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Premature Closure of the Cranial Sutures

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INDEX

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I. UNIVERSITY OF MINNESOTA MEDICAL SCHOOL
CALENDAR OF EVENTS

February 1 - February 4, 1947

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Saturday, February 1

7:45 - 8:50 Orthopedics Conference; Wallace H. Cole and Staff; Station 21, U. H.

9:00 - 9:50 Surgery-Roentgenology Conference; O. H. Wangensteen, L. G. Rigler, and Staff; Todd Amphitheater, U. H.

9:00 - 9:50 Medicine Case Presentation; C. J. Watson and Staff; M-515, U. H.

10:00 - 12:00 Medicine Ward Rounds; C. J. Watson and Staff; E-221, U. H.

10:00 - 12:50 Obstetrics and Gynecology Grand Rounds; J. L. McKelvey and Staff; Station 44, U. H.

11:00 - Anatomy Seminar; Anaphylactic mechanisms in encephalitis; Berry Campbell; 226 I. A.

Monday, February 3

9:00 - 9:50 Roentgenology-Medicine Conference; L. G. Rigler, C. J. Watson and Staff; Todd Amphitheater, U. H.

9:00 - 10:50 Obstetrics and Gynecology Conference; J. L. McKelvey and Staff; Interns' Quarters, U. H.

11:00 - Roentgenology-Medicine Conference; Veterans' Hospital.

11:00 - 12:00 Physical Medicine Conference; Fundamentals of Electrical Stimulation; William Kubicek; W-200 U. H.

12:15 - 1:15 Obstetrics and Gynecology Journal Club; M-435, U. H.

12:30 - 1:20 Pathology Seminar; Induction of mouse leukemia; Harry W. Mixer; 104 I. A.

12:15 - 1:30 Pediatrics Seminar; Irvine McQuarrie and Staff; 6th Floor Seminar Room, Eustis, U. H.

12:00 - 1:00 Physiology Seminar; Antigen antibody - neurotrophic virus disease; Berry Campbell; 214 M. H.

4:00 - School of Public Health Seminar.

Tuesday, February 4

9:00 - 9:50 Roentgenology-Pediatrics Conference; L. G. Rigler, I. McQuarrie and Staff; Eustis Amphitheater, U. H.

10:30 - Surgery Reading Conference; John R. Paine; Small Conference Room, Bldg. I, Veterans' Hospital.

12:30 - 1:20 Pathology Conference; Autopsies; Pathology Staff; 102 I. A.

2:00 - 2:50 Dermatology and Syphilology; H. E. Michelson and Staff; Veterans' Hospital, Bldg. III.

3:15 - 4:15 Gynecology Chart Conference; J. L. McKelvey and Staff; Station 54, U.H.

3:30 - Clinical Pathological Conference; Veterans Hospital.

3:45 - 5:00 Pediatrics Staff Rounds; I. McQuarrie and Staff; W-205, U. H.

4:00 - 4:50 Surgery-Physiology Conference; Inflammation, effective external pressure; Roger M. Reineke and Fred Kolough; Eustis Amphitheater, U. H.

5:00 - 5:50 Roentgenology Diagnosis Conference; at Veterans' Hospital.

Wednesday, February 5

8:00 - 8:50 Surgery Journal Club; O. H. Wangensteen and Staff; M-515, U. H.

8:30 - 10:00 Psychiatry and Neurology Seminar; Staff; Station 60 Lounge, U. H.

11:00 - 11:50 Pathology-Medicine-Surgery Conference; Gastric ulcer, myocardial infarction; E. T. Bell, C. J. Watson, O. H. Wangensteen and Staff; Todd Amphitheater, U. H.

12:00 - 1:00 Physiological Chemistry Journal Club; Staff; 116 M. H.

4:00 - 6:00 Medicine and Pediatrics Infectious Disease Rounds; W-205, U. H.

Thursday, February 6

8:30 - Surgery Grand Rounds; John R. Paine and Staff; Veterans' Hospital.

9:00 - 9:50 Medicine Case Presentation; C. J. Watson and Staff; Todd Amphitheater, U. H.

10:00 - 12:00 Medicine Ward Rounds; C. J. Watson and Staff; E-221, U. H.

10:30 - Roentgenology-Surgery Conference; Veterans' Hospital.

12:00 - 1:00 Physiological Chemistry Seminar; Chemical assay of hormones; Saul Cohen; 214 M. H.

4:30 - 5:20 Ophthalmology Ward Rounds; Erling Hansen and Staff; E-534, U. H.

4:30 - 5:20 Bacteriology Seminar; 214 M. H.

5:00 - 5:50 Roentgenology Seminar; Review of Radiological Society of North America Meeting; Walter H. Ude, J. Richards Aurelius, and W. K. Stenstrom; M-515 U. H.

Friday, February 7

9:00 - 9:50 Medicine Grand Rounds; C. J. Watson and Staff; Todd Amphitheater, U.H.

9:00 - 10:00 Pediatric Grand Rounds; I. Mc uarrie and Staff; Eustis Amphitheater

10:00 - 11:50 Medicine Ward Rounds; C. J. Watson and Staff; E-221, U. H.

