

**Iniencephalus.\***

BY

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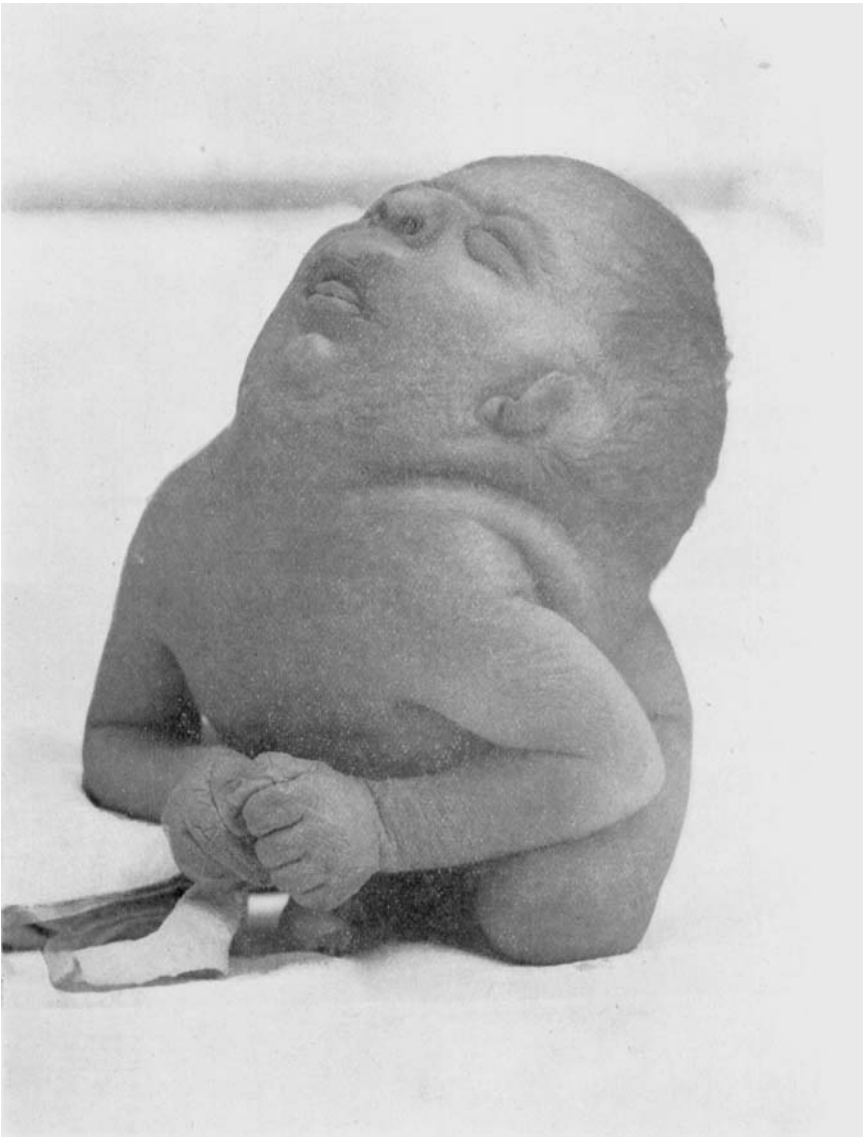
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INIENCEPHALY is very rare. The brain is situated to a large extent within the cranial cavity, but partly outside of it, behind and a little below the cranium, which is open in the occipital portion. Saint Hilaire only knew of one case. H. F. Lewis collected twenty-two cases, which he reported in the *American Journal of Obstetrics* in 1897. To this list, we are able to add a precise description of two others. 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' foetus is preserved.

