MICROCEPHALUS SECONDARY TO BIRTH TRAUMA*

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(Received for publication May 17, 1944)

THERE HAS BEEN little or no surgical interest in microcephalus since the turn of the century. It seems a bit unusual with the progress and contributions in the field of neurologic surgery that no one has attempted a revival of or any improvements on the surgery of the 1890's in behalf of this hopeless group of children. With the idea of possibly helping these babies by surgery, a review of the English literature was made by the author in 1935. This revealed a goodly number of articles dealing with the effects of irradiation upon the foetus, with resulting microcephalus. Many of these publications were controversial but the weight of evidence was that there were arrested developmental cerebral changes in these infants and as such the group did not invite any surgical interest.

Freeman in 1917 reported in detail the histologic changes in four cases of microcephalus and these were discouraging for any optimum surgical possibilities. He found “a disordered fat metabolism as a distinctive feature” and “the absence of any inflammatory vascular and degenerative changes striking.” Eley in 1933, in a review of neurologic conditions in children, was most discouraging in that “the treatment of this disorder (microcephalus) is ineffective.” He felt there could be little doubt “that trauma to the brain must be related to the production of microcephalus.” In support of this statement he cited the “frequency of a history of intracranial hemorrhage as a result of trauma or secondary to hemorrhagic disease of the new born” in these children. In 1926 Brushfield and Wyatt supplied an excellent statistical study on six per cent of 1545 mentally defective children: twelve per cent of these defectives had cranial circumferences smaller than normal, but they felt that only six per cent were true microcephalics. The measurements as reported by these authors have been followed in diagnosing the microcephalics to be discussed in this communication, although a disproportion of more than 4 centimeters between the greatest circumference of the baby’s head and chest measurements at the nipple line has been a practical criterion for such a diagnosis. These same authors in 1927 gave the autopsy findings in 10 cases, and in every instance “the meninges showed signs of chronic inflammatory changes . . . the dura was thickened and sometimes adherent to the pia arachnoid” microscopically . . . “the cerebral cortex showed a definite paucity of neurones; in the precentral gyrus for instance there were areas almost devoid of healthy pyramidal cells, etc. . . . Some brains under consideration showed definite evidence of a slight degree of atrophic sclerosis affecting both hemispheres.” Recalling the numerous in-

* Presidential Address, 13th Annual Meeting of The Harvey Cushing Society, New York City, May 17, 1944.

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stances in which dural-arachnoidal adherences in cases of tumor have been mistaken as the sole cause of the clinical symptoms of increased intracranial pressure, and the earlier explanations of pachymeningitis hemorrhagica on inflammatory bases, it occurred to mind that the chronic inflammatory changes described by Brushfield and Wyatt might well have been the result of subdural bleeding from birth trauma.

Lannelongue⁶ in 1891, before the 5th French Congress of Surgery, reported his results in 25 cases of craniectomies on microcephalics. He referred to them as obstetrical compressions due to forceps deliveries or difficult labor. He advocated linear or flap craniectomy and “detached the dura mater and allowed it to remain untouched.” In the 25 cases “the majority were improved both intellectually and in their ability to walk.” In this same year Keen⁵ reported 8 cases—3 of his own and 5 others whose records were furnished by his contemporaries. In none of these was the dura opened. One patient followed by Keen for a year “certainly improved steadily and considerably.” Wyeth (1891)¹⁰ noted that intelligence had greatly increased in a month following a bony incision. In 1900 Naylor⁷ sacrificed a large parietal flap without opening the dura and within a week noted improvement in the general spasms and that the spasm of the eyeball was less severe in extent and frequency. After 3 weeks his patient was moving the formerly spastic right arm and leg “quite freely.” The first report in which the dura was opened was by Stiles⁸ in 1901 (linear craniectomy). “The object of this was to prevent the subsequent obliteration of the artificial suture by the formation

Fig. 1 (left). Radiologic depictions of any changes other than premature bony closures excluded certain microcephalics for trephine exploration.

