Severe burn of top of head at seven months of age, followed by necrosis of entire osseous cap of cranium : at fourteen years of age detachment of the entire calvarium by circular craniotomy for epilepsy and defective mental development / by William Williams Keen.

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TABLE OF CONTENTS

ORIGINAL MEMOIRS.

TRANSACTIONS OF THE NEW YORK SURGICAL SOCIETY.

TRANSACTIONS OF THE CHICAGO SURGICAL SOCIETY.

REVIEWS OF BOOKS.

CORRESPONDENCE.

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* Read before the Philadelphia Academy of Surgery, February 4, 1907.

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HARRY H. W., æt. fourteen, was admitted to the Jefferson Medical College Hospital, December 7, 1904, at the request of Dr. W. F. Haines of Seaford, Del., with the following history: At seven months of age his parents left him wrapped up in a shawl in a rocking chair in front of a wood fire, which then consisted chiefly of coals, while they went to attend to some farm work. They also left an older child, about two years of age, to take care of him. They were absent from the house for about forty-five minutes. Upon their return they found that the baby in the rocking chair had begun to cry and the two-year-old child had tried to climb into the rocking chair to comfort him. In doing so the chair was overturned forward and the baby thrown into the fire, so that the top of the head was in contact with the live coals. As nearly as can be ascertained by cross-questioning the two-year-old child, and knowing the length of their own absence, the baby's head lay in the coals not less than twenty and it may have been thirty minutes. As a result of this severe burn, the scalp being thoroughly charred, the whole top of the head sloughed off about six months later, including a large portion of both frontal bones, the two parietal bones in their entirety, and a part of the squamous portion of the right temporal bone. The piece of the squamous bone was lost, but a photograph (Fig. 1) shows the other four pieces of bone their natural size. The four pieces of bone which have been preserved can easily be identified. They are of a dark brown color, the

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result both of the burn and suppuration. Placing them in position, they measure from front to back 17 cm., and from side to side 11 cm. When the bone sloughed away the dura was exposed, covered by that time with granulations. A year after the burn, the scalp was healed, and upon my recommendations (for Dr. Haines showed me the specimens and consulted me at that time) a tin cap covered with silk was made for the purpose of protecting the top of the head from blows, but it could not be used as it annoyed the child. Six months after cicatrix was complete, the scar broke down, and from that time till the present it has been alternately healed and open.