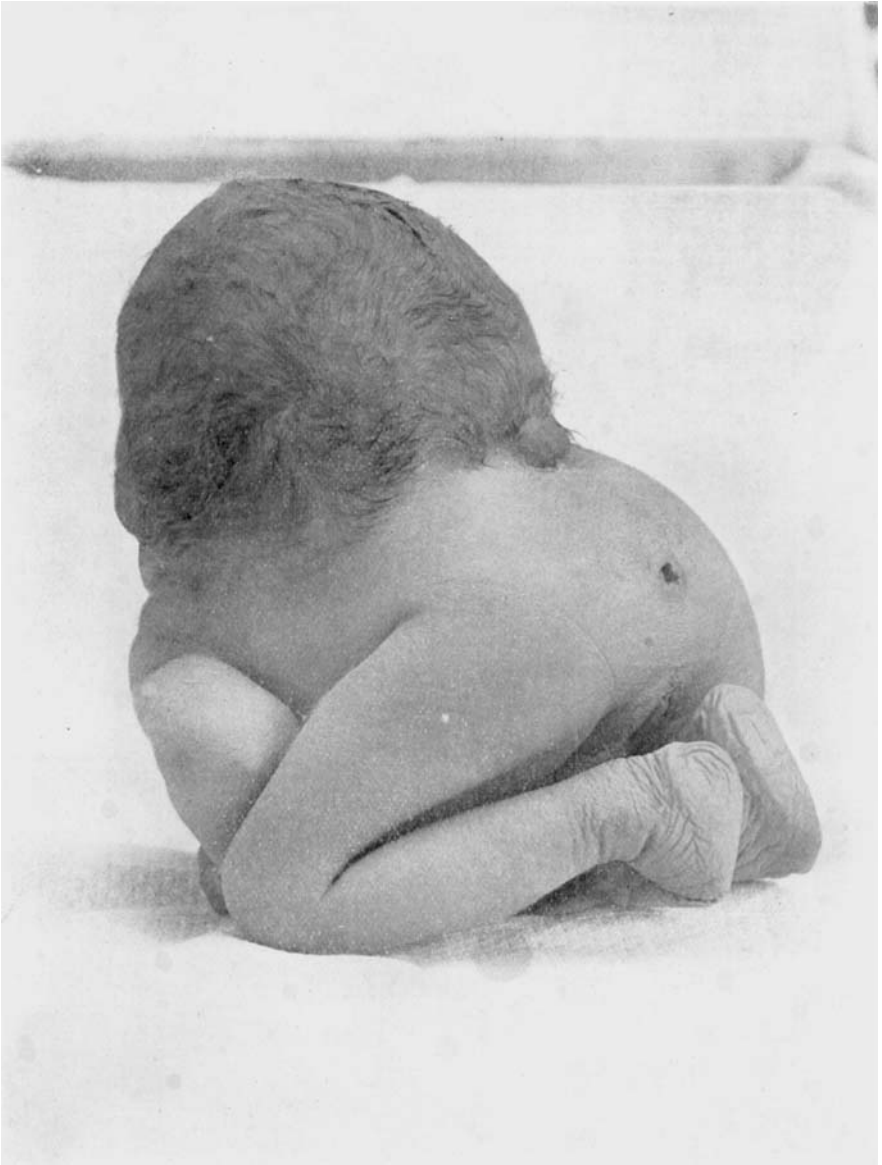
In studying this series of cases, various points in common are to be observed. For instance, the three chief characteristics are occipital defect, spina-bifida, and foetal retroflexion. Out of the 28 cases, the sex was mentioned in 23, and of these no fewer than 19 were females. In six there was a superabundance of liquor amnii, while hydrocephalus was co-existent once. Seven of the foetuses had reached full term, two  $8\frac{1}{2}$  months; two 8 months; one 6 months; one  $5\frac{1}{2}$  months; and one 5 months. The presentation was vertex in two, foot in two, face once and pelvic once. The monster 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 6, 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, these, with the latter, being true iniencephaly. Lastly, those cases where there is a large encephalocele, but where the other characteristics of

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**I.—Dr. Evans' Case. Anterior View.**



II.—Dr. Evans' Case. Posterior View.

iniencephaly are so well marked as to leave no doubt as to their character. This class he calls "*Iniencephalus Apertus*."

