The Histopathology of a Case of Hydranencephaly in a 30 Week Old Human Fetus

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Introduction

Kuhlenbeck and Globus ('36) described a case of Arhinencephaly with eversion of the forebrain of an infant four days after birth. Kuhlenbeck, Hafkesbring and Ross ('59) made some observations on a living decorticate hydranencephalic child. Their findings indicated a growth disturbance of the forebrain. Kuhlenbeck, Maher, Ross and Eastwood ('57) made some comments, with anatomical and clinical observations on three cases of hydranencephaly. Lindenberg and Swanson ('67) have reported five cases of infantile hydranencephaly in infancy, in which there were noted infarction of both cerebral hemispheres with softening of the cortex and the subcortical white matter of the cerebral hemispheres. There was noted relative gross preservation of the basal ganglia; but the globus pallidus and thalami were histologically damaged. There was noted gross preservation of parts of the lower temporal and occipital lobes and the contents of the posterior fossa.

Weiss, Young and McFarland ('70) described a case of hydranencephaly of post-natal origin. This child was normal at birth but it developed hydranencephaly in a period of 3 months. They suggested that this condition is acquired secondary to intrauterine trauma, infection, or hemorrhage. Appenzeller, Snyder and Kornfeld ('70) described two cases of hydranencephaly in which there was noted autonomic failure.

Halsey, Allen and Chamberlin ('68) described a chronic decerebrate state in infancy in eleven cases of long surviving cases of hydranencephaly. Halsey, Allen and Chamberlin ('70) stated that hydranencephaly may result from any of a variety of destructive or developmental abnormalities, which may act to produce the defect at any time
from the end of the third month of gestation to the second year of postnatal life. All these causes have in common the effect of markedly reducing the total mass of cerebral tissue. They indicated that hydrocephalus has only an accessory causative role. There was no evidence that obstructive hydrocephalus alone could reduce the cerebrum to membrane thickness. In their cases they found relatively normal basal ganglia, brain stem, and tegmentum. However, they noted thalamic atrophy, hypoplasia of the cerebral peduncles and the basis pontis; and absence of the medullary pyramids in nearly all their cases. The cerebellar abnormalities were also not uncommon in their cases. The hypothalamus was fairly well preserved in their long surviving cases of hydranencephaly. They indicated that the hydranencephaly begins in utero from the end of the third month to term, with exception of cases due to postnatal meningitis or trauma, which may occur as late as the second year.

Clinical History:

The mother, a 17 year old white female, was Para 0, blood type 0 Rh positive, STS-negative, and negative for antibodies. The pregnancy was 40 weeks, without significant complications. The membranes ruptured spontaneously with cervical dilatation of 1-2 cm and labor was induced. The delivery was operative and the fetus was delivered stillborn. Five hundred cc of clear cerebrospinal fluid was aspirated from the cranial vault in utero.

Gross Pathological Description

This case was that of a still born fetus of the clinical age of 40 weeks. The anatomical age was approximately 40 weeks. The weight of the fetus was 4050 gm. The crown-rump length was 58 cm. There was noted a severe over-riding of the cranial bones and enlarged fontanelles.

The facial features were unremarkable and the nares were patent. The mouth and teeth were also unremarkable, and the external canals of the ears were patent. The chest and the abdominal configuration was unremarkable. The liver edge was below the costal margin. The tip of the spleen was 2 cm below the left costal margin. The lungs were never expanded with air. There was no precordial fluid, and the heart size was unremarkable. The thymus was large and weighed 17.5 gm. The pulmonary artery was widely patent and the ductus arteriosus was likewise widely patent. The heart weighed 24 gm. The heart chambers were of normal size and configuration. The endocardium was smooth and unremarkable. The heart valves were smooth and glistening. The foramen ovale was probe-patent. The myocardium was
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unremarkable and the coronary arteries arose normally from the right and left cusps of the aortic valve.

The lungs together weighed 50 gm. The trachea and bronchi were patent and were without lesions. The diaphragm showed no hernias. The liver weighed 130 gm, and on section it showed a firm green surface.