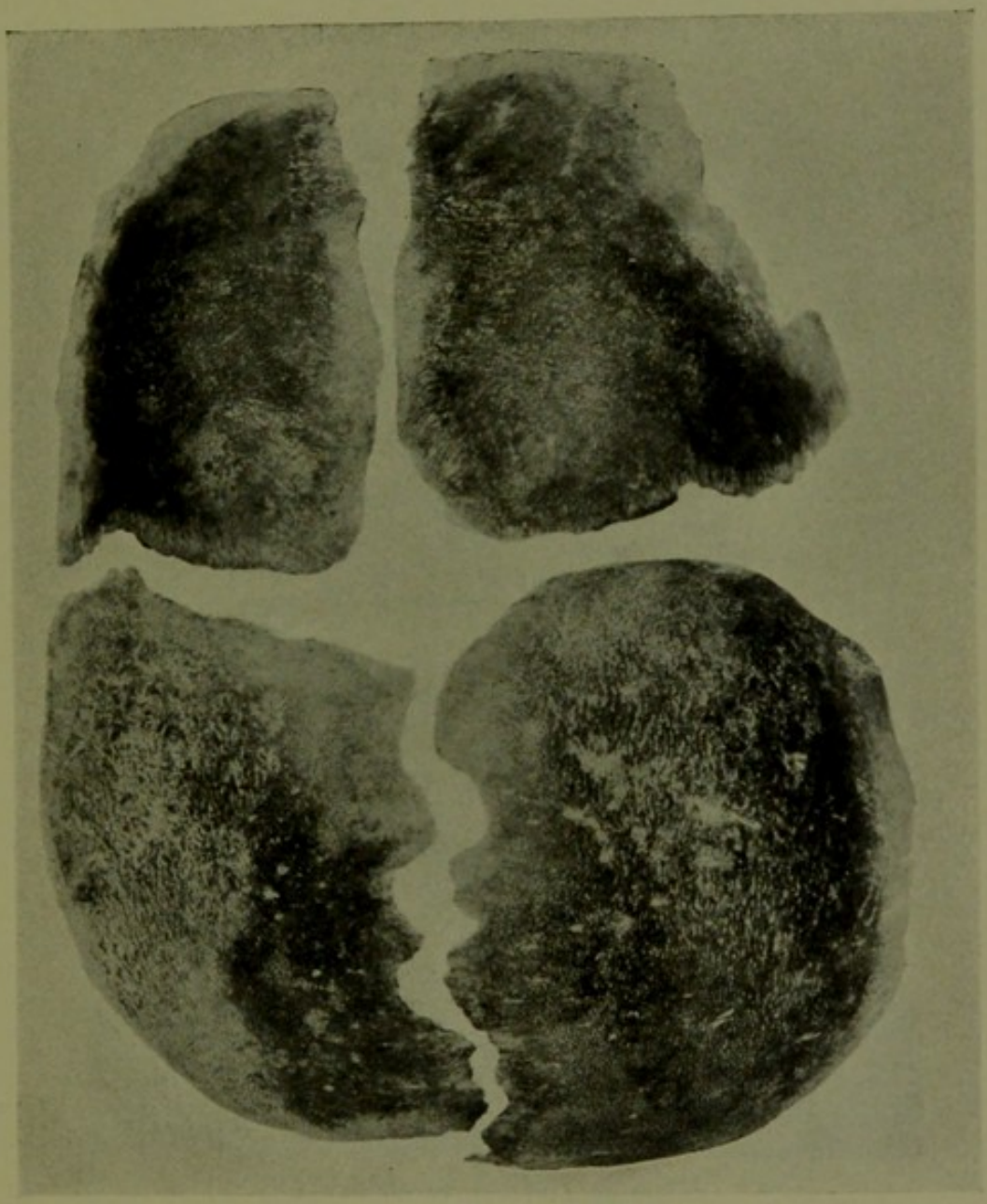
Soon after the accident he had nine convulsions. He was then free from them for over a year. Then he began to have distinct epileptic attacks. These have continued ever since and have increased in severity and frequency. They occur day and night regardless of any known influence, such as excitement, the direct sun's rays, etc. On an average, his father thinks he has about 400 attacks every year. Sometimes he goes several days without a spasm.

He began to go to school at seven years of age and appeared to learn rapidly. His memory was excellent till he was about eleven years old, when his epileptic attacks became more frequent and he became stupid. He was, therefore, removed from school, and he has forgotten most of what he learned and is becoming more and more deficient mentally. While at school he learned to read and write, but in the last three years he has lost the ability to do either.

Physical Examination on Admission.—He seems to be physically a well-developed boy of average height and weight, but his face presents a dull and stupid appearance. He responds rather indifferently to questions and talks, but can hardly be said to converse.