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

|   |            |
|---|------------|
| Anus to base of nose .....                      | 14.25 c.m. |
| Occipito-frontal .....                          | 9 c.m.     |
| Occipito-mental .....                           | 9 c.m.     |
| Sub-occipito bregmatic .....                    | 8 c.m.     |
| Bi-parietal .....                               | 9 c.m.     |
| Bi-temporal .....                               | 8.5 c.m.   |
| Fronto-mental .....                             | 6.25 c.m.  |
| Circumference of head at level of O.F. diameter | 28.5 c.m.  |

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 skiagram could be obtained owing to the saturation of the specimen by the mercurial solution in which it had been kept.

2. 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 foetus 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. 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 cm.    |
| Sub-occipito bregmatic ..... | 9 cm.     |
| Bi-temporal .....            | 8 cm.     |

The trunk is very well nourished. In the middle line of the back, about 2 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 short distance 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 skiagram, the basis cranii is seen to be high up and the occiput to be adherent to the bodies of the vertebræ in the lower dorsal region. There is a lordosis of the lumbar and lower dorsal regions.

3. The author's specimen was given him by Dr. A. R. Griffith, Montreal, in whose practice the case occurred. The radiograms were taken by Mr. Watson, radiographer to the Montreal General Hospital. The mother is a healthy ii.-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 large amount of liquor amnii, but the placenta was rather small. The measurements are:—

|                       |          |
|-----------------------|----------|
| Occipito-frontal..... | 9.25 cm. |
| Occipito-mental.....  | 10 cm.   |
| Bi-temporal .....     | 6.5 cm.  |
| Bi-parietal .....     | 8 cm.    |
| Vertex to heel .....  | 29 cm.   |



III.—Authors' Case. Anterior View.



IV.—Authors' Case. Posterior View.

## Head and trunk:

|   |          |
|---|----------|
| Length .....                                | 16 cm.   |
| Circumference around shoulders .....        | 34 cm.   |
| Occiput to anus .....                       | 5 cm.    |
| Cord to base of penis .....                 | 2.50 cm. |
| Shoulder to finger-tip .....                | 16 cm.   |
| Fold of the thigh to knee .....             | 7 cm.    |
| Fold of the thigh to external maleolus..... | 10 cm.   |

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 deep set, 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 1.12 cm. in its broadest part, which is above. This fissure is covered by a thin whitish membrane which extends out further on the right side than on the left. The spina 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 the cervical vertebræ as well as the upper six dorsal, while on the right side the attachment extends down as low as the 10th dorsal.

