CRANIOSTENOSIS
WITH NOTES ON A MODIFIED OPERATION FOR THE BRACHYCEPHALIC FORM

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Cranioskerosis is the premature synostosis of two or more membranous bones of the cranial vault. Although normally the sutures are apparently closed at the end of the first year because the serrated margins become interlocked, the x-ray examination reveals that the bones are not actually fused. During this period, growth of the skull is feasible because of the existence of the suture spaces. Later the skull growth is only by periosteal apposition and resorption, just as in basal synchondrosis. Ordinarily the brain doubles its weight in the first 7 months of life and triples its weight in 2½ years, completing 80 per cent of its entire growth in the first 3 years of life. For this reason cranioskerosis has practical importance when it occurs during the intra-uterine period or first year of life.

Based on anatomical and clinical facts we have suggested the following classification:

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\begin{align*}
\text{Cranioskerosis} & \Rightarrow \\
\text{Incomplete} & \Rightarrow \\
\text{Scaphocephaly} & \Rightarrow \text{Compensated} \\
& \Rightarrow \text{Decompensated} \\
\text{Incomplete} & \Rightarrow \\
\text{Brachycephaly} & \Rightarrow \text{Compensated} \\
& \Rightarrow \text{Decompensated} \\
\text{Complete} & \Rightarrow \\
\text{Oxycephaly} & \Rightarrow \text{Compensated} \\
& \Rightarrow \text{Decompensated}
\end{align*}
\]

Premature closure of the sagittal suture results in a long narrow skull (scaphocephaly) and early synostosis of the coronal suture develops a broad, short and high skull (brachycephaly). When premature synostosis affects practically all the sutures the result is a high peaked skull (oxycephaly).

Other changes in the skull may also appear. The anterior fossa becomes short, with its floor oblique, and the orbit becomes shallow, with prominence of the eyes.

Cranioskerosis is compensated when a compensatory growth of the skull occurs at the open sutures. Since this is the only evidence of this condition, a markedly abnormal shape of the head results which must be corroborated by the roentgenologic findings. Cranioskerosis is uncompensated when the

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patient develops chronic increased intracranial pressure, as evidenced by headache, vomiting, mental deterioration and convulsive seizures. Papilledema and optic atrophy with blindness may occur. In these cases an air injection is advisable to rule out a tumor or some other intracranial lesion.

**OPERATIVE PROCEDURES**

The first attempts at surgical treatment of craniostenosis were made by Krause in Berlin and Eiselsberg in Vienna, performing subtemporal decompression in some cases of decompensated craniostenosis as a palliative measure for relieving the increased intracranial pressure.

The first surgical treatment with curative purposes was done by Lane on August 28, 1888. He removed a strip of bone an inch wide, extending from the anterior to the posterior fontanelle. Then he removed on each side sections of the remaining parietal bones, and as Lane described it, the space after resection resembled a cross, of which the arms were of equal length and breadth. The child (9 months old) lived 14 hours after operation. Death was attributed to the prostrating effect of the triple alcohol-chloroform-ether anesthesia used. A year later he performed the same operation on another infant in whom the space after resection resembled a letter H; the results were better—the child survived and showed some mental improvement. Unfortunately, the 2 patients operated on by Lane were imbecile infants with microcephaly, and consequently the procedure fell into disrepute.

More than 35 years elapsed until Faber and Towne adopted Lane’s operation. By this time, as we will see later, it was used as a preventive measure in oxycephaly and allied conditions.

Subtemporal decompressions for oxycephaly were performed in Cushing’s clinic at least as early as 1913. One of these patients whose end result is known was operated upon by one of us (G.H.) on July 3, 1916. This was a 9-year-old girl who had decompensated oxycephaly, with bilateral exophthalmos, papilledema and headaches. She has been relieved of her pressure symptoms for more than 20 years.

Operations for craniostenosis prior to 1924 were performed only as palliative measures after increased intracranial pressure had developed and the usual irreparable damage had occurred. Although Messmer, in 1921, advocated extirpation of synostosed sutures, the operation was never performed. It was not until 1924 that Faber and Towne advocated the revival of the Lane-Lannelongue operation to be performed sufficiently early in life to prevent the effects of premature synostosis. In June 1924, they performed a linear craniectomy in a child aged 9 months, who showed a rapidly progressing deformity of the skull due to synostosis of the sagittal and right coronal sutures, with signs of increasing intracranial pressure. Two channels of bone, 1 cm. wide, were removed by means of a transverse scalp incision. The first was longitudinal, extending from the coronal to the lambdoidal suture and to the right of the sagittal suture, and the second extending from one squamoparietal suture to the other, posterior to the coronal sutures.
The result of the operation was excellent, the shape and development of the head becoming approximately normal. Considering the fact that growth of the skull and brain is very rapid during early infancy, Faber and Towne for the first time advocated the operation during the first 6 months, before malformation has progressed too far and signs of decompensation have appeared.