Fig. 2 (right). The rather consistent failure of air to enter the subarachnoid spaces in certain microcephalics might possibly be explained by the presence of the subdural membranes.
of new bone, which we know develops to a much greater extent from the dura than from the pericranium.” Eight weeks after Stiles had operated, the cranium on the side of operation was “perceptibly larger than the opposite side.”

These surgical reports were encouraging. The pathologic changes in Brushfield’s studies were not in every instance beyond the hope that with earlier surgical interferences certain possible secondary changes in the microcephalics might be prevented. The selection of patients in the present series was limited to those whose births were definitely pathologic, for certainly the hereditary group and those of normal birth were likely to have developmental defects. The next consideration was the age for investigation. Of course, the earlier the better if it were secondary damages that were to be obviated, but in the earlier cases, using physical measurements, skull disproportions were too minimal to be accurate about the true existence of microcephalus. In the first six months of life the circumference of the head is increased by three inches and it was usually after this age or later that the abnormally small heads were recognized, so that the earliest investigative ventures were carried out in babies past six months of age.

The presence of definite bilateral optic atrophy and objective evidence of blindness, irrespective of a history of birth injury, was used as a definite contraindication for any investigative or surgical consideration. Plain X-ray films of the skull when they revealed any changes (Fig. 1) other than premature bony closures were utilized in excluding further investigations. In pneumo-encephalographic experiences in an older group of microcephalics prior to 1935 various ventricular enlargements and deformities had been noted—in this same group the failure to visualize the subarachnoid spaces (Fig. 2) occurred in a greater percentage than had been the average in a comparable group of children who were not microcephalics. It is possible that chronic meningeal changes prevented subpial air entrance, and it was the proposal of introducing the air directly into the ventricles by way of the anterior horns that revealed the existing subdural changes. When these pathologic changes were more grossly

Fig. 3. It is most likely that the ventricular enlargements in this case were compensatory, although posterior fossa hematogenous membranes have been revealed in 2 cases with obvious hydrocephalus.
reviewed at a subsequent linear craniotomy, it was obvious that sub-
arachnoid air, had it been disseminated over the hemispheric surfaces, would
have unlikely depicted the presence of a pathologic subdural membrane. In
subsequent cases spinal air introductions have been carried out either pre-
or postoperatively for investigative purposes and not in the hope of depicting
the extra-cortical lesion. It is likely that preoperative encephalographic
studies (Fig. 3), should the ultimate surgical results justify their indications,
form a definite basis for the proper selection of these cases. In the cases to
be discussed in this communication trephine exploration has been the means
of demonstrating or excluding any subdural pathology.

OPERATIONS

There have been 17 patients operated upon. In five cases trephine ex-
posures were made; no extra-cortical pathology was seen and the scalp was
closed. In 12 instances a subdural membrane was exposed and the trephine
exposure was extended to a linear temporo-frontal craniotomy; eight of
these patients have had subsequent procedures on the opposite side of the
skull, so that bony and dural sacrifices, from zygoma to zygoma across the
vertex, have been effected. In no instance where a membrane was uncovered
on one side has there been a failure to expose a similar change over the op-
posite hemisphere. In one of the four cases in which only a unilateral ex-
posure was carried out there was a respiratory fatality six hours after opera-
tion. In another less than a month has lapsed since the initial operation. In
the third there was such obvious cerebral atrophy (external hydrocephalus)
that a second procedure was not considered. In a three-months observation
period the head circumference remained the same and the frequency and
character of the convulsions remained unchanged. In the fourth case there
occurred an increase of 1 centimeter in the head circumference in a four-
months lapse of time from the unilateral operation. The chest circumference
in this period had increased 5 centimeters. The decerebrate attacks had not
been influenced by the surgical efforts and the decompression area was con-
stantly a centimeter below the normal skull contour level.