10:30 - Medicine Grand Rounds; Veterans' Hospital.

10:30 - 12:20 Otolaryngology Case Studies; L. R. Boies and Staff; Out-Patient Otolaryngology Department; U. H.

11:30 - 1:00 University of Minnesota Hospitals General Staff Meeting; Vitamin D_2 in the treatment of cutaneous tuberculosis; John R. Haserick and Henry E. Michelson; New Powell Hall Amphitheater.

1:00 - 2:00 Dermatology and Syphilology; Presentation of Selected Cases of the Week; H. E. Michelson and Staff; W-312, U. H.

1:00 - Roentgenology-Neurosurgery Conference; H. O. Peterson, W. T. Peyton and Staff; Todd Amphitheater, U. H.

II. PREMATURE CLOSURE OF THE CRANIAL SUTURES

Donald R. Simmons
William T. Peyton

Introduction

Craniostenosis is a relatively rare anomaly in which the sutures of the skull close prematurely causing compression of the intracranial contents. Many variations of this anomaly occur depending upon which sutures close prematurely, and also depending upon what other anomalies may be associated with the craniostenosis. This variability in the clinical picture has resulted in a poor understanding of craniostenosis. Therefore in this report, in addition to discussing the types of craniostenosis, an attempt is made to classify the various forms of this condition in a manner which would help to eliminate some of the confusion which now exists.

Although the surgical treatment of craniostenosis is not new there is still little uniformity of opinion as to what operations should be employed in the treatment of this anomaly and some authors still express doubt as to the efficacy of any type of operation. This lack of confidence in the results which may be obtained from surgical intervention is not without basis. There have been few reports of long term successful results following operation.

It is probable that the surgical results might be more satisfactory if the indications for such operations were better understood, and if for each case a variable operative procedure was performed. Because suitable operations do not seem to have been utilized in all reported cases, the various surgical procedures which have been suggested for the treatment of craniostenosis are reviewed and, in addition, a modified operative procedure is described. This modification for the prevention of bony regeneration across the newly made skull defects should be effective in eliminating the chief cause of poor results following craniectomy for craniostenosis.

Reports in the literature of cases of craniostenosis which have been operated upon are not numerous, and for this reason reports on four additional cases illustrating many of the problems which the physician meets in dealing with craniostenosis would seemto be of value.

History

In 1851 Virchow[1] first described the relationship between premature closure of the cranial sutures and the distressing symptoms which result from such closure. To this condition he gave the name craniostenosis. In 1855 MacKenzie[2], in his textbook on ophthalmology, reported that there were certain cases of "hydrocephalus" in which the skull height was increased and the orbits were shallow, and that in these cases exophthalmos and blindness occurred. From his description there can be little doubt that he was dealing with cases of premature closure of the cranial sutures rather than cases of hydrocephalus. Von Graefe[3], in an article on neuroretinitis written in 1866, described the case of an eight year old boy who had a very high narrow head, visual impairment, epilepsy, exophthalmos, and papilledema. Von Graefe suggested that the papilledema was due to increased intracranial pressure secondary to the skull deformity. He was the first to recognize the importance of visual impairment in craniostenosis. Following this paper of Von Graefe many case reports of this anomaly were published especially in the German literature but there was a tendency at this time to confuse craniostenosis with microcephaly. It was not until after roentgenography was introduced that these two anomalies were separated with any degree of accuracy.

In 1890 Lannelongue[4] published the first report of an attempt to relieve the symptoms of this disease by removing strips of bone along the prematurely synostased sutures which would allow for expansion of the skull. In 1892 Lane[5] reported two cases upon which he had done linear craniectomies. In 1894 Jacobi[6] was able to find reports of 33 cases upon which operations had been performed to relieve craniostenosis but he severely criticized operations to correct cranial deformities. He pointed

out that while there might be some justification for operations in those patients who had a true premature synostosis of the cranial sutures, most of these operations were being done on true microcephalics in whom an erroneous diagnosis of craniostenosis had been made.

In spite of the paucity of published reports on the surgical treatment of this disease previous to 1900, the fact that Jacobi was able to collect 33 cases which had been operated is evidence that operations on the skull were performed rather frequently on patients with craniostenosis or patients who were thought to have craniostenosis. Because the difference between premature synostosis and microcephaly was not fully recognized at this time, and because the operative mortality in those cases which were operated was nearly 50%, all operations for cranial deformities fell into disrepute and even now there are some who still believe that surgical treatment offers no benefit in cases of premature synostosis of the cranial sutures.

In the last 30 years there has been published an extensive literature relative to craniostenosis, and during this period much has been learned concerning this anomaly, but a great deal of confusion still exists concerning the etiology, symptoms, classification, and treatment of craniostenosis. In more recent years there has been a revival of interest in the surgical treatment of craniostenosis, and several new operations have been suggested for the treatment of this condition.

Normal Skull Growth

There is commonly a slight separation of the cranial bones at birth but a firm fibrous union is present by the end of the fifth or sixth month. By the end of the sixth week the posterior and lateral fontanelles are obliterated and the normal closure of the anterior fontanelle occurs between the tenth and sixteenth months.