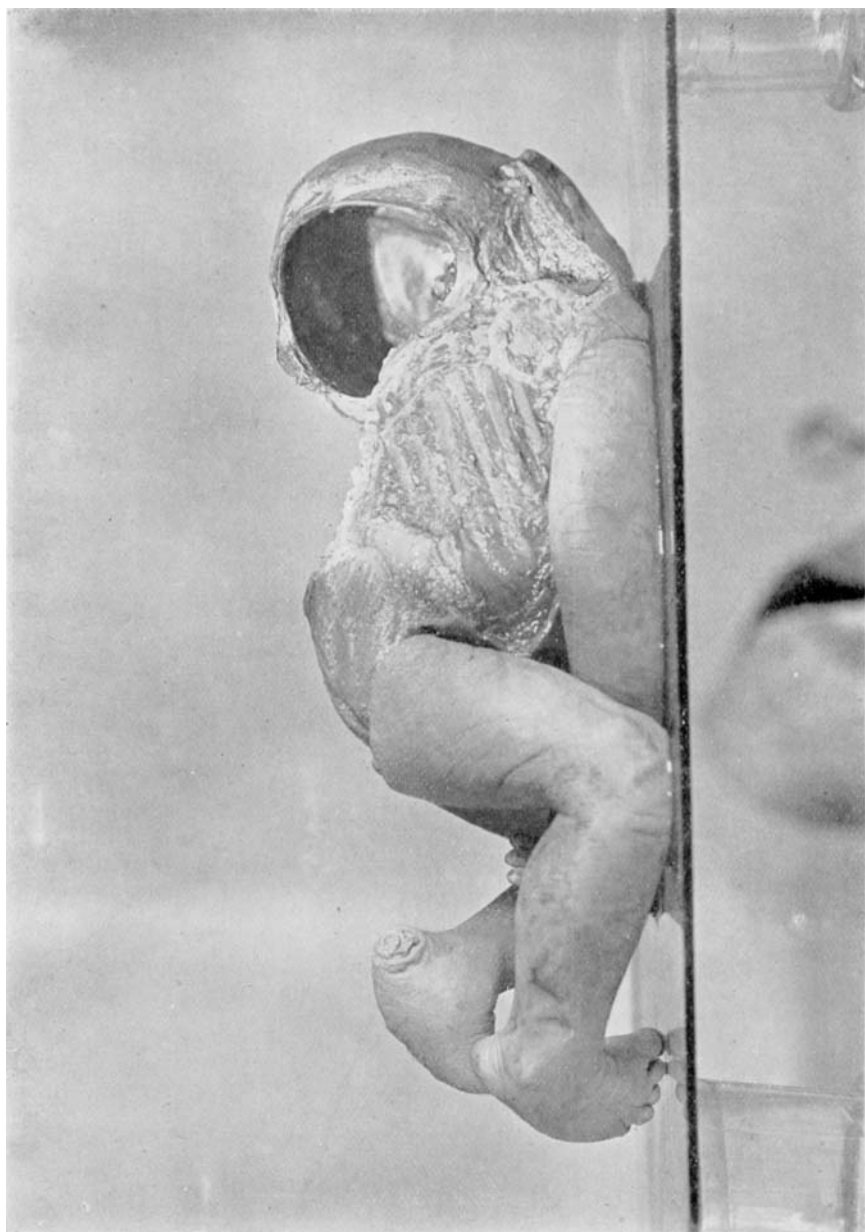
On opening the abdomen, the liver is seen to be larger than normal, and to be displaced downwards. It is attached to the anterior abdominal wall by a thin velamentous membrane, the other hepatic ligaments being normal. The left lobe is decidedly the larger of the two. The heart contained a patent foramen 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 encephalocele. This latter formed a bilobed 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 dura mater beneath. This latter structure formed the wall of the sac, which protruded from the cranial cavity. The sac contained thin chocolate-coloured fluid (2 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  $\frac{1}{4}$  in. thick around these cavities. Removal of the cerebrum showed the foramen in the occipital bone to be bridged across by another fold of dura mater, 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 enclosed 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 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 12th dorsal and 1st lumbar vertebræ, 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 12th dorsal vertebra. Owing to the flexion of the fœtus 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 1st and 2nd ribs, its upper boundary being



V.—Authors' Case. View of opening in occipital bone, showing interior of skull, lateral rotation of skull to the left and attachment of the left side of occipital bone to lateral processes of the cervical and 12th dorsal vertebræ.

formed by the clavicle, its lower by the 4th 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 end bent sharply down to meet the scapulæ, 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 1st arises from the middle of the shaft of the second; it is a small cartilaginous 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 2nd dorsal vertebra; the 3rd, as a bony process in common with that of the opposite side, in front of the body of the 3rd dorsal vertebra. These 2nd and 3rd ribs unite about the centre of their shafts to form a single broad flat bone, from which a single costal cartilage, becoming narrower as it passes to be inserted into the sternum just below the attachment of the 1st. These three ribs form a bridge across the upper third of the defect on this side. The 4th rib arises, with that of the opposite side, as a bony bridge in front and at the right of the body of the 4th dorsal vertebra. It is narrow and curves sharply downwards and forwards, forming the lower boundary of the defect. The 5th is broad and heavy, as are also the remaining ribs, and, except for a sharp curve at their origin, run straight forwards. The intercostal spaces are very broad, and there is a 13th (supernumerary) rib which is joined at its tip to the 12th, which latter is very long and broad.

On the left side, the 1st and 2nd arise from the corresponding dorsal vertebræ; the 3rd and 4th as a bony bridge, in common with those of the opposite side, in front of the bodies of the 3rd and 4th 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.