A CASE OF INFANTILE HEMIPLEGIA, IMBECILITY AND EPILEPSY. CRANIOTOMY.
MARKED IMPROVEMENT.¹

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MY object in presenting this case to the Association, aside from any interest that may attach to it as a contribution to cerebral surgery, is with a view to determine the extent of surgical interference necessary in cases of cerebral palsy with sequent symptoms of cortical irritation.

In a case of cerebral cyst, reported by myself to the neurological section of the American Medical Association, at its meeting last June, the lesion had existed from early childhood to adult life without causing further trouble than an atrophied condition of the right arm and hand. The brain in a measure had become tolerant of its existence, other convolutions assuming the function of the region affected.²

At a meeting of the New York Neurological Society in 1891, Dr. M. Allen Starr advanced the suggestion that removal of a section of the cranium over an affected area might relieve symptoms due to pressure.³

Dr. Roswell Park, of Buffalo, has also reported two cases in which simple craniotomy gave great relief.⁴

In the case about to be related it was decided for these reasons to limit the operation primarily to free removal of a portion of the calvaria, in view of the fact

¹ Read before the American Neurological Association, Washington, D. C., May 31, 1894.

² *Buffalo Medical and Surgical Journal*, April, 1894.

³ *JOURNAL OF NERVOUS AND MENTAL DISEASE*, July, 1891.

⁴ *Medical News*, December 3 and 10, 1892.

that it would be quite as easy at a later period to open the dura if need demanded.

December last, Dr. S. L. Elsner, of Rochester, brought me the little patient with the following history :

Simon G., six years of age, a Polish Jew, family history good and social condition satisfactory. His birth was difficult and prolonged, requiring instrumental interference. The application of the forceps produced a marked abrasion and depression on the forehead over the right eye, as well as upon the vertex. The abrasions healed with suppuration in a few weeks, but from the time of birth the mother noticed double ptosis for a period of eight days. Strabismus of the right eye also existed for a short time. During the first few weeks there was also noted an occasional brief tonic spasm of the right arm, while the fist was in a state of constant contracture, and the whole arm was helpless. Shortly the spasm ceased, recurring, however, at the period of primary dentition, although the loss of motion and contracture of the hand persisted. Subsequent to dentition, the tonic spasms of the right arm occurred at irregular intervals and were infrequent. They were more usually induced by fright or excitement, and attended by a marked congestion of the face and head. The hand and arm became spastic, cold and bluish, developing very slowly. The child only began to walk at the age of four, and was always lame on the right side. Even at the time I first saw him he could talk but little. During the early years of life he gave further evidence of weak intelligence, even of imbecility, by his irascibility, destructiveness and irresponsibility to control. Young as he was he made constant use of obscene words, would spit at his attendants and give utterance to a meaningless outcry at any effort to direct him. He was possessed of an inordinate appetite, and in any tantrum could best be appeased by its gratification. In July of last year the occasional tonic spasms began affecting the right leg; in December they became more general, and a month later were epileptiform, being attended with the characteristic cry, dilated pupils, frothing at the mouth and, at times, followed by sound sleep.

At my first examination in December, the general symptoms masked the signs of local irritation as a fog dims the outlines of a familiar landscape. Only after the use of a cerebral sedative, conjoined with the quiet-

ing influence of a hospital ward—its regular routine and regulated diet—could the focal indications be determined.

An examination February 11th, last, developed the following conditions:

The physiognomy was that of a peevish, mischievous semi-imbecile. In standing, the left leg was the main support, and the feet were wide apart. His gait was limping, slightly spastic and rather festinating. Skull measurement gave a circumference in the plane of the external angular process and occipital protuberance of 48.2 cm. The biauricular arc measured 31. cm. The antero-posterior diameter was 16.9 cm., the biparietal 13. cm., giving a resultant cephalic index of 77%. Hence, there was no marked departure from the normal mean dimension. However there was a perceptible flattening of the left parietal, between the bregma and parietal eminence. The teeth were jagged and irregular, and the arch of the palate unusually high.

He used the left hand entirely, the right being clenched, atrophied, cold and bluish. Forcible extension of the fingers of the right hand was painful and difficult.

