A CASE OF PORENCEPHALON IN WHICH TREPHEINING WAS DONE FOR THE RELIEF OF LOCAL SYMPTOMS; DEATH FROM SCARLET FEVER.

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PRELIMINARY REPORT BY DR. LLOYD.

G. P., aged seven years, was admitted to the the Home for Crippled Children, under the writer's care, in October, 1890. He had bilateral spastic hemiplegia of cerebral origin. His right arm especially was spastic and contractured, and the seat of athetoid movements. The legs also were spastic and the patellar reflexes were exaggerated; ankle clonus was well marked on the left side, but not on the right. The child had but little ability to walk; his legs doubled under him, and he had a tendency to rise on his toes. He had internal strabismus. His mental condition was slightly impaired. His general physical condition was good. The muscular development of the legs was perfect. There was no anaesthesia or atrophy. His head was evidently asymmetrical but there were no scars or depressions upon it. Very little history of the case was known, as, for instance, whether the child had had any trauma or any of the eruptive diseases. Nothing was known of his birth or whether he had had convulsions. About one month after admission, however, he had a series of severe convulsions, after which the athetoid movement in the arm was increased. No signal symptom was noted for lack of opportunity to observe the fits. After each convolution the child's mental faculties were perceptibly weakened. He was dull and listless, with increased athetoid movements in his arms for some hours or days. An examination of the eyes by Dr. de Schweinitz revealed normal pupils with convergent strabismus, the left eye being used preferably for fixation; fundi, with exception of absorption of pigment epithelium of the choroid, showed no abnormalities; high myopia.

The circumference of the child's head was 19½ inches; of the left side 9 3/8 inches; of the right side 11 7/8 inches. The prominent symptoms of the case were thus seen to be a local athetoid movement confined to the right arm, with severe epileptic attacks, increasing for some hours the athetoid condition and impairing the child's mental faculties. After two months' observation, during which time the child's course was progressively downward, a consultation was held with Drs. Willard,
The general opinion of the consultants was in favor of an exploratory trephining. It was thought that there was possibly some atrophy of brain tissue or thickening of membrane, or possibly a porencephalon. The motive for the operation was to explore and then to relieve, if possible, the local condition which was the immediate cause of the epileptic explosions and of the constant athetoid movements of the arm. It was even thought that the case might possibly be relieved, if in no other way, by the linear operation of Lannelongue. In case, however, of the existence of a porencephalon it was the intention to immediately close the wound. On December 12th the operation was performed by Dr. DeForest Willard, consulting surgeon to the institution, assisted by Drs. Keen and Taylor, in the presence of Drs. Mills, Deaver, Dixon, Davis, and Lloyd.

**Surgical Report by Dr. Willard.**

Exploratory trephining was proposed with the hope, not of improving the general mental condition, but for the relief of the athetoid movements and of the epilepsy.

Horsley's measurements were used and the fissure-meter employed to mark off the line of the fissure of Rolando. A delicate gouge drill was used through the scalp to mark the upper and lower ends of the fissure upon the skull before making the incision. A large flap with the base downward was raised, and the periosseum stripped off with an elevator. A one and one-half inch trephine was applied one and one-third inches to the left of the median line. As I have found on previous occasions in children, the skull was exceedingly thin, only one and one-half millimetres, and the centre-pin penetrated the dura before the serrated edge of the instrument had made a groove through even one-third of its circumference. On account of the shape of the skull the trephine was therefore abandoned and rongeur forceps were used to gnaw an opening in the bone.

As soon as the pin penetrated the dura a jet of cerebro-spinal fluid spurted through the puncture at each pulsation. When the opening in the skull was of sufficient size to admit the flat-bladed rongeur forceps, pieces were nipped away and the gap was made one and one-half inches wide. A straight incision was made in the dura, and the cerebro-spinal fluid flowed out to the extent of several ounces. A finger introduced into the opening entered a vast cavity without brain substance in any direction. This cavity will be described later by Dr. Lloyd. On looking into this deep well through the clear fluid a dark opening appeared at the bottom, apparently the ventricle, and it was at once seen that a large portion of the cerebrum was absent.

As no further operative procedures were possible it was decided to close and seal the wound. The wound of the dura was approximated with continuous catgut sutures, but it could not be entirely closed without tearing. The scalp wound was closed with silk sutures without drainage. Iodoform was dusted along the line of the wound and dry sublimate gauze applied with a tight bandage. Thorough antiseptic precautions were taken on the day previous as well as at the time of operation.

Within five hours after the operation the pulse rose to 156, and the temperature to 102°, which could not have been from septic cause. He
had but little pain, but vomited two or three times, evidently from the effects of the ether. He slept at intervals during the night, and on the following day the temperature was 103°, and the pulse from 150 to 170, yet he was perfectly conscious and answered questions with his usual intelligence, differing in no perceptible degree from his previous condition.