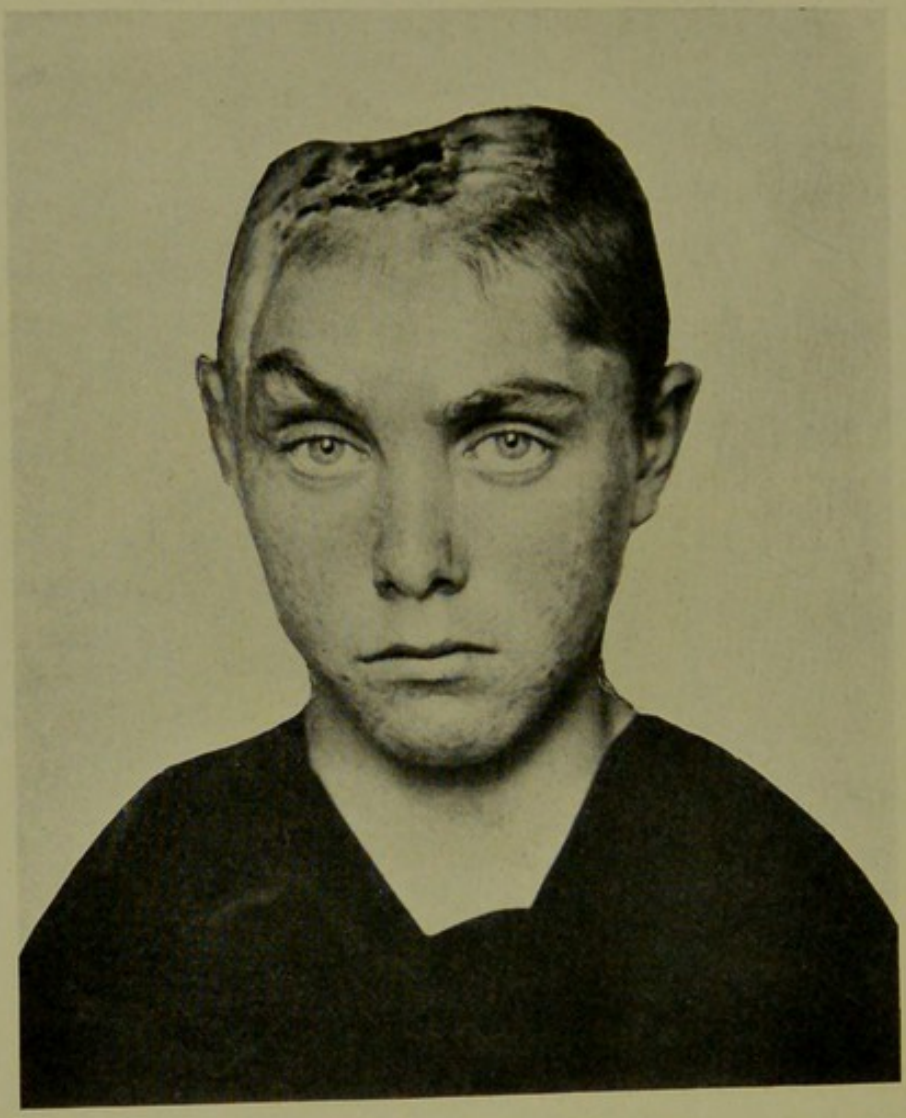
His heart, lungs, and abdominal viscera are, apparently, normal. The deformity of his skull is very marked (Fig. 2), showing a deep furrow a little to the right of the middle line, running obliquely from behind forward and to the right. On the top of the head there is a very large scar (Fig. 3). The oval line in this photograph is an ink line showing the present area under which there is no bone. This measures only 8 by 5 cm. Corresponding to this oval line the margin of the bones can be felt quite distinctly; under the scar, pulsation of the brain can be

FIG. 1.