The spleen weighed 12 gm. The splenic blood vessels were unremarkable. The esophagus, stomach, duodenum, intestines and mesentery were unremarkable. The pancreas was unremarkable, and on section, it revealed a firm, gray surface. The adrenal glands were small and pale. The right adrenal gland weighed 1.1 gm and the left adrenal gland weighed 0.85 gm. The adrenal cortex was whitish yellow, while the medulla was practically nonexistent. The adrenal glands were very small and the adrenal cortices lacked the fetal zone.

The right kidney weighed 10 gm and the left kidney also weighed 10 gm. On section, the kidneys were grossly unremarkable. The thyroid gland, pharynx, larynx, epiglottis, and the tongue were all unremarkable. The adrenal glands showed a hypoplasia of the adrenal cortex; and there was noted a hypertrophy of the thymus (17.5 gm). There were noted small subgaleal and subdural hematomas. A recent fracture of the right clavicle was also noted. The eyes and the orbits were unremarkable.

Cranial Cavity:

The scalp showed extensive subgaleal hemorrhages in the parietal and the occipital regions. The parietal bones overlapped each other. The sutures were wide and the fontanelles were quite enlarged. There was a small mass of brain tissue in the region of the basal ganglia. About 50 cc of hemorrhagic clot was noted attached to the falx.

Gross Description of the Brain:

The cerebral hemispheres were slightly smaller than the cerebellum, and showed extremely small fully formed cerebral hemispheres. There was observed a well defined cerebellum. There was seen a large subarachnoid hemorrhage connecting the hemorrhage within the hemisphere, within the dorsal surface of both hemispheres. The entire forebrain measured $3 \times 3 \times 4$ cm. The cerebral hemispheres were indistinct and no cerebral gyri were noted. At the base of the brain, two very thin optic nerves were seen. The oculomotor nerve at the midbrain was present. The pons was small and the cerebellum had two large cerebellar hemispheres; but it was slightly malformed. The superior vermis was large and extended one cm above the tentorium.

There was noted a relatively normal spinal cord, pons, and medulla. The medulla showed two very large olivary eminences. The seventh and the eighth cranial nerves were not seen. The spinal cord was
small, and its dura was attached and showed no evidence of defects. The midbrain showed small corticospinal tracts. The hypothalamus, thalamus, and the basal ganglia appeared to be grossly abnormal and rudimentary. There was noted a severe degree of cortical dysgenesis, with the absence of most of the cerebral cortex.

Gross Pathological Diagnosis:

Hydranencephaly with cortical dysgenesis, absence of most of the cerebral cortex, enlarged fontanelles, and severe over-riding of the cranial bones.

Materials and Methods

The brain of this specimen was embedded in celloidin, and it was sectioned serially at a thickness of 20 micra. The sections of the brain were stained with the Nissl method, the hematoxylin and eosin stain, and also with the Loyez modification of the myelin sheath stain.

Microscopic Description

The neocortex was markedly reduced in area. Thus, only part of the inferior temporal lobe was to be noted. The neocortex of the remaining part of the inferior temporal lobe showed a multilaminar six layered type of cortex. However, the cell layers were not as well demarcated, nor as well defined as in the normal state. There could be seen the hippocampus (Ammon's horn), and the dentate fascia. The pyramidal layer of the hippocampus was relatively normal, and the layer of pyramidal cells was relatively normal in appearance. The granule cells of the dentate fascia were normal in size and appearance. However, the dentate fascia was dysplastic and showed evidence of a considerable degree of reduplication and overgrowth. A fairly well defined small putamen which was relatively normal in appearance, was to be noted in the forebrain. However, a caudate nucleus could not be demarcated in this case. A globus pallidus, which appeared to be relatively normal, could be noted.

A large subthalamic nucleus could be seen, which showed evidence of overgrowth and was larger than in the normal state. Just dorsal to this nucleus there could be seen a relatively normal zona incerta. A relatively normal fasciculus H-1 of Forel or the fasciculus thalamicus could be noted. In addition, a lenticular fasciculus (H²) was likewise observed at this level. A small part of the parahippocampal cortex could be seen in the forebrain.

In the epithalamus a rather poorly demarcated habenular nucleus could be observed; but no definite degree of differentiation of the medial
or lateral habenular nuclei could be noted. In the periphery of the
dorsal thalamus a pars dorsalis of the lateral geniculate nucleus could
be seen. It was rather poorly differentiated, and did not show any
evidence of lamination as is present in the normal state. The third
ventricle, particularly its ventral part was dysplastic. In the remainder
of the grisea of the dorsal thalamus no evidence of any specific dorsal
thalamic nuclei could be noted.