Careful mensuration of the upper extremities are noted for the sake of comparison:

R. upper arm, acromion to olecranon	-	22.5	cm.
L. " " "	-	21.5	"
R. fore " olecranon to carpus	-	14.4	"
L. " " "	-	16.5	"
R. hand, carpus to end of metacarpus	-	3.8	"
L. " " "	-	4.9	"
Circumference R. upper arm, origin of deltoid		15.8	"
" L. " " "		16.4	"
" R. fore arm	- - -	15.6	"
" L. " "	- - -	16.5	"
" R. carpus	- - -	10.8	"
" L. " "	- - -	12.	"
" R. hand	- - -	12.8	"
" L. " "	- - -	14.5	"

Similar measurements of the legs and feet indicated only a very slight difference, the right leg being larger and longer. There were no definite physiological marks. Pyschically, he was weak minded, had a bad disposition, was destructive, and a terror to the other children in the ward, while to the nurses he was a veritable "pest."

He exercised no control over urination or defecation, rather from a lack of decency than volition. His mother had long ago learned to give an early-morning enema to control his bowel action, while she had become indulgent to mere wetting of the bed and clothing. He was noisy and obstinate, only amenable at meal time.

Sensation was everywhere normal. The superficial reflexes were increased and the knee-jerks equally exaggerated. Vision was good and the eye ground of no moment. The pupils were equal and responsive. Usually there was marked dilatation. There was, however, inability to control the eye muscles, with resultant though not persistent strabismus, a condition of inco-ordination rather than a deficiency of muscle power.

During the period of three weeks for which he was under observation, he had characteristic epileptiform attacks daily; some complete and severe, others less marked. They occurred at night as well as during the day and were general rather than focal in character. They were easily induced by aggravating or teasing him and seemed in a measure dependent upon increased cerebral tension.

The history and examination determined a diagnosis of pressure in the neighborhood of the hand centre of the left motor area, while I was led to believe by the birth history that the trouble was due either to a hemorrhagic plaque or cyst.

It was decided to trephine and remove a sufficient portion of the cranial vault as a tentative operation, providing the conditions present did not demand more radical measures. It was hoped relief from the evident pressure would alleviate the more serious symptoms, so that the greater risk of opening the dura need not be assumed.

The operation was performed March 13th last, by Dr. Mulligan, of the Rochester City Hospital staff, with thorough aseptic technique. A tourniquet about the head satisfactorily controlled bleeding from the scalp incision. A three-fourth inch trephine was applied directly over the hand centre, determined in the usual manner. The skull was very thin and easily penetrated. Upon lifting the button of bone, the dura bulged slightly into the skull opening, but showed no pathological change. Pulsation was present, but slight. By means of a faradic current, the electrodes being one cm. apart, the dura was stimulated with a resultant muscular con-

traction in the right forearm, a very slight current being sufficient. Palpation of the dura indicated the existence of fluid, and the suspicion of the presence of a hemorrhagic cyst was confirmed by use of a small syringe, half a drachm or more of reddish cystic fluid being withdrawn.

The opening in the skull was enlarged in every direction until the limits of the cyst were reached, an opening of $5 \times 6\frac{1}{2}$ cm. being made. The vault of the overlying cranium had become markedly thin, doubtless from the pressure. Not only does the button of bone, presented for inspection, show this, but as the limit of the cystic area was reached the skull walls became much thicker, presenting the usual proportion of diploic substance. The cyst was ovoid in form and was rather in front of and above the middle third of the Rolandic fissure covering the central portion of the ascending frontal region and extending toward the second frontal convolution. It was probably shallow, since it rather interfered with the growth of the corresponding hand and arm than caused entire arrest of the development.

The readiness with which the centre reacted to a mild current with the slight diffusion of current possible, might also be taken as an indication of the superficial character of the cyst. Inasmuch as opening the dura and removing the cyst added so greatly to the risk incurred, it was decided to carry out the plan originally proposed, in order to ascertain to what extent craniotomy would relieve the patient.

The margin of the opening was carefully trimmed, the dura irrigated and the scalp replaced and stitched, a slight oozing through the minute aspirator puncture being disregarded.