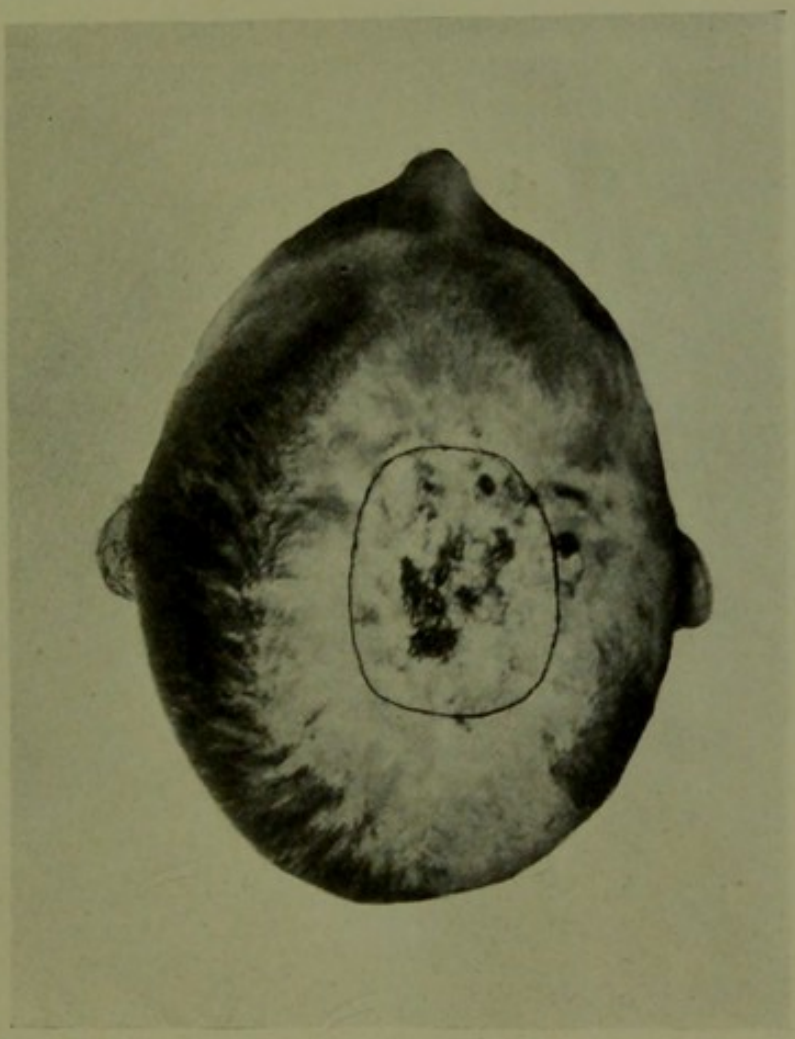
Photograph of the necrosed frontal and parietal bones, natural size, and measuring when approximated, 17 x 11 cm.

FIG. 2.