In the ventral thalamus a relatively normal zona incerta could be
seen. In the ventral thalamus the subthalamic nucleus of Luys could
also be noted. It was relatively normal in form but was larger than
in the normal state. A nucleus amygdala could be seen, but no evidence
of differentiation of specific amygdalar nuclei was observed. The hypo-
thalamus showed no evidence of differentiation of its specific hypo-
thalamic nuclei.

In the medial part of the rostral level of the hypothalamus, just
lateral to the third ventricle there was noted a profound gliosis, con-
taining areas of ependymal rosettes, abnormal hypertrophic astrocytes
and small nests of macrophages; as well as a number of heterotopic
ependymal lined tubules. This constitutes an area of dysplastic tissue
structures of border line neoplastic character. Similar lesions have
been described by Kuhlenbeck and Haymaker ('46 and '47), and Kuhlen-
beck ('50). Similar dysplastic changes were noted in the lateral part
of the hypothalamus at this level. However, the astrocytic gliosis was
not as profound as in the medial part of the hypothalamus. Only a
few degenerating medium sized neurones were seen in the lateral part
of the hypothalamus. Similar dysplastic changes were seen in the
medial and lateral parts of the hypothalamus at the middle and caudal
levels of the hypothalamus. No evidence of any specific nuclear dif-
ferentiation was observed in the middle or caudal levels of the hypo-
thalamus.

At the pretectal level of the brain stem a relatively normal medial
geniculate nucleus could be noted. The caudal part of the lateral
geniculate nucleus could be seen, but it was poorly differentiated and
was not laminated, as in the normal state. Both red nuclei were present,
and were normally formed. However, they were larger than in the
normal form. A central tegmental tract was present and it was rela-
tively normal in size. It was almost fully medullated. A large sub-
thalamic nucleus of Luys could be demarcated at this level. It showed
evidence of considerable overgrowth. A relatively normal substantia
nigra present, in which a pars compacta and a pars reticulata could
be demarcated.

A thin and very hypoplastic basis pedunculi could be seen at this
level. At the ventrolateral extremity of the brain stem, and just peri-
pheral to the basis pedunculi, a relatively normal and partly medullated
optic tract could be seen. The rostral level of the superior quadri-
geminal colliculi were relatively normally formed. A pretectal nucleus
could not be demarcated in this case. Ventromedially the infundibular
recess could be noted. A pulvinar could not be demarcated in this
case.

At the upper and middle levels of the midbrain there could be noted
forking of the inner wall of the aqueduct of Sylvius. There could also
be seen folds of the inner part of the wall of the aqueduct of Sylvius.
At the middle level of the midbrain, the red nucleus was seen, which
as at the upper midbrain level was larger than normal. The substantia
nigra was of relatively normal size and appearance. The oculomotor
nucleus at this level was likewise relatively normal. The central teg-
mental tract was quite well medullated and was relatively normal. A
capsule of medullated fibers derived from the brachium conjunctivum
was noted at the periphery of the red nucleus. Lying just dorsal to
the substantia nigra, a group of fairly well medullated strio-nigral
fibers were observed. These fibers appeared to be relatively normal in
appearance.

Just lateral to the medial lemniscus there could be seen the medial
geniculate nucleus, which was relatively normal in form and size. Lying
ventral to the substantia nigra there could be noted the basis pedunculi.
As at the upper level of the midbrain the basis pedunculi were thin
and hypoplastic. No evidence of any medullated fibers were seen in
the basis pedunculi. In Nissl stained preparations, a marked gliosis
consisting of various glial elements could be seen in the basis pedunculi.
In Nissl stained preparations a relatively normal substantia nigra,
a pars compacta and a pars reticulata could be seen. However, the
pars reticulata contained fewer cells than in the normal state.

There was noted a stenosis of the aqueduct of Sylvius. At the
upper level of the midbrain, just dorsal to the aqueduct of Sylvius
there was noted a fairly large area of periaqueductal gliosis. However,
the aqueductal ependyma was relatively normal. The changes present
in the area of periaqueductal gliosis consisted of a gliosis of various
neuroglial elements. These changes are commonly seen in cases of aqu-
ductal stenosis. There was no evidence of any inflammatory reaction
in the aqueductal ependyma, or in the periaqueductal region. At the
level of the middle of the midbrain, the stenosis of the aqueduct of
Sylvius was more pronounced, and the periaqueductal lesions were
more striking.