In the eight cases in which bilateral operations have been carried out the
second procedure was based on several requisites: (1) that at the time of the
primary procedure the exposed hemisphere did not exhibit any grave gross
abnormal changes; (2) that the convulsive seizures had positively been in-
fluenced, with either a decrease in frequency and severity or evidence that
they were firing primarily from the opposite cortex; (3) that signs of de-
cerebration, when present before exploration, had been materially lessened;
and (4) a measurable increase in head circumferences in three cases where
symptomatic evidence of improvement was feebly established.

CLINICAL HISTORIES

The clinical histories of the children who have these hematogenous sub-
dural membranes are all so similar and the seizures which have been per-
sonally observed in almost every instance are so much alike that a clinical picture can be described which, with a few minor individual variations, is typical of all of them. They all have been first-born. The deliveries have been pathologic, with a short or difficult labor, or the use of pituitrin or forceps, and more than half of the infants required artificial respiration. General convulsive seizures occurred within the first two or four days of life. Eight of the 12 had bloody spinal fluid on lumbar punctures. Symptomatic treatment was instigated, and by the time the mother's post-obstetric recovery was assured the baby's symptoms had subsided and the maternal fears of a handicapped child were abated.

At the age of two to three months focalizing irritations began. The patterns of progress in these attacks were not always the same: first one side

was involved and then the other—sometimes the mother's observations were those of clonic seizures of the head and eye—the initial onset would be in one extremity, then another. Although the irritations may have been bilateral, severe generalized convulsions occurred in only two cases. Sedatives played little role in controlling the focal irritative phenomenon, and 20 to 30 attacks a day were recorded in some of the mothers' diaries.

A difficulty in the baby's ability to grasp objects and to hold up its head at three to four months of age was a characteristic paternal observation in almost every instance. In the children over eight months old an early fontanelle closure had been noted by the parents in the majority of cases, their

Fig. 4 (left). This 9-months old baby had all 4 cranial physical characteristics of microcephalics.

Fig. 5 (right). This 4-months old baby girl had only the over-riding anterior sutures and a bifrontal narrowing. Facial features and expressions were good and the posterior contour of the skull could not be classed as abnormal.
observations being that there was no fontanelle at three months of age, or complete disappearance before the age of six months.

The attacks of these children occurred with such frequency that it was possible to observe many of them prior to trephine exploration. There are two characteristics: (1) Jacksonian seizures, mild in character, definite in pattern and short in duration; (2) spontaneous rigidities, either quadra-extensor in character or unilateral extensor and crossed flexor types. While physical measurements and early fontanelle closures have been the criteria for classing these children as microcephalics, not all of them have the birdlike appearances, the flattened occiputs and the two marked angles (1) of the facial center and (2) at the vertex of the head. Most of them have had one and many two of these four physical characteristics of the microcephalic group (Figs. 4 and 5).

**METHOD OF OPERATION**

The surgical procedure used in these cases was a linear craniotomy from the zygoma to the vertex across the longitudinal sinus. A 3-centimeter sacrifice permitted. Care was exercised against opening the arachnoid (Fig. 6).

**PATHOLOGY**

The subdural pathology revealed on these exposures has been uniformly the same and this rather than the surgical technique or results of operation may well be the only justification for this publication. In the initial trephine exposure the dura is likely to be thickened in comparison with the dura in cases of tumor or in traumatic craniotomies in babies. Free cerebral pulsations are not grossly obvious. When the dura is opened one encounters a greenish or purple-tinged membrane, in direct contact with the inner dural surface. Its appearance is very much that of an older chronic subdural hematoma membrane, and its adherence to the dura is identical. Using a small grooved director or a small dissecting blade this membrane can be freed from the dura without any bleeding. On cutting through this friable membrane its depth will approximate that of the dural thickness. Although in three specific instances a thinner transparent pial membrane was demon-
strated as a separate structure, no collection of old free liquefied blood was seen in any of the 20 exposures. As the dura is opened toward the temporal lobe the pathologic lesion is likely to thin out and disappear beneath the temporal bone. On extending the dural exposure to the longitudinal sinus the membrane increases in thickness. One wonders from this constant finding if the origin is from a torn bridging vein that drains into the sinus or from a tear in the sinus with an escape of blood outside the pia arachnoid.