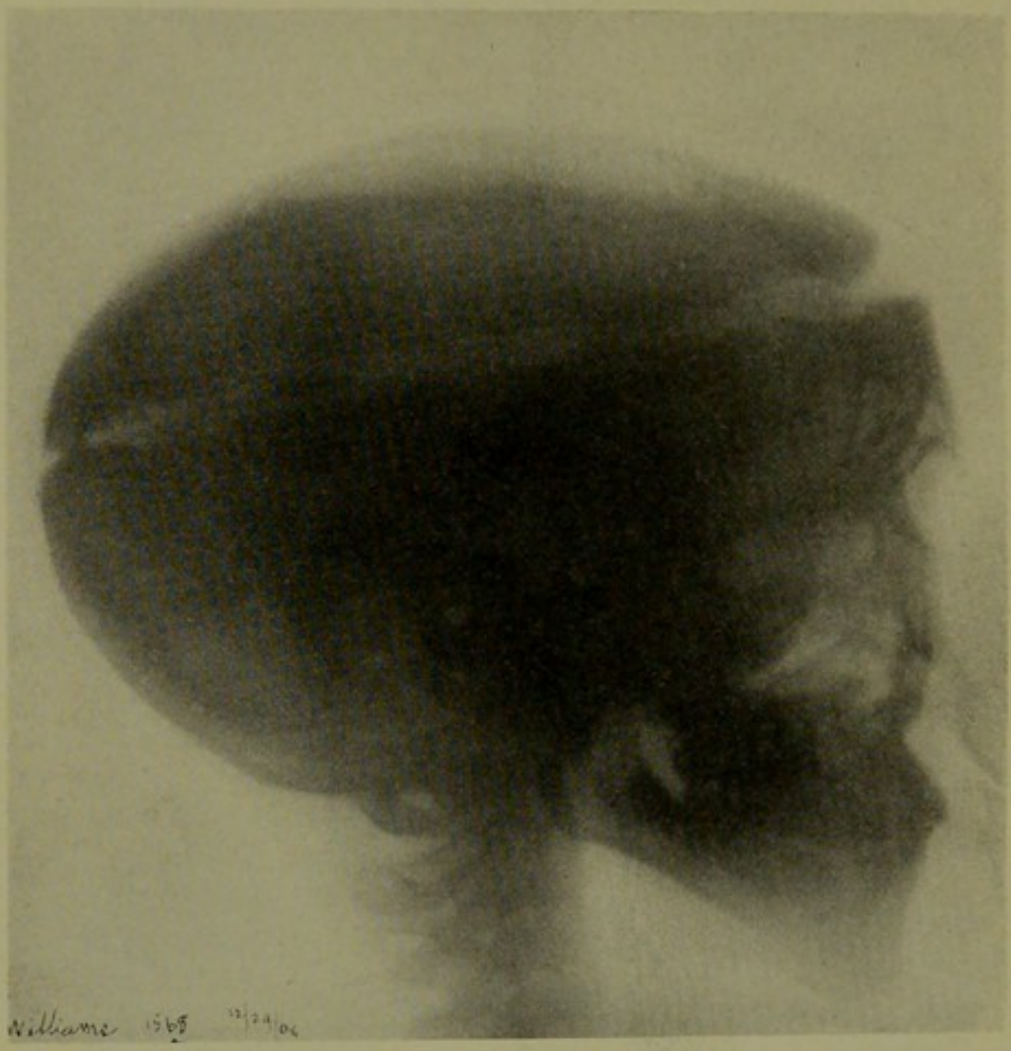
Photograph of the boy at 14 years of age.

FIG. 3.



Photograph of top of head. The dark line corresponds to the present opening in the bone and measures 8 x 5 cm. The original opening when the bones sloughed away at 13 months of age measured 17 x 11 cm. (see Fig. 1). While his head has increased in size with his growth, the defect in the skull has contracted 9 cm. antero-posteriorly, and 6 cm. transversely.

FIG. 4.



Skiagraph (Jan. 10, 1905) showing the gap left in the bone by the complete circular subcutaneous craniotomy. Note also the evidence of loss of bone on top of the skull.

seen; pressure on the area where there is no bone causes pain. There is also a scab at two or three ulcerated points. The scalp is as tense as a drum head over the entire top of the head.

His convulsions as observed in the hospital were at times chiefly manifested in the left leg and arm, at other times in all four extremities. There was no localization of the convulsions.

Urine: turbid, straw-colored, 1017, reaction acid, no albumin or sugar was found, urea 1.6 per cent.; no crystals, but amorphous urates, squamous epithelium, and a few leucocytes; no blood or pus. Dr. Wm. M. Sweet examined his eyes and reported as follows: Normal pupils; normal ocular movements. Optic nerves good color, vertically oval. Arteries and veins normal; smaller twigs tortuous. The arteries in the right eye-ground are a trifle small in proportion to the veins.

Dr. Bochroch examined him from the neurological standpoint and reported as follows. Knee jerks are equal; no asteriognosis; no Babinski; no ankle clonus; no impairment of sensation below the knees and no impairment of the muscle sense. No trophic ulcers; he stands equally well on both legs. There are ecchymotic spots on the arms, impeded circulation, cold sweaty hands; the radial arteries suggest hardening. The left hand, which was also burnt, is smaller than the right. The grasp is equally good in both. No atrophy of shoulder girdle muscles. No thermal anæsthesia. Pupils respond to light and accommodation. High arched palate; fairly good dentition. Hears the ticking of a quiet watch at about ten inches. Tendency to nystagmus laterally with the pupils turned to the right. No impairment of the sensation of taste.