Normal growth of the skull and obliteration of the sutures of the skull is directly dependent upon normal brain growth. Thus, if the brain remains small the skull will be correspondingly small, and the sutures may be obliterated early. If, on the other hand, the brain expands more rapidly than normal, the skull becomes larger than normal and the fontanelles and sutures remain open for a longer period. That this is true is shown by the fact that in microcephalus there is commonly premature obliteration of the fontanelles and premature closure of the sutures. In hydrocephalus, on the other hand, obliteration of the fontanelles and closure of the sutures is delayed. Further evidence that growth of the skull is directly dependent upon brain growth is shown by the fact that in cases in which there is atrophy or lack of growth in one cerebral hemisphere in early childhood, the corresponding side of the skull is smaller than on the normal side. Growth of the brain has been demonstrated by Scammon and Dunn[7] to form a parabolic curve when plotted graphically, the period of rapid growth lasting for only the first two to three years. Since the growth of the cranium is dependent upon the growth of the brain, the rate of growth of the skull parallels the rate of growth of the brain. After the age of three years there is only slight increase in intracranial capacity, and any increase in the head size is due largely to thickening of the bones of the skull, and growth of the accessory sinuses and the bones of the face.

Giblin and Alley[8,9] believe that skull growth occurs in two ways. First: growth occurs by accretion of bone on the external convex surface of the skull with simultaneous resorption of bone from the inner concave surface. Growth also occurs in a lateral direction from the sutures of the skull. Giblin and Alley have demonstrated experimentally that this lateral growth from the sutures does occur. Proitsky also has demonstrated experimentally that growth of the cranial bones in a lateral direction is from the suture lines, and that there is no interstitial growth of the bones of the skull. This experimental observation is corroborated by the clinical impression that following synostosis of a suture, no growth takes place perpendicular to that suture. Giblin and Alley have shown that in experimental animals the cranial sutures will fuse

if normal movement of these sutures is stopped by placing a single bridge of bone across the sut re line. Thus, we see that even in normal skulls synostosis of a suture once it starts may tend to perpetuate itself until the suture is completely obliterated. Bolk[10] has shown that the sutures may be completely obliterated in what appear to be perfectly normal skulls at a much earlier age than is usually considered normal. In a study of 1820 skulls ranging in age from three to twenty years he found a synostosis of one or more sutures in 343 of the skulls. The lamboidal suture was the one most commonly affected and this was obliterated in 10% of the skulls he examined. The sgggital suture was synostosed in 2½% of the skulls, and the other sutures were less commonly affected. Greig in his discussion of oxycephaly classified certain cases of premature closure of some of the cranial sutures without cranial deformity as false oxycephaly. Apparently, these cases of false oxycephaly of Greig and the premature closure of cranial suture described by Bolk are the same. It seems unwise to use the term false oxycephaly to describe what apparently is a normal variation of skull growth because the term false oxycephaly, suggests a pathological condition. Bolk believes that when this premature synostosis occurs, it usually does occur between the ages of four and six years, because the percentage incidence of premature obliteration of the sutures remains constant after the sixth year. This physiological early closure of the cranial sutures occurs, then, at a time when the brain has practically attained its adult size, and it is probably for this reason that no symptoms result in these cases.

Classification of Premature Synostosis

One of the chief difficulties in arriving at a clear understanding of premature closure of the cranial sutures is the complex descriptive terminology which has accumulated. The terminology is confusing because different names have been applied to identical forms of the anomaly and also because many superfluous subdivisions have been made to separate minor varients. In addition, as previously stated, pathological terms have been applied to what are apparently variations of normal skull growth.

Some of the terms used to describe this anomaly are: premature synostosis, synostosis cranii, craniostenosis, oxycephaly, delayed oxycephaly, false oxycephaly, pyrgocephaly, turricephaly, turmschadel, steeple-skull, turret-skull, sugarloaf skull, scaphocephaly, plagiocephaly, clinocephaly, trigonocephaly, leptocephaly, acrocephaly, craniofacial dysostosis of Crouzon, and acrocephalosyndactlysm.

To add to the confusion of this complicated terminology some investigators would separate and classify as an entirely different type of cranial anomaly types, which in reality should be classified as varients of synostosis cranii, Thus Greig[11], for example, believes that oxycephaly is a disease process separate from the other forms of premature synostosis of the cranial sutures because he believes that in oxycephaly true suture lines are never developed. He believes that in cases of oxycephaly "where the bones touch they fuse", and therefore any skull growth which does occur takes place before the bones of the skull have approximated, but fusion immediately follows approximation so that at no time are suture lines ever formed. Sear[12], on the other hand, states that he has observed occasional cases of severe oxycephaly in which certain of the suture lines remained open. This difference of opinion may be due to the fact that Greig was studying museum skulls, while Sear's observations were made on clinical cases. Whereas it is true that the oxycephalic skull has a shape which is distinctly different from the other forms of craniostenosis, there does not appear to be sufficient evidence to indicate that it is different disease process. In fact, there is considerable evidence to prove that oxycephaly is closely related to the other types of craniostenosis. It has been observed frequently that other members of a family in which there is a member with typical well-advanced oxycephaly may have definitely abnormal but not oxycephalic skulls. The deformities commonly associated with oxycephaly are also found associated with other forms