At this level there could be noted within the periaqueductal glial
tissue ependymal cell nests and rosettes of ependymal cells. The peri-
aqueductal gliosis was more noticeable than at the upper level of the
midbrain. At this level there was noted no evidence of any inflam-
matory reaction in the aqueductal lining tissue or in the periaqueductal
lesions. Only a few neurones of the mesencephalic trigeminal nucleus could be seen at this level. A relatively normal oculomotor nucleus and an Edinger-Westphal nucleus could be observed.

At the level of the trochlear nucleus the number of neurones in the mesencephalic trigeminal nucleus was very small. At this level of the midbrain a heavily medullated relatively normal decussation of the brachium conjunctivum, which was larger than normal, was noted. The number of cells in the central gray matter was smaller than in the normal state. A relatively normal supratrochlear nucleus and a relatively normal trochlear nucleus were observed at this level. The periaqueductal lesions and the stenosis of the aqueduct were similar to that seen at the higher levels of the midbrain. In the oculomotor nerve at the base of the middle level of the midbrain there were noted a small number of ectopic primary sensory type of neurones. At the level of the rostral extremity of the pons, in the caudal part of the aqueduct of Sylvius the periaqueductal lesions were not as pronounced as at the rostral levels of the midbrain. At the level of the rostral part of the fourth ventricle, a large nodule of a suprapendymal tumor like area of gliosis was noted. This glial mass contained numerous glial cells, which were closely arranged.

The folia of the cerebellum were relatively normal in appearance and in the content of granule and Purkinje cells, as well as having a normal molecular layer. A well formed relatively normal dentate nucleus was noted. There was also noted a relatively normal nucleus emboliformis and a nucleus globosus. A small relatively normal nucleus fastigii could be demarcated in this case. In the periphery of the cerebellar folia, there was observed the superficial transitory granular cell layer of Obersteiner; which is normally present in the fetal period.

In the dorsal part of the middle level of the pons a relatively normal and heavily medullated brachium conjunctivum was noted. A relatively normal and well medullated medial longitudinal bundle could also be observed. Relatively normal motor and sensory trigeminal nuclei could be seen. A heavily medullated central tegmental tract could be observed. The central tegmental tract was relatively normal. A relatively normal partly medullated medial lemniscus could be demarcated. A trapezoid body, which was relatively normal could also be noted. There could be observed a relatively normal and heavily medullated lateral lemniscus.

A small but relatively normal brachium pontis, which was only partly medullated could be noted. At the middle and lower basilar pontine levels a number of medullated deep transverse pontine fibers were seen. These (fibers) were pontocerebellar fibers. There was no cortico-pontine fiber system to be noted. It evidently did not develop in this case. However, the pontine nuclei did develop in this case. In
the normal new born foetal state no medullated deep transverse fibers are to be noted in the pons. However, in this case a number of medullated deep transverse fibers were noted in the pons. This was an incidental finding in this case. While there exists a considerable variation in the degree of medullation of the fiber tracts in foetuses of similar age; nevertheless the degree of medullation noted in the deep transverse fibers of the pons in this case represents a more advanced degree of medullation of this fiber bundle than that noted in the normal state. In the ventral part of the basilar part of the pons a small number of lightly medullated superficial transverse fibers were seen. In the lateral part of the middle level of the pons, heavily medullated relatively normal trigeminal nerve roots were noted.

The basilar part of the pons appeared to be relatively normal. A small but relatively normal motor facial nucleus was noted in the caudal level of the pons. A relatively normal abducens nucleus was likewise seen in the caudal level of the pons. In the medulla at its middle level relatively normal hypoglossal and dorsal motor vagal nuclei could be seen. A relatively normal nucleus ambiguus could also be noted at this level of the medulla.