In 1932, Bauer described an operation for oxycephaly consisting of a circular resection of the skull, leaving the upper segment of the skull free to rise beneath the scalp flap, thus relieving the increased intracranial pressure. Keegan,1 in 1934, performed a bilateral subtemporal decompression in 2 stages, removing the bony channels as suggested by Faber and Towne, and opening the dura only on the right side. In 1935, Keegan performed a modification of Bauer’s operation on another patient. Instead of doing a complete circular resection of the skull he spared a distance of about 3 cm. in the occipital region to permit some fixation of the skull cap.

King6,7 in 1936, devised an operation for decompensated oxycephaly, making a mosaic of the bones of the cranial vault. Fragmentation of the skull on each side created about nine fragments of bone by cutting between burre holes.

In 1943, Faber and Towne3 reported 5 cases of compensated craniostenosis in which an early operation was done for the prevention of symptoms of increased intracranial pressure and sequelae which presumably would have developed. They modified their former surgical procedure, removing strips of bone about 2 cm. wide parallel to the closed suture. In the case of scaphocephaly the bony channels were removed on either side of the sagittal suture, connecting both defects through a strip of bone removed at a point just anterior to the lambdoidal suture. For brachycephaly they excised a strip of bone posterior to each of the closed coronal sutures, extending from the anterior fontanelle to the posterior squamous portion of the temporal bones. In oxycephaly they performed a transverse and longitudinal excision parallel to the closed coronal and sagittal sutures. Recently, Ingraham,8 based on experimental observations, advocated the use of polyethylene to delay closure of artificial cranial sutures created by operation.

We were unable to find any mention in the literature of surgical procedures performed in cases of decompensated brachycephaly. The patients with brachycephaly operated on to date had compensated craniostenosis. An early operation was done to prevent the symptoms of increased intracranial pressure. Therefore, we consider it worth while to present the case report of a child who had a decompensated brachycephaly.

A 3-year-old white boy was admitted for the first time to the New England Deaconess Hospital on Nov. 14, 1947. He had had convulsions that appeared shortly after he was born. The convulsions were of generalized type with loss of consciousness lasting for about 2 minutes. He was able to walk at the end of his 1st year. He was able to speak a few words at 9 months but progressed no further. About 3 weeks before admission he lost the use of his legs. For 3 days he was unable to stand at
all; gradually he regained the use of his left leg and in a few days his right one. At the time of admission he was able to stand and walk, but only a few steps. He had always had a peculiarly shaped head.

Examination. The neurologic findings were negative except for bilateral choked disks. Examination of the head revealed flattening in the anterior and posterior plane with the cranial wall abnormally high. The forehead was excessively prominent and the eyes were placed far apart because the root of the nose was widened. Macewen's sign was negative.

X-rays of the skull showed that the anterior fossa was shallow with marked convolutional impressions. The coronal suture was obliterated. The middle and pos-

terior fossae were unusually deep, with the remainder of the skull appearing normal for his age (Fig. 1).

On Nov. 20, 1947, ventriculography was performed. Approximately 10 cc. of clear CSF were obtained from each side and replaced by filtered air. The subsequent roentgenograms disclosed good filling of the ventricular system, which was normal except for some symmetrical dilatation of the frontal horns (Fig. 1).

From the clinical picture and roentgenologic findings the diagnosis of decompensated brachycephaly was made.

1st Operation. Following ventriculography a right frontotemporal parietal craniotomy was carried out. The dura was extremely adherent along the closed coronal suture, which was represented by a distinct white line through the bone, but there was no particular difficulty about turning up the flap. The bone was thickened in this suture area, not only above but particularly down toward the region of the
pterion where the bone made a much deeper projection inward than usual between the posterior end of the frontal lobe and the temporal lobe. A block of bone here was taken out where it indented the dura to such a great extent (Fig. 2 b), and also a considerable amount of bone was taken out in the temporal region from the lower portion of the bone flap and from the bone below the flap as well. Likewise, the area of the coronal suture superiorly was removed (Fig. 2 a), and the strip of bone which remained anterior, after taking out this piece, was replaced over the dura, leaving a gap in the bone between this strip and the bone anteriorly as well as between it and the anterior portion of the bone flap, so that expansion could take place here. Furthermore, the dura was opened over the temporal region where bone had been removed in order that a liberal decompression could be secured. The posterior and upper portion of the bone flap was then replaced, after which the skin and muscle flaps were replaced and careful closure carried out in two layers with silk, without drainage.