of craniostenosis. Cases of oxycephaly are seen in whom the sutures are present early and when the sutures close the skull assumes a typical oxycephalic shape. Greig[11] refers to these as cases of delayed oxycephaly, a process which he considers as unrelated to true oxycephaly. All forms of premature synostosis of the cranial sutures are in this report considered to be merely variants of a single disease process. This concept is not new! Bertolotti[13] in 1914 suggested that all these deformities be grouped together, and called the cranial dystrophies. Sear[12] was of the same opinion that all of these deformities should be considered as a single anatomic-pathologic entity.

In spite of this confusion concerning the classification of these anomalies, a simple workable classification can be made by eliminating duplications in terminology and by disregarding the minor variations which are probably of little significance. The following classification seems to be reasonably simple, and yet it separates the important groups making up the condition.

A. Complete early premature synostosis of the cranial sutures (oxycephaly, turricephaly, turmschadel, etc.)

   1. Oxycephaly without facial deformity.
   2. Craniofacial dysostosis of Crouzon
   3. Acrocephalosyndactylism
   4. Delayed oxycephaly (onset after birth.

B. Incomplete early synostosis of the cranial sutures.

   1. Scaphocephaly - premature closure of the saggital sutures.
   2. Brachycephaly - premature closure of the coronal sutures, or of the coronal and lamboidal sutures.
   3. Plagiocephaly - asymmetrical premature closure of the sutures.
   4. Mixed.

C. Late premature synostosis of the cranial sutures after the skull has reached or nearly reached adult size so that no deformities and no symptoms result. (This is included only to show its relation to true craniostenosis and to make it clear that surgical treatment is not indicated. This process should not be considered pathologic.)

Etiology

The etiology of craniostenosis is not known, but many possible causes have been suggested. Virchow[1], in his original description of the disease in 1851, suggested that premature synostosis might follow inflammation of the meninges. Others have also suggested that intrauterine meningitis might be the etiological agent. Rickets, syphilis, glandular dysfunction, and birth trauma have all been mentioned as possible causes. Young[14], in studying two cases of oxycephaly found the blood calcium levels to be 18 mgm% in one and 20 mgm% in the other. Because of these findings he suggested that the disease might be on the basis of an endocrine imbalance with calcium retention. There have been no other studies to confirm his views.

Park and Powers[15] believed that the disease was due to some inherent defect in the germ plasm rather than being due to any mechanical factors. There is some evidence to substantiate this point of view. There is no doubt that at least in certain instances the disease is familial and also craniostenosis is frequently associated with other congenital defects.

Greig[11] and Ovilive and Pasel[16] have suggested that the development of craniostenosis may be related to disturbances in growth of the sphenoid. Ogilive and Pasel, in studying a case of scaphocephaly, found that the greater wings of the sphenoid were small, and the lesser wings were larger than normal, but the reverse was true in cases of oxycephaly, the greater wings of the sphenoid being much larger than normal. They felt that this enlargement of the greater wings of the sphenoid probably accounted for the wide skull, temporal bulging, and shallow orbits, abnormalities which are present with oxycephaly. Greig and Ogilive and Pasel have noted similarly that in hypertelorism the lesser wings of the sphenoid were over-

developed. Interestingly enough hypertelorism and craniostenosis are very commonly found in the same patient.

Mann[17], in discussing the development of oxycephaly, stated that there was failure of normal growth of the lateral portions of the base of the skull which develops from the visceral mesoderm of the first visceral arch. On the other hand, he felt that the central portion of the base of the skull which arises from the periaxial mesoderm develops normally.

Morselli[18], Rieping[19] and Sear[12] have suggested that premature synostosis of the cranial sutures is due to the fact that the centers of ossification in the adjacent bones are in too close proximity. Rieping[19] thought that there was in addition some fault in the blastodermal matrix separating the bones. Giblin and Alley[8] have demonstrated in experimental animals that synostosis of the cranial sutures is influenced by the mobility of the bones when they demonstrated that synostosis of a suture will occur if the bones are immobilized by a bony bridge across the suture. This being true, a premature synostosis may be self perpetuating once it starts.

It has long been recognized that heredity might be an important factor in the etiology of this disease, and the hereditary or familial tendency of craniostenosis is shown by numerous reports of this process occurring in siblings or in succeeding generations in the same family [16,20,21,22,23,24,25,26,27,28]. The mother of one of our cases (Case #2) of craniofacial dysostosis, had a high narrow head, hypertelorism, divergent strabismus and prognathism. Gunther[29], in a review of the literature on oxycephaly, found in several pairs of twins a typical oxycephalic deofmity occurring in both.

Symptoms

Premature closure of the cranial sutures occurs much more commonly in males than in females. Sharpe[30] has stated that males are affected five times as frequently as females.