![Image](image-url)

**FIG. 7.** Section from a membrane in a baby 4 months of age. (Hematoxylin-eosin × 6)

The microscopic characteristics of these subdural membranes are similar to those of pachymeningitis hemorrhagica and are shown in detail in the microphotographs (Figs. 7, 8, and 9).

**RESULTS OF OPERATIONS**

The surgical results in three instances have been very striking. The dominating Jacksonian seizures were promptly minimized when the first

![Image](image-url)

**Fig. 8 (left).** Same specimen as Fig. 7. All of the histologic studies have been characteristic of the changes in chronic subdural hematomas. (Hematoxylin-eosin × 75)

**Fig. 9 (right).** This was removed when the baby was 3 months of age. (Hematoxylin-eosin × 75)
operation was performed. The opposite side then dominated until the second procedure was carried out.

Case I (3523). This is the oldest child in the series. At the age of 8 months she was operated upon in 1935 by a right-sided procedure because of dominating left spasticities (10 to 30 attacks per day). There was an increase of 1½ cm. on hemicircumferential skull measurements within 30 days after operation. The mother and attending nurse had observed no left-sided attacks during this period. The left craniotomy was performed 6 weeks after the first one. There were no Jacksonian seizures during the postoperative hospitalization and when she returned for observation 2 months later the mother and nurse reported an absence of any jerking attacks or phenomenon suggesting decerebrate seizures. The head circumference on this date had increased 3½ cm. from the initial measurements, which at that time were 5 cm. below normal.

Seen a year after the second operation there had occurred, according to the mother, an occasional flexor attack involving the right arm, and in walking with support there was a suggestive equinovarus of the right foot. She subsequently learned to walk, and to talk at the age of 3 years.

At 4 years of age her cerebration was quite below normal; she had been trained to good bath-room habits; the equinovarus deformity was much more in evidence in her locomotion; and her mother described attacks of a spastic character involving her entire right side. These were said to be transient in duration and as frequent as 10 per day and as infrequent as once a month.

At 6 years of age she was entered in a school for defectives. The right-sided spasticities continue as transient seizures, but on last report she had had no generalized convulsions and no further Jacksonian attacks.

Case II (3652). The second oldest case in the bilaterally operated group had a similar follow-up course. His original dominating Jacksonian features involved the muscles of the left chest and abdominal wall, at times being so continuous as to result in a body torsion to that side. He was first seen at the age of 8 months, with a 7-cm. disproportion in the size of his head. Following the initial operative procedure the left-sided symptoms were reduced to an occasional twitching with no status seizures.

He was seen at intervals during the successive 6 months. He was able to hold up his head and move it freely within a month after operation. Four months after operation he could sit alone, but when placed on his abdomen he was unable to turn himself over. His attacks were 1 to 2 per day, were momentary, and from the mother’s description were more like decerebrate seizures with the right extensor pattern dominating. The circumference of the head increased 1½ cm. in this 6 months. The left linear craniotomy was done at this time.

A month later, having had no spastic upsets during his immediate postoperative hospital convalescence, his mother wrote that he had on first awakening each morning some attacks that simulated his former decerebrate seizures but that there was no domination of one side over the other. A year from the first operation he had learned to take steps with support, and had learned one or two baby words. He continued on awakening to have rigidity phenomena. He died at the age of 20 months from pneumonia, undoubtedly an act of mercy.

Case III (36211). This child, operated upon in 1936, had a head that was 4½ cm. smaller than normal measurements. He was 7 months of age when first seen. Permission for operation was not given by the parents until the child was readmitted to the hospital at 11 months of age. The head circumference had increased 1 cm. in this interval, while the chest circumference at the nipple line had not changed. He had generalized convulsive seizures beginning with a rotation of the head and eyes to the right side. A right-sided procedure was carried out. There was a moderate external hydrocephalus.