Relatively normal dorsal and ventral cochlear nuclei were noted. The inferior olivary nuclei were relatively normal. A medial accessory olivary nucleus could be seen, and a small dorsal accessory olivary nucleus could also be noted. The pyramids of the medulla, at this and the other levels of the medulla were considerably thinner than normal. No evidence of any medullated pyramidal fibers was seen in the pyramids of the medulla. However, some fine medullated transversely running ventral external arcuate fibers could be seen in the pyramids of the medulla. In the pyramids of the medulla there was noted in Nissl stained preparations an advanced degree of gliosis.

A heavily medullated relatively normal restiform body was observed at the middle level of the medulla. Medial to the restiform body a relatively normal nucleus of the descending vestibular tract was seen. A relatively normal fairly well medullated descending vestibular tract was noted. Just medial to this nucleus a relatively normal medial vestibular nucleus was seen. Almost fully medullated relatively normal solitary tracts were present on both sides of the medulla. A fully medullated normal olivocerebellar tract was observed; and a nearly fully medullated ventral spino-cerebellar tract was seen. In the ventro-medial extremity of the medulla a relatively normal well defined arcuate nucleus could be noted.
Discussion

In contrast to the cases of hydranencephaly described by Lindenberg and Swanson (’67), no large areas of softening due to necroses of the cerebral cortex and of the subcortical white matter of the cerebral hemispheres were noted in the present case which was studied. However, as in two of the cases noted by Lindenberg and Swanson, there was seen a relative gross preservation of the cortex of the lower temporal lobes and the contents of the posterior fossa.

In spite of the advanced degree of agenesis of the neocortex, the archicortex or the hippocampal cortex and dentate fascia were relatively normal in this case. Thus, the pyramidal cell layer of the hippocampal cortex was normal in this case. The dentate fascia was relatively well preserved; but showed evidence of a considerable degree of overgrowth. This was similarly noted by Marburg and Warner (’46) in their case of cyclopia. According to Marburg and Warner (’46) this was evidence of the independent development of the archicortex or hippocampus. According to Fischel (’21), every part of the brain is determined in the germ plate, and may develop independently of other parts.

In the cases of Lindenburg and Swanson (’67) the globus pallidus and the thalamus could be delimited macroscopically but were histologically damaged. However, in the present case of hydranencephaly the globus pallidus and putamen were relatively normal. In some of the cases reported by Halsey, Allen and Chamberlin they noted relatively normal caudate nuclei, putamen, and globus pallidus. In the present case of hydranencephaly no caudate nucleus was seen. However, a small but relatively normal putamen and a relatively normal globus pallidus was noted. The dorsal thalamus was dysplastic in this case and showed only a slight degree of dorsal thalamic nuclear differentiation.

As in the case of hydranencephaly reported by Halsey, Allen and Chamberlin (’71), no pulvinar was observed in the present case which was studied. In the epithalamus a poorly differentiated habenular nucleus was noted, but no evidence of medial and lateral habenular nuclei were seen. In the present case of hydranencephaly a small and poorly differentiated pars dorsalis of the lateral geniculate nucleus was seen. However it did not show any evidence of any lamination of the pars dorsalis of the lateral geniculate nucleus, such as is seen in the normal state. No pretectal nuclei were noted in this case.

No other nuclei could be demarcated in the dorsal thalamus of this case. In the ventral thalamus a relatively normal zona incerta could be noted, as well as a very large subthalamic nucleus of Luys. The hypothalamus was completely dysplastic, and showed in its lateral and medial parts a dysplastic state of a borderline neoplastic state. No
specific nuclei could be noted anywhere in the hypothalamus of this case. This resembles the findings of Halsey, Allen and Chamberlin ('71), who noted atrophy of the thalamus and severe disruption of the hypothalamus in some of their cases of hydranencephaly. In one of their cases the hypothalamus was completely destroyed while in other cases the hypothalamus was poorly formed. In contrast to some of the cases of Halsey, Allen and Chamberlin a well differentiated and relatively normal medial geniculate nucleus was seen in the present case of hydranencephaly. In the cases of Halsey, Allen and Chamberlin relatively normal subthalamic nuclei, red nuclei, substantia nigra and brain stem were noted in some of their cases.

The subthalamic nucleus of Luys in the present case of hydranencephaly was considerably larger than in the normal state, and showed evidence of a marked degree of overgrowth. The red nuclei were also relatively normal, and were larger than in the normal state. The brain stem and tegmentum were relatively normal in this case. The oculomotor and trochlear nuclei, as well as the substantia nigra were present and were relatively normal in this case.