It is apparent that the severity of the skull deformity and the severity of the symptoms resulting from premature synostosis depend not only upon which sutures are obliterated but also upon the age at which this synostosis occurs, a severe degue of deformity occurring only if the process begins early in life. It is probable that no symptoms will occur if synostosis begins after the age of three years. As has been stated previously, these cases should probably be classified as a variation of normal skull growth rather than a pathological premature closure of the sutures. Although mild deformity of the skull may result these cases probably never need treatment.

All cases of premature synostosis have abnormally shaped heads, the shape depending upon which sutures are prematurely closed, and also in what order they close. If the saggital suture closes doliocephaly results, but if the coronal sutures close the head becomes brachycephalic. Closure of the sutures on one side will cause an asymmetry of the skull, and closure of all of the sutures will cause the skull to be tower shaped because of the pressure upward against the open anterior fontanelle by the still expanding brain.

Because the change in the shape of the skull from normal to definitely abnormal is very gradual, the parents usually do not realize that a pathological process is present until the deformity is far advanced. However, frequent observation by a physician may establish the diagnosis at an early stage of the disease, especially if head measurements are taken routinely. Early recognition is of the greatest importance since the results of sugrical treatment in this disease depend in large measure upon the promptness with which surgical intervention is initiated.

Certain other abnormalities of the skeletal system are commonly associated with premature synostosis of the cranial sutures. There may be shallow orbits resulting in exophthalmus. Premature synostosis of the sutures of the facial bones may occur resulting in the typical facial appearance described by Crouzon[23],

characterized by exophthalmus, divergent strabismus, a saddle shaped nose, decreased prominence of the maxillary processes, a prognathous jaw, a very high arched palate, and dental abnormalities due to the small maxilla. Many, especially the oxycephalic variety, have a divergent strabismus and increased distance between the eyes as described in 1924 by Greig[31] when he reported two cases presenting this deformity and suggested it be called hypertelorism. Neither of Greig's cases, however, had a premature closure of the cranial sutures. He had the opportunity to examine the skull of one of these cases, and from his examination he concluded that the deformity was caused by an exaggerated growth of the lesser wing of the sphenoid, whereas the greater wing remained smaller than normal. Greig felt that hypertelorism should be considered as an entirely separate type of craniofacial deformity rather than a form of craniostenosis. Hypertelorism may be seen not only in cases of craniostenosis, but also associated with other diseases or it may even be an isolated finding. Braithwaite[32] has reported a case of hypertelorism in a mongol. Hypertelorism is an excellent descriptive term and should be retained for this condition, but it is doubtful if it should be considered as an entirely separate disease. It is more probable that it may be one of the characteristics of craniostenosis, but at times it may occur as an isolated finding or it may be associated with diseases other than craniostenosis.

Syndactylism associated with oxycephaly was first described by Wheaton[33] in 1894, and since that time others have also discussed this syndrome and applied to it the name acrocephalosyndactylism. The syndactylism may affect either the hands or the toes, and it may be associated with other congenital anomalies in the bones of the extremities. Some authors believe that acrocephalosyndactylism should be considered a separate disease rather than a form of oxycephaly with associated anomalies. As in the case of hypertelorism, it is more probable that this syndrome is one of the variants of craniostenosis rather than aseparate disease process. Many other congenital deformities including naevi, mongolism, hairlip, hypoplasia of the genitalia, pituitary dysfunction, kyphosis, spina bifida, genu valgum, pes planus, osteogenesis imperfecta, hydrocephalus, and hemolytic icterus have been reported associated with premature closure of the cranial sutures.

Except for deformity of the skull itself, the most common symptoms of craniostenosis are those related to the eyes. For this reason much of the early writing on this disease is to be found in the ophthalmologic literature, and even now many of these cases are seen first by the ophthalmologist.

Exophthalmus is a common symptom, and is due to a shallow orbit which results from the orbital roofs being more vertical than normal. External strabismus is commonly associated with the exophthalmus, the external strabismus being due to the fact that the orbital axes assume a downward and outward direction. At times the exophthalmos may be of such a severe degree that the lids cannot be closed and corneal ulceration occurs.

Loss of vision secondary to optic atrophy is frequently found in severe degrees of craniostenosis. Gunther[29], in reviewing the literature, found that 67 of a total of 74 cases had optic atrophy. Several theories have been proposed as a cause for the atrophy. Schloffer[36] and Van der Hoeve[37] have suggested that the optic foramina are smaller than normal, and that the pressure on the optic nerves by the walls of the constricted foramina cause optic atrophy and blindness. Behr[38] and Beaumont[39] believe that the optic atrophy is due to an angulation of the optic nerve caused by the combination of a deep middle cranial fossa and an orbital roof which assumes nearly a vertical plane resulting in an overhanging superior rim of the optic foramen. This causes the angulated optic nerve to be compressed between this overhanging roof of the optic foramen and the pulsating internal carotid artery below. The most commonly accepted theory is that the blindness is secondary to papilloedema sssociated with generalized increased intracranial pressure. Bennett, Keegan and Hunt[40] have found intracranial pressures ranging as high as 80 mm.

of Mercury, and Gunther[29] has reported cases having pressures of 450 mm. of water. Papilloedema is commonly seen, and one would expect that optic atrophy would follow on this basis alone, especially since the increased intracranial pressure is of long duration. There have been reports of cases in which papilloedema occurred in adulthood long after the brain had reached its adult size. It is difficult to explain the occurrence of papilloedema in these adult patients who apparently have had no previous evidence of increased intracranial pressure.

