Linear craniotomy (miscalled craniectomy) for microcephalus / by W.W. Keen.

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Keen, William W. 1837-1932. Royal College of Surgeons of England

Publication/Creation

Philadelphia: Lea Bros., 1891.

Persistent URL

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FOR



MICROCEPHALUS.

BY

W. W. KEEN, M.D.,





REPRINTED FROM THE
AMERICAN JOURNAL OF THE MEDICAL SCIENCES,
JUNE, 1891.

PHILADELPHIA: LEA BROTHERS & CO., 1891.



Extracted from The American Journal of the Medical Sciences for June, 1891.

LINEAR CRANIOTOMY (MISCALLED CRANIECTOMY) FOR MICROCEPHALUS.¹

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I have entitled this paper "Linear Craniotomy" instead of "Craniectomy," the word proposed by Lannelongue and formerly used by myself. The operation is practically a long incision in the skull, the removal of a part of the bone being merely incidental to its being a hard instead of a soft tissue. The termination "-ectomy" usually and of right signifies the entire removal of the part preceding this termination, e. g., oöphorectomy, omphalectomy, nephrectomy, etc. Removal of the cranium (as craniectomy means) being scarcely intended or done, our nomenclature should fit the facts. Dr. Bauer uses the word craniotomy in reference to his case (see below), but trephining is, I think, the proper one to describe his operation.

In The Medical News of Nov. 29, 1890, I published a case of linear craniotomy for microcephalus. The object of the present paper is to report the later history of this case and two additional cases that I have had, as well as brief notes of one kindly furnished me by Drs. B. Sachs and A. G. Gerster, of New York, and another, similar, furnished me by Dr. J. C. McClintock, Professor of Surgery in the Kansas Medical College, making, with Lannelongue's two cases and Wyeth's case, eight in all that have been done so far; and to refer to two other cases done before my own, for the same disorder and with the same intention, but by methods which I cannot think should be classed as craniotomies.

Case I.—(For earlier history see *The Medical News*, Nov. 29, 1890.) Since the operation this child has certainly improved steadily and considerably, but not at all to the extent of the case reported by Lannelongue. She screams scarcely at all, sleeps better, notices a watch when held before her, observes things about her, and has used a number of words occasionally, but not constantly. She droots but little, and has

¹ Read in the Surgical Section of the American Medical Association, Washington, May, 1891.

In The Medical News for April 25, 1891, is a notice of a recent report by Lannelongue, of twenty-five cases, which I saw too late to incorporate into my paper, hence I can simply call attention to it in this way. There was but one death.

almost lost the restless wringing of the hands which was so marked a

peculiarity before the operation.

February 17, 1891. A precisely similar operation to the first was done upon this patient at the Jefferson Hospital. The improved forceps enabled me to do it in thirty-five minutes instead of an hour and a quarter. She was entirely well in five days. No drainage was used.

March 24. The child is slowly improving. I do not see that her pro-

gress has been any greater since the second operation than before.

Case II.-K. K., girl. Patient of Dr. F. X. Dercum.

Condition, May 23, 1890.—Age, one year (when first brought to Dr. S. Weir Mitchell at the Orthopædic Hospital and Infirmary for Nervous Diseases). Very small. Makes no attempt to walk or sit; moves arms well; feet and hands cold; muscles flabby. Circumference of head 14½ inches; diameter, biparietal, 4 inches; occipito-frontal, 4.5 inches. Kneejerk slight, sensation good, no clonus, no rigidity. Fontanelle closed. Nose bridge very wide. No mental development; hearing and vision good; does not speak at all; disposition good. Passes water and stools without notice. Spine straight. Five teeth. A general restlessness comes on at times; usually excessively cheerful.

Both parents alive and well. Father and mother are first cousins. No venereal history; no consumption in the family history except a sister of the father who contracted consumption. The mother has been married five years and has two children, of which the patient is the younger. Her brother is four years of age, and is strong. He was

breast-fed; is hydrocephalic; circumference of head 53.3 cm.

The patient was born at term. Labor lasted twenty-four hours and was very hard. No instruments were used. Patient was very small in size. No palsy, but very weak. No fontanelle existed at birth and her head has not grown since, though her body has. She was breast-fed for three months, and since then bottle-fed. She had a cold in her head for six months.