After considering the possibility of doing any operation on the top of the head, I decided that that held out little hope of relief from the pressure, and as the covering of the top of the head consisted of the dura and scar tissue intimately adherent together, it would be very dangerous and probably fatal to attempt any operation there. Moreover, I supposed that probably the superior longitudinal sinus might be blocked as a result of the burn.* I decided, therefore, to do a complete linear craniotomy, so as to separate the entire top of the skull from the lower portion. To do this by an open incision of the entire scalp would almost certainly produce gangrene of the scar tissue of the top of the

* The operation showed that this was not the case.

head. I therefore decided to make several incisions, say 4 to 5 cm. above the ears, and then by my craniotomy forceps to gnaw away a portion of bone about 7 mm. in width. I found that the scalp moved loosely over the skull at about the level indicated all around the skull, excepting at a small area over the right temple. I could, therefore, by undermining it, detach the scalp from the skull through the small openings and then, having made a small trephine opening in the bone, could detach the dura from the bone and do the linear craniotomy.

Operation, December 14, 1904.—I carried out my plan as above described, making the first incision a little back of and above the left ear. I got along without trouble (excepting that it was tedious on account of having to do a large part of the operation without the aid of sight) till I reached the middle line of the forehead. Here, unfortunately, the superior longitudinal sinus was caught in the bite of my rongeur and torn. I immediately checked the quite violent hæmorrhage by some iodoform gauze, extended the incision somewhat across the forehead to the left, rapidly made a trephine opening at this point and gnawed away the bone till I reached the point of the tear. I was able then by my finger to check the flow of blood sufficiently to see the bone well, and complete the craniotomy in the middle line. I packed some iodoform gauze into the opening, which effectually checked the hæmorrhage, and then discontinued the operation, having completed nearly one-half of it, and determined to do the other half a few days later.

In thinking over the matter I feel quite clear that the tear of the sinus was due to the fact that I did not adopt the proper method in approaching this portion of the bone. I should have continued the gnawing away of the bone till I reached almost to the middle line, then have made a trephine opening on the left side and gnawed away the bone on that side nearly to the median line, have exposed the sinus, and then by guarding it with my forefinger or some other suitable shield, such as the handle of a knife, I am quite sure I could have removed this piece of bone which projected inward more deeply than usual, at least 4 or 5 mm., with safety and I would not have torn the sinus.

December 20, 1904.—He has done so well that I completed the craniotomy to-day. Warned by my former experience, I attacked the superior longitudinal sinus posteriorly first from one side and then from the other, as just described, gnawing away the

bone over the sinus itself last and without any trouble. The hæmorrhage was not at all severe. Eight incisions were made in performing the complete craniotomy.

Two days after the first operation, and one day after the second, he was sitting up in bed with a backrest. In the interval between the two operations he had two convulsions, December 18 and 19, with but very little twitching. Before the second operation was done it was clearly noted by the resident, his father and several surgeons who had seen him repeatedly, that his mental condition seemed to be distinctly improved even by the first operation. I hardly think that the wish was father to the thought, but, of course, it is difficult to express an unprejudiced judgment. The boy himself said that his head felt much better than before the operation. Very little pain followed either operation. His temperature after each operation only once exceeded 100 degrees.

January 9, 1905, a skiagraph was taken (Fig. 4.) This shows well the absence of bone on top of the head and also the line of my linear craniotomy.