His temperature remaining high, it was feared at the end of forty-eight hours that there might be some difficulty with the wound. The dressings were accordingly removed and the large flap was found to be entirely united throughout its extent. There had been absolutely no oozing of cerebro-spinal fluid; there was not even a stain on the dressing. The entire serous stain upon the gauze, over the large wound, would probably not exceed a quarter of a drachm; there was no pus nor redness along the incision nor the slightest sign of irritation of any kind. The wound was absolutely clean.

On the fourth day the temperature rose rapidly to 104.2°, accompanied by an eruption over almost the entire surface of his body. This eruption was slightly elevated, with thickening of the derma. A few hours later there was a profuse crop of sudamina, especially on the inner and anterior portion of the thigh and lower abdomen. These miliary papules soon filled with serum and became turbid; the vesicles ruptured within twenty-four hours; this was followed by rapid separation of the epithelium in patches, which epithelium speedily dried in situ in large flakes. This eruption was accompanied with a moderate degree of itching. The tongue was polished, red, and strawberry in appearance. The eruption was more prominent upon the lower extremities than upon the breast, face, and neck; in fact, there was but little upon the face. On this day, for the first time, the dressings became soiled by oozing. They were accordingly removed, and the wound was found to be healed throughout its entire extent. The clear spinal fluid oozed through the needle punctures of the scalp, the sutures acting as capillary drains, and conducting the fluid through the tissues. The stitches were accordingly removed in the hope that such action would permit the points to heal. The drainage through these points was probably about two ounces. The wound was absolutely free from inflammatory secretions and was in every respect healthy. Antiseptic dressings were applied and firm pressure made.

On the day following the eruption the vesicles had dried. The eruption was very slightly present on the body, but the epithelium was already beginning to desquamate. The temperature ran to 104.4°, it having sunk at one time on the day previous to 99°. The child was perfectly conscious and as intelligent as before the operation. The tongue presented no change, and there was but little redness of the fossae. The pulse was 180. Serum again oozed through the dressings. They were again removed, and it was found that the suture openings had all closed except one in the posterior part of the wound. Intra-cranial pressure had burst through the weakened cicatrix at one point, which had been perfectly healed, but which was now opened to the extent of one eighth of an inch, through which the serum constantly exuded. The wound was perfectly sweet and aseptic. The serum was slightly tinged with blood. Iodoform collodion was freely applied over several layers of gauze in the attempt to arrest this oozing and prevent this continuous drain of spinal cerebral fluid.
On the seventh day the temperature was 101° F.; the pulse 160. There had been no oozing from the wound since the collodion was applied. The child was perfectly rational and suffered less pain than yesterday. The skin felt dry and harsh. The epithelium desquamated at various points, but was still a little red with slightly elevated points. There had been no marked vomiting; no hardness of the glands of the neck nor indurations behind the jaws. The dressing was not disturbed.

On the eighth day the child was desquamating slightly over the entire surface of the body. The temperature fell to 101°. The child cried at times with pain apparently in his ear. He was more bright, more intelligent, talked in a much better tone of voice and much more intelligently than before the operation. The pulse went down to 136; respiration 24. He fretted and worried, but took food well. The urine contained no albumin; it was heavily loaded with urea.

The intracranial pressure was so great that when one opening was closed by collodion, a portion previously healed would be pushed through by the oozing of the cerebro-spinal fluid. No evidences whatever of inflammation were present. The wound was re-sealed and re-dressed. The patient desquamated rapidly and large masses of the epithelium came away. The child steadily failed and lost flesh. Diarrhea set in with involuntary discharges, and he sank steadily until the nineteenth day with all the symptoms seen in severe grades of diphtheria or scarlet fever. The wound remained perfectly healthy. The child evidently died from the poisoning of scarlet fever.

**Clinical Report and Autopsy by Dr. Lloyd.**

The child's first movement on coming out of the ether was with the right or athetoid arm. His condition immediately after the operation was fairly good. He cried loudly. The temperature was good; pulse improving. After the first few hours, however, his condition was never good. His temperature began to rise the same afternoon and reached as high as 102°. His pulse became very rapid; he was restless, slept badly, vomited frequently, and had twitching and jerking movements in the arm. His mental condition, however, was clear and notably better than before the operation. The features of the case, during the remainder of its course, may be briefly summed up in the fact that on the third day after the operation the child was found to have developed a distinct scarlet fever. The surgical condition remained good and continued fairly good to the end. Not a drop of pus or any undue inflammation was observed about the wound. In spite of this favorable surgical condition, however, the child sank lower and lower under the influence of the scarlet-fever poison. A history of this aspect of the case has been published already by the writer and need not be dwelt upon here.¹ The case pursued a somewhat typical course of scarlet fever. The eruption was characteristic, and marked by but one peculiarity—the presence of sudamina. The sudamina were at first lightly sprinkled over the body, but later they appeared in very thick crops on the lower part of the abdomen and inner part of the thighs. The temperature rose to 104°, even 105°; the rash persisted for several days, gradually fading out. It covered almost the entire surface, the