Convulsive seizures frequently occur in craniostenosis but usually have their onset late in the course of the disease.

There is still a difference of opinion among investigators as to the mental status of these patients. Many authors, even in recent years, have insisted that the disease is not necessarily associated with mental deficiency, and many cases have been reported to confirm this opinion. Some have said that these patients may even be of superior intelligence. Obviously, one reason for such statements is that these authors were including cases in which there was a late development of premature closure of the sutures, cases which may have no symptoms, and as previously stated, these cases should not be considered as pathologic.

So many cases of synostosis reported have been associated with mental deficiency that there seems to be no question but that the two are concomitant. It is true that many cases of true synostosis have been presented in the literature in which the authors stated that the mentality was normal, but it is apparent that most of these statements were based on casual observations of the patient without psychological testing to give an accurate measure of the intelligence of these patients. In reviewing the literature on this disease only one case was found in which it was quite evident that the patient's intelligence was above normal. This patient was one of the cases of Faber and Towne[41,42] upon whom a linear craniectomy had been performed. Because this patient had good growth of the skull following the operative procedure he cannot be considered as showing the usual symptoms of a patient with craniostenosis.

Roentgenographic Findings

In all cases of premature synostosis of the cranial sutures, the obliteration of the sutures can be most readily demonstrated on roentgenographic examination. The bone immediately adjacent to the obliterated suture shows an increased density and it is of greater thickness here than elsewhere in the skull. The characteristic shape of the skull, depending upon which sutures have been prematurely synostosed, is readily apparent in the roentgenogram. In fact, the abnormal shape of the skull is frequently more apparent on roentgenographic examination than on physical examination. Especially is this true in older children and in adults in whom the hair hides the characteristic deformity. In addition to abnormalities in the sutures and shape of the skull certain other associated changes are frequently visible in the roentgenogram. The orbits may be shallow and the orbital roofs may assume a nearly vertical position. The middle fossa may be abnormally deep and the auditory meati assume a low position. The superior orbital fissure is short and narrow. The frontal sinuses are small. The bones of the skull are much thinner than normal. There may be signs of increased intracranial pressure as evidenced by digital markings over the vault and decalcification of the dorsum sella. Sear[12] has reported a case in which there was a Luckenschadel. The skull is usually thinned but occasionally it may be thicker than normal. The thinning, if it does occur, is at the expense of the diploe. Sear has shown that the basal angle is altered in these patients. In scaphocephalus there is a platybasia with an increase in the basilar angle, and in Crouzon's disease there is a basilar kyphosis with a decrease in the basilar angle. Decrease in the size of the optic foramen has been reported in a few cases.

Careful study of the roentgenograms is especially important if surgical intervention is considered because the surgical procedure which is chosen may in large part depend upon which sutures are synostosed.

Diagnosis

When craniostenosis is severe the diagnosis of this disease is a relatively simple matter. The typical cranial deformity, the associated deformities of the bones of the face and of the extremities, and the signs of increased intracranial pressure, the evidence of cerebral damage, and the typical roentgenographic findings all serve as unmistakable signposts to a correct diagnosis.

However, if treatment is to achieve maximum results the diagnosis must be made prior to the time that irreparable damage has been caused to the brain by the nonyielding skull. In the early stages of craniostenosis the diagnosis is much more difficult. Roentgenographic evidence of obliteration of one or more sutures must be present in order to make a diagnosis of craniostenosis. However, roentgenographic evidence of premature closure of suture lines is not in itself irrefutable evidence of the presence of craniostenosis. This is true because of the difficulty at times of satisfactorily demonstrating the sutures roentgenographically in normal children, especially if the skull is thin so that the contrast between suture and bone is poor.

The shape of the skull itself is of great importance and measurement of its expansion over a period of time should be carefully plotted to see how closely it approaches the normal growth curve. The shape of the skull in early cases will not be as definitely abnormal as it will at a later date, and the great variability in the shape of normal skulls adds to the difficulty in making a correct diagnosis. Still one is not justified in temporizing until the head is unmistakably under size or until it is extremely abnormal in shape before arriving at a correct diagnosis and instituting surgical treatment.

The typical deformities so frequently associated with craniostenosis if present may be of the utmost value in arriving at the diagnosis. Thus, if the typical facial deformities or deformities in the extremities are present they establish the diagnosis even with minimal cranial vault deformity.

Careful family histories may reveal that similar deformities are present in other members of the family and this, together with even minimal findings in a patient is enough to make a diagnosis early in the course of the disease.

Visual difficulties and signs of increased intracranial pressure occur late in the course of the disease, and, if possible, surgical treatment should be instituted before these occur. By the time these symptoms are present there has undoubtedly been damage to the brain and the optimum time for surgical intervention is past.