In the cases of Halsey, Allen and Chamberlin the cerebellum was abnormal in four of their cases. However, in the present case the cerebellum and its nuclei were relatively normal. In the cases of Halsey, Allen and Chamberlin ('71), the medullary pyramids were completely absent and the pyramidal tract was absent. In the present case the medullary pyramids were markedly hypoplastic. The basis pedunculi were likewise completely hypoplastic, and were considerably smaller than normal. No evidence of any medullated pyramidal fibers was seen in the basis pedunculi. A marked gliosis was noted in the basis pedunculi. In the medullary pyramids some fine medullated fibers were noted, which were probably ventral external arcuate fibers. Thus, the medullary pyramids and the basis pedunculi both showed a marked degree of hypoplasia. This resembles the findings of Halsey, Allen and Chamberlin ('71) in their cases of hydranencephaly.

Lindenberg and Swanson ('67) postulated that hydranencephaly may result secondary to trauma, or infections, namely hemophilus influenzal meningitis and pneumonitis. According to Lindenberg and Swanson ('67), hydranencephaly may result from any of a variety of destructive developmental abnormalities, such as intrauterine trauma or infections, which may produce the defect at any time from the end of the third month of gestation to term; or in rare cases may occur as late as the second year of postnatal life.

Lindenberg and Swanson ('67) support the etiological theory of damage as opposed to a developmental arrest, at some stage of the developing brain. Weiss, Young and McFarland ('70) hold similar views as to the etiology of hydranencephaly; namely that the pathogenesis of
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Hydranencephaly consisted of damage to previously normal cerebral structures. Weiss, Young and McFarland ('70) described a case of hydranencephaly in a child which was normal at birth, but shortly thereafter sustained massive damage to the brain. An intraventricular hemorrhage occurred shortly after birth. This hemorrhage after three months after birth did not progress. They were of the opinion, that this vascular insult initiated the pathological changes in hydranencephaly.

This patient died about 2 years after birth.

Kass reported a case of an infant who died at the age of eight weeks, whose mother had hepatitis during the first trimester of pregnancy. Muir ('59) described two infants, one of which had congenital lues; and the other which had toxoplasmosis in association with hydranencephaly. Johnson ('51) reported a case of hydranencephaly whose mother had received pelvic radiation during the first trimester of pregnancy. Halsey, Allen and Chamberlin ('71) were of the opinion that that hydranencephaly is an end result of diverse morphogenetic entities.

Halsey, Allen and Chamberlin ('71) indicated that hydranencephaly may result from any of a variety of destructive or developmental abnormalities, which may act to produce the defect at any time from the end of the third month of gestation to the second year of postnatal life. They also stated that many cases of hydranencephaly could not be attributed to congenital vascular occlusions. Halsey, Allen and Chamberlin further indicated that the processes leading to hydranencephaly begin in utero, from the end of the third month to term; with the exception of postnatal meningitis and trauma, which may occur as late as the second year.

According to Kuhlenbeck ('73), the condition known as hydranencephaly consists of two main types, namely, type 1, the sort caused by extrinsic destructive encephaloclastic processes; undoing or secondarily modifying the results of normal morphogenesis, (including histogenesis). The second type was caused by intrinsically arrested or defective developmental processes. Kuhlenbeck ('73) indicated that the groups 1 and 2 of hydranencephaly cases might show intermediate or overlapping forms of from 1 and 2 of hydranencephaly. The present case of hydranencephaly which was studied, would probably belong to the group two of this classification of hydranencephaly cases. Thus, this present case of hydranencephaly was probably caused by intrinsic arrested or defective developmental processes. According to Kuhlenbeck ('73) the available evidence remains in favor of histogenetic disturbances independent of exogenic effects; which are factors as yet unknown.

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Summary

1. The neocortex was markedly reduced in area and size.
2. The hippocampal cortex was fairly well preserved and the dentate fascia showed a marked degree of overgrowth in this case.
3. The globus pallidus and the putamen were relatively normal.
4. No caudate nucleus was noted in this case.
5. The thalamus was atrophic and the entire hypothalamus was dysplastic.
6. An absence of the pulvinar and the pretectal nuclei was noted in this case.
7. A poorly differentiated pars dorsalis of the lateral geniculate nucleus was noted.
8. A relatively normal medial geniculate nucleus was noted.
9. The subthalamic nucleus of Luys was present. It was larger than normal, and showed evidence of a marked degree of overgrowth.
10. The red nuclei were relatively normal, but were noticeably larger than in the normal state. The substantia nigra was present and was relatively normal.
11. In the present case of hydranencephaly relatively normal structure could be noted in the basal ganglia, cerebellum, brain stem, tegmentum, and the medulla.
12. There was noted an absence of the pyramidal tract; this fiber tract being completely hypoplastic.