He had one attack during his 10-day postoperative stay in the hospital. For 2½ months his convulsions were at 12 to 14 day intervals and he would have only one attack on these
days. He then began having as many as 6 per day. These continued, and when seen the child
was definitely idiotic in appearance, more spastic than relaxed, and the spasticities were
greater in the right extremities. Eight months after the first surgical procedure the character
of the seizures had changed from generalized convulsions to momentary spastic attacks pre-
cipitated at will with loud noises; the right arm and leg were obviously more involved than
the left extremities. A month later the left craniotomy was done.

The parents reported 2 months later: "His spells don't bother him very much but every
day or so he has a very light one." Fourteen months after the second craniotomy the baby
was still idiotic. He could sit alone, had no words in his vocabulary, and "the spells have left

him entirely but now and then he will get awfully nervous." He died of pneumonia at 6 years
of age, never having learned to walk, feed himself, talk, or cerebrate, but he was free from
generalized convulsions during the last 4 years of his life. Permission for autopsy was impos-
tible to obtain.

Case IV (38150). A 7-months-old baby girl began at 2 months of age to have attacks oc-
curring from 6 to 20 times per day. Many of these attacks were observed. They had their
onset with a twitching of the right facial muscles, then spread to involve the right side of the
tongue, then the shoulder and ended with a spastic extension of the arm. There was a 7-cm.
disproportion in the circumference of the head. A zygoma to zygoma incision was made across
the vault and the bone rongeured away for a width of 2 1/2 cm. The dura and its attached sub-
dural membrane were sacrificed except over the longitudinal sinus.

She was dismissed from the hospital 2 weeks after operation and one month later the

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**Fig. 10 (left).** The left temporal bulge is not to be mistaken as evidence of cerebral expan-
sion. This photograph was taken 6 weeks after the craniotomy, at which time the arachnoid was
accidentally opened, and part of the temporal expansion is the result of fluid escape under the
scalp exposure.

**Fig. 11 (right).** The recent selection of babies has been on the basis of focal irritations, int-
elligent appearances and freedom from any suggestive spasticities.
mother reported 3 to 4 light attacks for that month involving the right eye and mouth, with no spread into the extremity. The child held up its head and turned it decently, had use of her hands and arms in reaching for and holding objects, took certain recognitions of her surroundings, and supported her weight on her lower extremities with assistance. When 5 years of age she could walk unassisted without any frank spasticities, her vocabulary was very limited, her bath-room habits were well established, and transient mild irritative involvement of the right face and eyes occurred on an average of 5 to 6 times a day. She had no generalized convulsive seizures during this time interval.

Cases V, VI, VII and VIII. These children have all been operated upon within the past 12 months. They have been younger and selected with more neurologic care (Fig. 10) than in any of the previous cases. Cases V and VI were 4 months of age. Case VII (Fig. 11) was 5 months old at the time of the initial craniotomy. Case VIII is the youngest and most recently operated upon, having just had the second unilateral procedure. These four patients have not had any generalized convulsions since the initial ones at birth. Focal seizures, even though in some instances initiated in first one part of the body and then another, have all been observed in more than one attack in every case. In these four most recent cases, surgery seems to have had some definite beneficial effects upon the irritative symptoms. The ultimate progress will be routinely followed.

COMMENT

It appears that birth trauma is a definite factor in some cases of microcephaly. The subdural hemorrhagic membrane in these cases is analogous to the membrane of pachymeningitis hemorrhagica of the chronic subdural hematomas in adults. It is possible that this membrane in babies may be a secondary factor in the production of certain cortical histologic changes and might even be a factor in the failure of the cerebrum to expand. Autopsy studies compared with examinations of the cortex removed by biopsies should ultimately shed some light on the first possibility, and further follow-up physical measurements should net some information regarding the second possibility. When one is familiar with the intrinsic cerebral damages resulting from birth injuries one cannot become very enthusiastic in advocating any surgical procedures in these handicapped children, but if something can be done to minimize or prevent secondary damage, one might consider one’s efforts more effective.

REFERENCES