Mental development is retarded in these children, and this may be the primary indication for surgical therapy. However, it is often extremely difficult to differentiate synostosis from microcephaly which is also characterized by decreased expansion of the skull and early synostosis of the cranial sutures. In many instances excellent clinical judgment will be necessary to determine whether or not a true craniostenosis is present.

Treatment

In the latter part of the last century the first attempts were made at surgical intervention in cases of children with small heads. In 1890 Lannelongue[4] reported the case of a four-year old girl with microcephaly and idiocy, on whom he performed a linear cranictomy paralleling the saggital suture and reported improvement in mentality following the operation. In 1892 Lane[5] reported two cases in which cruciate types of linear craniectomies were performed. A strip of bone was removed parallel to the saggital suture and another strip just posterior to the coronal suture. The first case died 14 hours postoperatively, and in the second case he reported there was some improvement in mentality following the operation. It is now evident on reading the case histories reported by these surgeons that the children operated upon had microcephaly rather than craniostenosis, but the authors did not recognize the difference between these two conditions. In 1894 Jacobi[6] was able to collect re-

ports of 33 cases that had operations performed in America either for premature synostosis or microcephaly. Of this group there were 14 postoperative deaths. He criticised severely those who were performing operations on microcephalics, although he admitted that operative procedures might be useful in cases of premature synostosis. Surgery for craniostenosis fell into disrepute, because most surgeons apparently did not recognize the difference between craniostenosis and microcephaly, and because there was a high operative mortality. There are still many who feel that successful treatment of craniostenosis is impossible.

In 1908 Dorfmann[43] reported a case of a 4 year old girl with oxycephaly and severe papilloedema. This case was trephined by Eiselberg and Dorfmann felt that the operation was successful in saving the patient's vision.

Since that time Sharpe[30], Bedell[20], Watts[44], Skipper[27], and Bennett Keegan and Hunt[40] have all reported cases in which either unilateral or bilateral subtemporal decompressions have been done to relieve symptoms resulting from increased intracranial pressure. Bedell[20] had one case that died immediately postoperatively, but the other reports have all been favorable, and would indicate that the operation has merit. However, the total number of cases in whom this procedure has been done is small, and the patients have been followed for too short a time after surgery to arrive at any definite conclusions as to the results of subtemporal decompression in this disease.

In 1913 Schloffer[36] first recommended a resection of a portion of the roof of the orbit adjacent to the optic nerve to relieve the visual symptoms in these cases. He felt, as have other investigators, that the optic atrophy and papilloedema so commonly seen in cases of craniostenosis was due to direct constriction of the optic nerve rather than to generalized increased intracranial pressure. Hildebrand[45], in 1923 reported four cases in whom a modified form of the Schloffer operation was performed to relieve increasing visual impairment and he reported that the results were satisfactory in all four cases. In 1924 Elschnig[46] performed this operation on three patients but he reported no follow-up studies on the patients. Schloffer's operation has not been used outside of Germany, and most investigators believe that the operation has no value because it is based on the false premise that the visual difficulties are due to direct optic nerve involvement rather than to increased intracranial pressure.

Bauer[47], in 1932, reported on one case in which a circular type of craniectomy was performed separating the vault from the remainder of the skull, thereby permitting an increase in intracranial capacity by elevation of the separated portion of the vault. The dura was not opened. Bauer claimed that there was no new bone formation six months after the operation, but the published reproductions of the roentgenograms on this patient seem to show that new bone is present at the site of the craniectomy. Although Bauer has been given credit for developing this operation, Sharpe[30] reports that Cushing in 1911 performed the same operation but abandoned it because of the rapid regeneration of bone which prevented adequate expansion. Bennett, Keegan and Hunt[40] in 1936 used this circular craniectomy in one case. Results of these operations are not known.

King[48], in 1938, proposed a procedure for the treatment of craniostenosis which he calls the morcellation operation. Multiple grooves are cut through the skull separating the cranial vault into a checkerboard pattern. King believed that this procedure would allow for a more symmetrical expansion of the brain than any of the previously suggested procedures. King[48,49,50] and Woodhall[51] have reported a total of seven cases in which this morcelation operation was performed and have found the results to be satisfactory.

Faber and Towne[41,42] have again advocated the use of a linear craniectomy as used by Lane[5] and Lannelongue[4] during the last century. They have reported on five cases in which this operation was performed, and their results have been encouraging. They have been able to observe one of the patients for a period of 14 years following operation, and the result in this case has been

excellent. This is the only reported case in which there has been an adequate postoperative period of observation so that the eventual result after operation could be evaluated. Faber and Towne[41] were the first to point out that early operation is imperative if these cases are to receive maximum relief of symptoms. This is true because the period of rapid brain growth occurs in the first few months of life, and if operation is delayed until after six or eight months of age the damage to the brain may be irreparable, and operation may be of little benefit.

Dandy[52], in 1943, reported a new operative technique for the treatment of craniostenosis. He turned a large frontoparietal horseshoe shaped flap with its base hinged at the midline. The flap is then elevated from its bed and held in this elevated position like a trap door by a small piece of bone which is fastened to the inferior edge of the flap, and the squamous portion of the temporal bone. This operation would appear to be technically more difficult than others which have been proposed, and it would not seem to be any more effective in increasing the volume of the intracranial cavity.

