forming on each side an absolutely empty space. In view of the macerated condition of the subject, it was impossible to dissect in detail the muscles of the posterior region, which seemed to us to be profoundly modified, this being easily explained by the extensive deformity of the skeleton of the neck and the upper part of the back. At the level of the external occipital protuberance, the skull was fused to the vertebral column at the level of the seventh dorsal vertebra. There was no trace of articulation but complete fusion of all parts of the vertebral column situated above the seventh dorsal with the inferior surface of the occiput. On the other side anteriorly and always at the level of the seventh dorsal, the spinal column seems to bi-furcate from below upwards so as to

form an inverted V, each of whose branches send out a rib from its external face. There were thus six pairs of ribs which seemed to originate at the base of the skull. The distance which separated anteriorly the two homologous ribs was the greater as it rose from the first to the sixth ribs. There was no trace of the cervical vertebrae, except some rugosities echeloned along the base of the skull, which seemed to continue the direction of the spinal column. The cavity of the skull was completely closed. The brain, although much softened, seemed well formed. The cavity of the rhachis was also closed. The bulb presented an exaggerated length; the basilar measuring 27 cm., more than half of the length of the interior base of the skull. The channel of the rhachis opened with the skull cavity into the lower part of this channel, at the level of the seventh dorsal. We did not observe any visceral deformity. This malformation must have been the result of coalescence, during the first stages of embryonal life, of a part of the rhachidian and cranial cavities. The channel of the rhachis could not form itself posteriorly in the upper part in consequence of its fusion with the occiput. We thus explain the spreading of the first six dorsal vertebrae and the almost complete disappearance of the cervical. The absence of the neck behind and on the sides accounts for suberabundance of integuments and their laxity in the upper part of the body. This de-

formity, if the infant had arrived at full term, would doubtless have been a cause of dystocia by hindering flexion of the head.

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