References


**Explanation of Plates**

**Abbreviations**

I. — Molecular layer
II. — External granular layer
III. — Layer of pyramidal cells
IV. — Internal granular layer
V. — Ganglionic deep layer of large pyramidal cells
VI. — Layer of polymorphic cells
Aqu. Sylv. — Aqueduct of Sylvius
Arc. Nu. — Arcuate nucleus
Ar. Sup. Ep. Gl. — Area of supraependymal gliosis
Ar. Trans. P. A. — Area of transition between the putamen and the nucleus amygdala
Bas. Ped. — Basis pedunculi
Brach. Conj. — Brachium conjunctivum
Br. Pont. — Brachium pontis
Br. Stm. — Brain stem
C. Gen. L. D. — Pars dorsalis of lateral geniculate nucleus
Cau. D. Thal. Gr. — Caudal part of dorsal thalamic grisea
Caud. Ep. — Caudal extremity of epithalamus
Cent. Teg. Tr. — Central tegmental tract
Cerebel. — Cerebellum
Ch. Pl. III. — Choroid plexus of third ventricle
Den. Fasc. — Dentate fascia
Dors. Coch. Nu. — Dorsal cochlear nucleus
Dors. Thal. — Dorsal thalamus
Dp. Tr. Fib. Po. — Deep transverse fibers of pons
Dysp. Tiss. — Dysplastic tissue in medial part of rostral level of hypothalamus
For. Br. — Forebrain
Gl. Pal. — Globus pallidus
Hipp. — Hippocampus
Hypop. Bas. Ped. — Hypoplastic basis pedunculi
Inf. Ol. Nu. — Inferior olivary nucleus
Infund. Rec. — Infundibular recess
Plate I

Fig. 1. Ventral surface of brain of this specimen.

Fig. 2. Horizontal, slightly oblique section of the forebrain and diencephalon, 20 micra, Myelin sheath stain.
Plate II

Fig. 3. Horizontal, slightly oblique section of the diencephalon, 20 micra, Hematoxylin and eosin stain.

Fig. 4. Horizontal, slightly oblique section of the diencephalon, at a higher magnification than that shown in figure 3, 20 micra, Hematoxylin and eosin stain.
Plate III

Fig. 5. Transverse section of neocortex of the inferior temporal lobe, 20 micra, Nissl stain.

Fig. 6. Horizontal section of the hippocampal cortex and the dysplastic dentate fascia, 20 micra, Nissl stain.
Plate IV

Fig. 7. Transverse section of the medial part of the hypothalamus, showing its dysplastic character, 20 micra, Hematoxylin and eosin stain.

Fig. 8. Transverse section of the pretectal level of the brain stem, 20 micra, Myelin sheath stain.
Plate V

Fig. 9. Transverse section of the aqueductal and periaqueductal areas of the upper midbrain, 20 micra, Nissl stain.

Fig. 10. Transverse section of the aqueductal and periaqueductal regions of the upper midbrain, 20 micra, Nissl stain.
Plate VI

Fig. 11. Transverse section of the middle of the midbrain, 20 micra, Myelin sheath stain.

Fig. 12. Transverse section, at middle level of midbrain, showing the substantia nigra and the basis pedunculi, at a higher magnification than that shown in figure 10, 20 micra, Nissl stain.
Plate VII

Fig. 13. Transverse section at the rostral level of the fourth ventricle, showing a supraependymal glial module, 20 micra, Nissl stain.

Fig. 14. Transverse section of the dorsal part of the middle level of the pons, 20 micra, Myelin sheath stain.
Plate VIII

Fig. 15. Transverse section of ventral part of the pons, 20 micra, Myelin sheath stain.

Fig. 16. Transverse section of the middle level of the medulla, 20 micra, Myelin sheath stain.