The boy reacted well, had no rise in temperature beyond that which at times had been noted prior to the operation. With the exception of a single light convulsion next morning, he had no fit for over a week. On the seventh day there was a sharp rise of temperature. It reached 103° the following afternoon and was accompanied by a hard epileptiform seizure. The dressing was removed and the scalp was found bulging, although union was complete. A probe inserted into the opening of the skull from behind gave vent to a quantity of sanguineous fluid. This was speedily followed by a marked fall in the temperature and an improvement in the general symptoms. To an overtight head bandage, causing

œdema, the whole trouble was due. This, of course, filled up the cranial opening with serum, ultimately resulting in sufficient pressure upon the dura to develop an old-time convulsion.

With the remedying of this, the improvement was rapid and continuous. The mishap was of interest, however, in demonstrating the direct effect of pressure upon the motor area in producing a convulsion. Incidentally the high temperature, 103° , must have been due to the same cause, as throughout the whole period there was no inflammatory action.

With the exception of this single severe seizure, there has been no convulsion since. The boy became quiet, manageable and developed a much better disposition. He began using the right hand, although in an awkward and spastic manner. This was something he had never done before and gave his mother much satisfaction. Control of the eye muscles became much better and his expression greatly improved. The spasticity in gait was lessened and he improved in walking very much. However, there is a decided talipes equinus, with marked contracture of the adductors of the right leg. Later the tendons will be cut thoroughly with a view to still further improvement.

During the remainder of his stay in the hospital he was easily taught to notify a nurse when he wanted to urinate or have a dejection: a very notable improvement over his former indifference.

He would play with blocks quietly by himself, and refrained from teasing the children. At the present time, nearly three months after the operation, he has continued to improve, although more slowly. He has had no convulsion, is far more tractable than formerly and is no longer obscene in speech. He plays quietly about and in various ways shows a brightening intelligence. The scalp over the opening in the skull is depressed and palpation shows in the dura no sign of bulging. For this reason I am, perhaps, the more hopeful of success, since it would seem there was no communication between the cyst and the cavities of the brain.

Curiously, he has lost the rapacity of the appetite formerly marked, and is no longer destructive of his clothing.

I am aware it is rather early to predict permanent improvement, nor do I overlook the possibility of a further operation being required. But bearing in mind the

toleration shown by the brain in similar instances and considering the little risk incurred in simple craniotomy, I believe it is a wise procedure to be content with the lesser operation until the need of a graver one is demonstrated. Should that be required it is an easy matter to reflect the scalp with a view to complete the operation by removing the cyst.

October 1. The patient was seen two days ago and found to be steadily gaining. There has been no convulsions whatever, while the mental improvement, wholly spontaneous, has been very marked.

A Case of Friedreich's Disease.—H. Krause, a medical student, describes a case of this rare disease in a young peasant of twenty years, who presented no family history of nervous diseases. His father is alive and well. One of his brothers died of consumption. His mother was addicted to strong drink from her youth, and of late years has had now and then convulsive attacks, as well as delirium tremens. Her only sister, who died while some thirty years old, suffered from her seventh to eighth year from a similar uncertainty of gait. In the latter part of her life she was nearly helpless, and could only with difficulty speak. His mother's parents were healthy, yet her paternal grandmother, in the last years of her life, was disturbed mentally. His father has been married twice, and the children of both marriages are healthy. The patient was a full-term child; when twenty-one months old he was very sick, but the nature of his sickness was not to be determined. He learned to walk at the usual time, but from its beginning an unsteadiness was noticed, which increased slowly but progressively. During the last two to three years it has gone forward more rapidly, but without any acute febrile aggravation. As to the diagnosis there was no uncertainty, as all the classic symptoms were present if not all distinct: Ataxia of all four extremities, contractures of the feet, disturbances of speech, hystagmus, absence of tendon-reflexes, as well as paresis and disturbances of sensibility, pains and involvement of the sphincters, as usual, were lacking. The characteristic simultaneous involvement of several members of the same family was absent, yet the other symptoms and its beginning at the usual age—the seventh to the eighth year, place it under this heading.—*Hospitals-Tidende*, No. 30, 1893. F.H.P.