Condition, December 1 (nineteen months of age).—Cannot sit alone; constantly in motion. Twelve teeth. Circumference of head 36 cm. (14\frac{1}{4} in.). Biparietal, 10.3 cm. (4\frac{1}{16} in.); biauricular, 10.2 cm.; bitemporal, 8.3 cm. (3\frac{1}{2} in.); occipito-frontal, 11.9 cm. (4\frac{3}{4} in.). Circumference of chest, 38.2 cm. (15\frac{1}{6} in.). Height, 69.8 cm. (27.5 in.). Weight, 12.5 pounds. [I append the following normal measurements at birth from Schroeder's Lehrbuch der Geburtshülfe: Length, 50 cm. Head—Bitemporal, 8 cm.; biparietal, 9.25; occipito-frontal, 11.75 cm.; circumference, 34.5 cm.] Top of head quite prominent. Can hold things in her hand; cannot feed herself. Moderate contraction of flexors of both feet; legs not paralyzed, knee-jerk absent, no ankle clonus. Idiotic expression in repose. Frontal lobe retreating. Two convulsions this summer, two last winter, each time with teeth. Has the expression of a precocious idiot. Attention can be won, but only momentarily. Mind and eye flit from one thing to another quickly.

Operation at the Infirmary, December 3, 1890.—An incision was made one inch to the left of the middle line, parallel to the sagittal suture and six inches in length. A curved incision was then made from the anterior end of this line downward so as to lift a frontal flap, the scar of which would be hidden by the hair. A half-inch button of bone was removed by the trephine, and from this anteriorly and posteriorly a furrow a quarter of an inch wide was cut out of the bone, extending to

within an inch of the supra orbital ridge and an inch above and to the

left of the inion. The length of the furrow was five inches.

The amount of hæmorrhage from the scalp was very slight—much less than I have found it in adults. The bone was very thin, about one to one and a half mm. only, but bled freely. Opposite the parietal boss the dura was very adherent to the bone, but at all other points was separated easily. When the point of a pair of scissors was put under the flap of bone thus loosened and the handle of the scissors let down gently, the simple weight lifted the flap perceptibly. The periosteum corresponding to the bone removed was cut away. The dura had not been opened, and appeared normal.

A few strands of horsehair were placed in the furrow and the wound dressed. The operation lasted half an hour. Temp. at its close 98°.

13th (10th day). The wound healed kindly and quickly and in five days the stitches were out. During the process of healing the child showed unusual fluctuations of temperature, the highest being, however, only 100.8°, for which no apparent cause could be found. After keeping the child a few days longer in the hospital to be sure of a safe recovery, she was sent home to-day. The hospital attendants are decidedly of the opinion that she is quieter and claws at her head much less than she did before the operation, though for myself I can scarcely see much difference.

March 2, 1891. The child has improved very much in general mental condition, but not so rapidly as I could wish. Accordingly, to-day I did a linear craniotomy on the other side of the head in precisely the same manner as the former operation. The operation was wholly completed

in twenty minutes by the new forceps.

24th. The evening of the operation her temperature suddenly rose to 104.6°, falling to normal in four days. The rise in temperature was of course too sudden and too great to be the result of the operation. The cause of it was soon discovered to be a marked intestinal disorder which had begun the day before the operation: this fact was not communicated to me by her mother. No drainage was used, and the wound was entirely well and the stitches out in five days. Her progress is

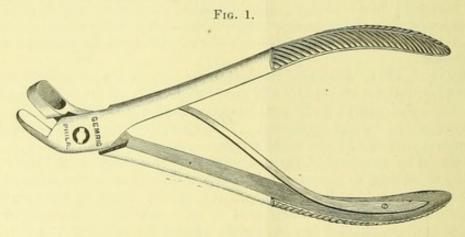
very much the same as in Case I.

Case III.-J. L. H., male, aged sixteen months. Was first seen by me on January 10, 1891. In the fifth month of her pregnancy his mother's friendly sympathies were deeply stirred by seeing the dead baby of a friend, and in the seventh month of her pregnancy she was startled by being nearly run over. Her labor was normal, lasting three hours; no instruments were used. The child's weight was between seven and eight pounds. The anterior fontanelle was very small at birth, and closed at or before seven months of age. The child was breast-fed for three weeks, and afterward bottle-fed. At about three weeks, six weeks, and three months of age he had one or more convulsions. He was very peevish and fretful up to twelve months of age, but of late has been much less so until some recent trouble, presumably his teeth. His parents state that he was a "blue baby" for a year, but Dr. S. Striker, his physician, informs me that it was not discoloration due to a patent foramen ovale, but a constant and very marked mottling of the skin from defective circulation. The child has had attacks of very poor circulation, but of late these have diminished very much. He has also

been subject to attacks of apparent pain and of restlessness continuing

for a day or two.

Status præsens, January 10, 1891.—A well-nourished, apparently hearty boy; no contracture or other deformity excepting his head Viewed in front the skull is markedly conical, face broad, top of skull narrow and arching. Circumference on the shaven scalp, 38 cm. (15 in.). Diameters: Occipito-frontal, 13.5 cm. (5\frac{5}{16} in.); biparietal, 9.8 cm. $(3\frac{7}{8} \text{ in.})$; bitemporal, 8.6 cm. $(3\frac{7}{16} \text{ in.})$. The child droots all the time, has never attempted to stand, falls unless held up, and scarcely even holds his head up. He has never talked. He moves his extremities, but very sluggishly; moans a little, sleeps poorly; head and eyes commonly turned to the left and upward, except in the attacks of apparent pain, when they go to the right, and, as his father expresses it, "he twists himself all up." These attacks are not epileptic in character. He vawns often. His attention can be attracted, but with difficulty, and only for a moment. On shaving the head the broad furrow behind the coronal suture is very marked and the occiput protrudes posteriorly much more than usual.