On January 10, 1905, just before his discharge, Dr. Bochroch again examined him and made the following report: The patient's face expresses apprehension and lack of intelligence. A considerable interval elapses between his answers to such questions as "Where do you live?" "How many brothers and sisters have you?" etc. There is apparently no paralysis of the muscles of the face; he is, however, unable to draw his cheek from either side in order to show his teeth. Most likely this is due to lack of understanding of what he is expected to do. The eye-balls have a tendency to twitching, or a slight jerky movement; possibly more marked in the right than the left eye. When following an object, especially toward the right side, lateral nystagmus is distinct. The pupils are somewhat dilated, but respond promptly to both light and accommodation. There is a fine tremor of the tongue; also a fine tremor of the hands, more marked in the right than in the left. Grasp good and equal. His walk suggests the "steppage gait;" this is exaggerated when walking with his eyes closed. During this test he always walks to the right. He has no Rombergism, but he stands with difficulty on either leg, with his eyes closed. The knee jerk on the right side is exaggerated, on the left side rather minus. No Babinski or ankle clonus. The reflexes in the upper extremity,

wrist, biceps and scapulo-humeral, are exaggerated. Tactile and thermal sense normal, though he occasionally gives evidence of paræsthesia. No asteriognosis.

He left the hospital on January 7 to visit an uncle in the neighborhood, but returned to the hospital on the tenth and then went home. His peculiar gait mentioned in Dr. Bochroch's last examination was improved, and his general and mental condition also were improved.

After the second operation, his convulsions were as follows: December 23, 5 minutes; December 24, 5 minutes; December 28, 7 minutes. They were chiefly on the right side and the mouth was drawn to the right. December 30, two attacks, 6 and 3 minutes long respectively, similar in type to the one on the twenty-eighth. December 31, one attack, duration 5 minutes. There were no movements on this occasion on the right side, but only a clonic spasm of the left arm and leg, and the face was strongly drawn to the left.

October 26, 1906.—He was shown to the Society of Clinical Surgery in a clinic which I held at the Jefferson Medical College Hospital. His father states that he has had fewer attacks and that his intelligence is slowly improving. The ulcers on the top of his head are rather worse than when I last saw him two years before and cover the central half of space where there is no bone.

A new skiagraph taken at this time shows persistence of the gap seen in the first skiagraph, but the edges of the gap are, of course, rounded off and less sharply defined. The width of the gap in the bone is the same as immediately after the operation. Dr. Haines writes me, January 25, 1907, that the top of the skull does not seem to him to be movable.

REMARKS.

That the baby did not die from the accident is extraordinary, but it is not a cause of astonishment that he should develop an abnormal shape of his head or an abnormal mental condition accompanied with epilepsy.

That popular myth, "pressure on the brain," is certainly realized in this case, as shown by the deep furrow on top of his head and by the measured contraction of the original defect in the skull. His head, though of very abnormal shape, is of the average size for a boy of fourteen. Hence the head has

enlarged very much since the bones exfoliated thirteen years before. But instead of the opening left by this exfoliation enlarging pari passu with the growing head, it has greatly contracted. Adjusting the necrosed bones accurately together and exclusive of the lost piece, the aperture left by their exfoliation must have been 17 by 11 cm. At fourteen years of age this opening had contracted to 8 by 5 cm. Not only had contraction taken place in the horizontal plane, but the deep furrow on top of the head shows that a marked contraction had taken place in the vertical plane.

That the epilepsy and mental dulness have been caused by the contraction and consequent pressure, and by the physical alteration in the structure of the cortex itself by the burn, I think there can be no doubt. The only wonder is that he is not wholly idiotic as well as epileptic.

While I had little hope of benefitting the boy by any operation, it seemed to me he ought at least to have the possible chance of benefit from the relief of pressure, provided such an operation would not be almost certainly fatal. As described in the notes, my idea was to make the entire calvaria movable so that it could be lifted like a lid on top of the head. If, then, the brain had any power of expansion it might lift the calvaria and so get more room.

The apparent immediate result seemed to promise considerable improvement, but after two years I fear that this will be slow in its progress and will not be as great as could be desired. Yet the lessened frequency of his epileptic attacks is a positive improvement and he is certainly somewhat less dull than he was when I first saw him.