¹ Transactions of the Philadephia County Medical Society, 1891.
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back especially, which was free from sudamina, showing a typical scarlatinuous eruption. The face was much flushed, but around the mouth the white skin was preserved, as is so common in scarlet fever. The throat was flushed but not ulcerated. The cervical glands were little, if at all, involved. The tongue had the characteristic "strawberry" appearance. The patient became slightly more restless and difficult to manage, complaining constantly of pain in the head. The escape of cerebro-spinal fluid from the needle-wound continued very profuse, but otherwise the wound was healing and was thoroughly aseptic. The patient had vomited several days before the eruption appeared. Desquamation began on the fourth day of the eruption at the place where the sudamina had been thickest, and became general and profuse. The child's condition improved, but only to a certain point. The urine was not albuminous. After the eighth day of the eruption, about twelve days after the operation, the patient sank lower and lower. Desquamation proceeded perfectly and the urine was never albuminous, but the patient passed into a dyscrasia, with variable temperature, involuntary stools, diarrhœa, increasing mental torpor, continued head-pain, dread of light, aversion to food, and restless, disturbed sleep. He failed gradually and died eighteen days after the operation.

The autopsy was made a few hours after death. The brain showed not a trace of septic infection. There was no meningitis. The wound was aseptic although it had not healed at all the needle-points. The body presented nothing of special interest, except the brain, which alone will be described. The left hemisphere presents an immense porencephalon. This porencephalon involves the Rolandic region. It extends anteriorly beyond the pre-frontal fissure, downward almost or quite to the operculum; backward to include the superior parietal lobule; in other words its area coincides very closely with the distribution of the middle cerebral artery, the superior convolutions of the temporal lobe alone having escaped. Its edges and sides are formed of the convolutions of the
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brain which have sunk downward and still preserve, especially in the anterior part, their form and relative positions. In the mid-region of the cavity is a large crater-like opening, extending into the lateral ventricle. The membranes at the edge of the cavity are shrivelled up, and form a distinct line or ridge, separating the cavity from the surface of the brain. The gray matter of the mesial aspect of the brain is atrophied and almost destroyed. The gyrus fornicatus is preserved. The left hemisphere is smaller than the right. The measurement from the tip of the frontal to the tip of the occipital lobe shows a difference between the two hemispheres of about three-quarters of an inch. The structures at the base of the brain on the left side are slightly atrophied.

Of the various theories which have been proposed to explain this condition of porencephalon, three only need claim our attention. First of these is the theory of Strumpell of a polioencephalitis analogous to the poliomyelitis of children. The case before us is not explained by this theory, as neither the convolutions nor the membranes covering them present the appearance of having been destroyed or injured by an inflammatory action. Second, the theory of Richter of a basilar kyphosis. According to this theory a defective cartilaginous union, probably of rhachitic origin, causes a twist of the base of the skull, the sella Turcica being thus deflected. The corpus callosum is brought into contact with the falk, which causes atrophy radiating from the corpus callosum into both hemispheres. An obstruction of the veins of Galen is also caused; consequent dilatation of the ventricles and destruction of tissue ensues. This theory may or may not be considered by some to be illustrated in our case. It seems to me, however, that the theory is far-fetched, and that even if it will explain some of these cases, it is not available to explain all of them, especially cases where the porencephalon is unilateral, as in our case. Third: The last, and to my mind the most conclusive theory, is that of vascular disease or injury. Anyone who looks at this brain must be convinced that the left hemisphere has been undermined in the Rolandic region and that its top and sides have caved in. The diseased processes which might produce this condition are several. It might be caused by an obstruction of a large artery of supply. Its location, especially in the area of the middle cerebral artery, lends countenance to this view. Such embolus is not uncommon in several of the infectious diseases of childhood, the most common of which are rheumatism and scarlet fever and the other eruptive diseases. Pneumonia, which is probably infectious in some cases, has been known to be followed by grave cerebral disorders. The protracted summer diarrheas of children have also presented occasionally these sequelae. Venous thrombosis has also been supposed, with reason, to give origin to these serious degenerative and destructive lesions. Finally cerebral hemorrhages have played a very important part in the production of these cerebral palsies of children. The asphyxia of the
newborn babe is a potent and frequent cause. This asphyxia, which too often has been ascribed erroneously to pressure upon the head during labor, or to the action of the obstetric forceps, is probably more frequently the result of compression of the placenta in protracted labor, interfering with the proper aeration of the child's blood. In this condition ecchymoses have been observed, not only beneath the membranes of the brain, but also in the pleural cavity and in the capsule of the liver. This compression of the placenta in protracted labor has not been recognized sufficiently as the cause of serious or even fatal injuries to the brain of the newborn. In its practical bearings, which cannot be discussed here, it suggests timely interference, even with instrumental aid, as conservative, rather than dangerous, to the child's brain.
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