Operation, January 16th.—Present: Drs. Striker, J. C. DaCosta and Mills. Dr. W. J. Taylor, as usual, assisted me. I thought that the child's circulation had at times been so impaired that especial care was necessary in administering the anæsthetic, so I requested Dr. Coplin to give the ether, on account of his large experience in the Jefferson College Hospital. After the same method as in my former cases I cut a groove three-quarters of an inch to the left of the middle line, reaching from three-quarters of an inch above the supra-orbital ridge well back, nearly to the inion. Its length was six and three eighths inches. In biting the bone I used a pair of forceps devised by me for the purpose, which answered admirably, so that instead of an hour and a quarter, as in my first case, and fifty minutes in my second, I did the operation in thirty minutes. The bone bled quite freely but not alarmingly, and the bleeding stopped spontaneously. No other incident occurred during the operation except that the child's breathing at one time was sighing and he had apparently about the same time a slight convulsion. There was tremor of the extremities during the operation. At no time was he deeply anæsthetized. Shortly after the close of the operation I left the child in its crib in charge of a competent nurse and the mother. He was slightly pallid, but no more so than one would expect after an operation, and there was nothing in the respiration or pulse to cause

anxiety. Dr. Taylor did not leave the house till an hour after the operation, when the child's pulse and respiration were entirely satisfactory. An hour and a quarter after the close of the operation the child gave a few gasps and died instantly, presumably from heartfailure. It had not recovered consciousness after the operation. No post-mortem examination could be obtained in spite of the most earnest efforts.

Case IV. (Gerster and Sachs).—A. F., female, aged four and a half years; labor normal; first child; began to walk after the age of two years; in third year began to speak a few words; but these she would use properly. When thirteen months old had measles. At fifteen months two distinct convulsive attacks without paralysis. Other convulsive attacks at the age of twenty-two months and at twenty-nine months; none since. After these attacks grew more idiotic; lost what little speech she had, became unruly, fretful and ill-tempered; slept little. The fontanelles had disappeared. The measurements taken on shaved head were: Circumference, 31 cm.; fronto-occipital, from root of nose over top of head to occipital prominence, 30 cm.; biauricular, 32½ cm. The child was operated under chloroform, the period of anæsthesia lasting not more than fifty minutes; linear craniotomy was done on the left side, from in front of the position of the coronal suture and beyond the lambdoidal suture. The opening in the skull was semicircular. The child's pulse was weak as soon as chloroform began to act; recovered nicely, however. Soon after operation was conscious, and pulse rallied. Without the further accident the child died suddenly three and a half hours after operation, from acute anæmia.

Case V.—In the New York Med. Record, Feb. 21, 1891, Dr. John A. Wyeth reports a case of a male child, eleven months old, operated on by a median incision of the scalp from the nose to the occiput. Two lateral trenches, one-fourth of an inch wide and three-fourths of an inch apart, were made from just above the eyes to the occipital protuberance. At each end a transverse cut was made in the bone on each side and a similar transverse cut on each side at the middle. By the fingers these four bone flaps were then forcibly torn loose from the dura mater, widening the trenches from one-fourth of an inch to one inch each. No injury seems to have been done to the dura. Time required, an hour and a half. Wound well without incident in ten days. A month later the improvement was "surprising and

gratifying." The intelligence had greatly increased.

Cases VI. and VII.—Two cases reported by Lannelongue in L'Union Médicale, July 8, 1890, and of which a résumé appears in my clinical lecture

in The Medical News of Nov. 29, 1890.

case VIII. (Dr. J. C. McClintock's case.)—A girl, aged three years and eight months, though appearing to be not over one year of age. Premature birth at eight months; no instruments. Anterior fontanelle closed very early. Totally blind; not the faintest signs of intelligence. Two years ago the orbital plate of the frontal bone on the right side gave way, displacing the right eyeball from the orbit. The knee-joint became very painful a few months ago, and the arms and lower limbs contracted. Right tibia and fibula and left femur curved. Head narrow, forehead low. Linear craniotomy (March 28, 1891) on each side of the longitudinal sinus, with lateral grooves at the two ends. The tips of the fingers were then passed into the grooves, and the bones spread on each side until the quarter-inch groove became an inch wide. The dura was not opened. Next morning the temperature was 103° with rapid pulse, but soon subsided to normal. Entire union at the end of a week. The time is too brief as yet to judge of the result, but Dr. McClintock reports that the mother says that "since the operation the child is not so restless, does not cry nearly so much, and has required much less care and attention. The extremities are not so tender, and the child will try to play with any little article that is given to it." Before the operation she could only lie on one side; now she will lie on either side.