The postoperative course was normal and the wound healed satisfactorily. By the time the patient left the hospital, there was marked improvement in his mental status as evidenced by the fact that he was less irritable, more alert and was taking nourishment better. Also, he was able to stand and walk merely by holding on to the edges of chair and table.

He was readmitted Feb. 20, 1948. At that time the child was alert and more cooperative. The gait was fair, but greatly improved as compared with that on his 1st admission. The decompression area was bulging and moderately tense. The optic disk margins still showed slight blurring, but there was no elevation.

Discussion. The head of this child was flattened in the anteroposterior plane and the vault was abnormally high, as in oxycephaly. The most striking difference lay in the breadth of the head, which is reduced in oxycephaly but was increased in our patient. It was evident in this case, as has been proved by the pathologic report on the segment of bone removed from the coronal suture at the 1st stage of the operation (Fig. 2), that the deformity was due to the premature closure of the coronal suture, and that the skull in this child had expanded laterally (Fig. 3) and vertically (Fig. 4), because growth in the anterior and posterior planes was impossible. The question arose, therefore, as to how it would be possible to enlarge the capacity of the skull so that the brain might grow anteriorly and posteriorly. The two problems in this case were, (1) to relieve the increased intracranial pressure and thus preserve vision, and (2) to plan a second plastic procedure on the left side to correct further the abnormal shape of the head. None of the various operations advised for craniostenosis would permit the solution of the problem in this particular patient since all previous operations had been performed in cases of oxycephaly or scaphocephaly, but not for cases of decom-
CRANIOSTENOSIS

Fig. 3. The head is abnormally broad and short with marked diminution of the anteroposterior diameter.

Fig. 4. The head is flattened in the anterior and posterior plane, with the cranial wall abnormally high.

pensated brachycephaly. In view of the roentgenograms and the general configuration of the child's head it was thought that the operation should be based on a modification of King’s procedure.

2nd Operation. A good-sized skin flap was turned down over the left temporoparietal region, proceeding well anteriorly to the closed coronal suture line and posteriorly terminating above the mastoid process. The area of the closed coronal suture, in addition to being thickened, showed that the suture was represented by a distinct white line through the bone just as on the right side. Burr holes were made around the periphery of the exposed skull about 3 to 5 cm. apart, and several other burr holes were made in the central part of the exposed skull. The holes were connected, using a Montenovesi forceps so that the area exposed was cut into 3 rectangular pieces of bone (Fig. 5). A small bridge of bone, holding the fragments together, was left until most

Fig. 5. Area of cranial vault cut into 3 rectangular pieces of bone.
of the operation was completed. The bone was thickened in the coronal suture area, as mentioned above, but particularly down toward the pterion where the bone made a much deeper projection inward than usual between the posterior end of the frontal and temporal lobe. A moderate amount of bone was removed in the temporal region as an additional helpful measure. This completed, the remaining bridges of bone were removed. The dura was not opened, owing to the fact that there was no papilledema at this time. Strips of gelfoam were placed between the fragments of bone. The skin flap was replaced and closed with two layers of interrupted silk sutures. It was noticed that there was some expansion of the skull wall, particularly over the frontal area, in comparison to the opposite side, at the completion of the operation. A dressing was applied with an outside protection of plaster shell.

The postoperative course was uneventful and the wound healed satisfactorily. The patient was discharged on Mar. 12, 1948 (Fig. 6), showing marked improvement both in physical condition and mental development, the physical improvement being pronounced as far as his gait was concerned.

Three months later the patient was seen at the clinic (Fig. 7); he showed constant pronounced improvement over his condition before he was operated on. He was able to walk freely and had begun to say a few words. The shape of the head on the left side was practically normal. The right decompression was full but not tense.
CRANIOSTENOSIS

SUMMARY

A new classification of craniostenosis, based on anatomical and clinical facts, has been suggested.

An historical review of the evolution of surgical treatment of craniostenosis is discussed and analyzed.

A modified operation for the decompensated brachycephalic form is offered.

REFERENCES