The paralysis has almost disappeared, so that the left hand is used almost as readily as the right.

In considering these cases one is struck by the fact that two of them were followed by speedily fatal results. I presume it is only to be expected that children of such feeble cerebral development accompanied as in my own case (Case III.) with faulty circulation, should be less rugged and less able to stand the shock of an operation. The cause of death in my own case was very clearly, I think, heart-failure, and in Sach's and Gerster's case it was acute anæmia. This mortality, which is quite unusual, of course, in ordinary brain operations, should induce us to state the risk to the parents in stronger terms than we would were it an ordinary case of trephining. For myself I cannot but think it a very fortunate thing, for if such children cannot be helped it seems to me that death is preferable rather than to linger in this world in such a defective and helpless condition.

It should, however, make us especially careful as to the administration of the anæsthetic, and we should use all possible means to shorten the operation as much as possible. The forceps I devised for the operation certainly answer the purpose very much better than those I first used, so that the time required for the last operation was only twenty minutes, instead of an hour and a quarter as in my first. In using them it is better to cut alternately a little to the right and left (as one turns his toes out in walking) to prevent their binding. It will be noticed that the upper blade is perforated. This perforation widens from the cutting edge upward, so that each piece of bone bitten out pushes its predecessor loose. In a recent case of spinal laminectomy I used the same forceps to remove the vertebral laminæ, and I found them far superior to any of the other instruments I had on hand, and tried in turn.

I have not yet operated on both sides of the skull, and in view of our present experience I certainly would deprecate a simultaneous operation on both sides. Probably the additional shock would be very unwise and would add greatly to the mortality. Whether two lateral operations done successively will improve the condition of such children more than one remains to be seen.

The results as to mental condition in the six cases that have survived may be stated briefly as follows: Lannelongue's second case was reported almost immediately, too early to judge of any results. So, too, of McClintock's case. In the other four children there can be no question of the improvement, very rapid in Lannelongue's case, slower but sure in both of my own and in Wyeth's case. We have therefore, I think, sufficient reason to encourage us to operate in other cases, and it is this fact and the fatal result in two which has led me to report my cases so early, before the final results are obtained. This will require several years, and meantime we ought to make known the immediate results as a guide in other cases.

In the St. Louis Clinique of Physicians and Surgeons, April and May, 1890, Dr. Louis Bauer reports the case of a young woman on whom he did a craniotomy for microcephalus. Her age, the measurements of the head and the date of the operation are not given. Two buttons were removed from the right parietal bone and the intervening bridge chiselled away. On May 9th, presumably 1890, a second operation was performed on the opposite side of the skull. Prior to operation there was spastic paresis of the muscles with great tremor, which after the first operation had so diminished as to enable the patient to thread a needle. There is no statement as to mental condition. She recovered from the second operation, but it is too early to judge definitely of results.

judge definitely of results.

In The Medical News of Jan. 3, 1891, Dr. Trimble, of Baltimore, reports the case of a child, three years of age, whom he trephined on Nov. 8, 1890. On the right side of the middle line two buttons of bone, one inch in diameter and half an inch apart, were removed, so that the opening measured two and a half by one inch. Some improvement was noted on Dec. 6, 1890.

I have not included these cases under the head of linear craniotomy, since it seems to me that they should be considered simply cases of trephining for microcephalus and idiocy, as has previously been done by Fuller and others. The essential difference between trephining and linear craniotomy is this: that in linear craniotomy the intention is to make the entire side of the head a bony flap, as it were, whereas the trephining in the cases alluded to simply removed two buttons of bone and the intervening bridge and so produced a difference in pressure and possibly allowed, simply at the point of trephining, a little bulging of the brain itself. As to the results, so far as the brief time in all the cases will allow us to judge, they seem to be quite as good after trephining as after linear craniotomy proper, and if so, it may be a matter of indifference which is done. But at the present time it seems to me more logical to do the craniotomy than the simple trephining.

The operation of Wyeth seems to me unwise and likely to be followed by a fatal result in more cases than if the less heroic method usually employed is followed. Even this has resulted in two deaths already, and if a double operation is done and the two sides of the cranium are forcibly separated the danger would seem to me to be far greater. The dura might easily be torn, especially as it is so adherent in children. Moreover, the brain cannot suddenly follow the widening bone, but must be persuaded, as it were, to occupy a larger room by favoring its gradual growth. This, I think, is better obtained by the process I have followed than by Wyeth's method. The same remarks apply to McClintock's case.