Even in recent years it has been stated that there are no therapeutic measures which are successful in relieving the symptoms resulting from craniostenosis, but this has certainly not been the experience of most surgeons who have operated on these cases. Although the results are not perfect a certain measure of relief of symptoms has been obtained in most cases, and in at least one case[42] the final condition many years after operation was very satisfactory.

The large majority of cases of craniostenosis are not of severe degree and in these cases no therapy is necessary. However, there are certain very definite indications for surgical intervention. Probably the most important indication for surgical intervention in cases of craniostenosis is mental retardation. This indication for surgical therapy has received too little emphasis in the literature. Mental retardation is one of the earliest symptoms and probably the only symptom throughout life in many cases of craniostenosis. Every child with craniostenosis should be watched closely for mental retardation and surgical therapy should be instituted at the first sign of such retardation. It cannot be emphasized too strongly that if the surgery is to be of maximum benefit it must be done at an early age. Faber and Towne[41,42] have recommended that these patients be operated before the age of six months because if surgery is delayed longer than this irreparable damage is sustained by the brain. Blindness is one of the most common symptoms in this disease, and surgery should be done at the first indication of failing vision. Gunther[29] found that in 74 cases collected from the literature 67 had optic atrophy and 45 amaurotic. Brac[53], in reviewing the literature on oxycephaly, found that of a total of 85 cases, 25 had a bilateral total amaurosis, and in 30 additional cases there was a unilateral amaurosis. Occasionally surgery may be necessary for the relief of severe and persisten headaches which accompany the disease.

Many operative procedures, several of which have merit, have been recommended for the treatment of the disease. Each individual case must be evaluated and the operative procedure chosen which seems most applicable to it. In infants the linear type of craniectomy as used many years ago by Lane and Lannelongue and as advocated by Faber and Towne in recent years appears to be most useful. Because the bones of the skull in infants are thin and flexible the linear type of craniectomy will allow for adequate expansion of the brain, and will allow the head to assume a normal shape. After linear craniectomy, especially in children, the bone tends to form bridges across the artificial suture lines and expansion is again arrested. This growth of bone across the craniectomy is probably one of the reasons for poor results following surgical intervention in these cases. Tantalum foil interposed between the bone edges should prevent this regeneration and allow continued expansion without additional operations. It has now been used in two cases (Case No. 1 and Case No. 4.)

The morcellation procedure proposed by King is useful in older children in whom the skull is so thick and rigid that a linear craniectomy alone will not allow for adequate expansion of the brain and resumption of a normal shape by the skull. Subtemporal decompression is useful in adults with visual difficulties and headaches, and may be preferable to the morcellation operation because of its simplicity. The operations of Bauer[46] and Dandy[49] are not recommended because they seem to be technically more difficult, and they are probably no more effective in increasing the intracranial volume than are the other operations.

Decompression of the optic canal as proposed by Schloffer[36] has not been widely accepted but it is possible that in certain cases visual impairment is due to direct pressure on the optic nerves, and if so this operation should give relief. At least, it should be considered if loss of vision is progressive after increased intracranial pressure is eliminated by one previously described operation.

Prognosis

There is very little in the literature concerning the prognosis in this disease. Since the large majority of the cases reported have been in children and young adults one might suspect that this indicates that life expectancy is less than that of normal individuals but there can be no doubt that premature synostosis is at times associated with longevity. Fletcher[24] said, there is no direct evidence that the disease shortens life, but rarely are cases seen among people over 50 years of age. The prognosis for life probably depends in large measure upon the severity of the disease process. There seems little doubt that there will be an impairment of intelligence in a large number of these patients. Again, this undoubtedly depends largely on the severity of the process. Certainly, in every case of severe premature synostosis there is a danger of optic atrophy and blindness. This usually occurs in childhood, but it may occur later. Elschnig[45] reported a case of a 50 year old man with visual impairment of increasing severity. Thus, the danger of loss of vision is not past when the brain has attained its full growth but it is most prevalent during the period of rapid cerebral growth which in turn produces increased intracranial pressure.

To what extent the prognosis can be altered by surgical therapy is not definitely known. This is true because of the small number of cases that have been operated upon and followed for a sufficient period of time to be accurately evaluated. Long term follow-up study has been reported in only a single case[41]. This child was operated upon in 1927 and followed him until 1941, when he was perfectly normal in every respect. This case makes one hopeful as to the results which may be obtained by surgical intervention, but certainly we are not yet justified in drawing any conclusions as to its efficacy. The most one can say is that as a method of therapy it appears to be rational, and that in isolated instances the results as far as they are known appear to be encouraging.

Case Reports

Case No. 1.

An eleven month old male infant was admitted to the hospital on 1-22-44. The parents had noticed no abnormality until the child was eight months old, when he began to have periods during which his head would bob up and down. Birth history revealed that the labor lasted three days but the delivery was spontaneous and normal. The child weighed 9 lbs. 3 oz. at birth. He did not sit alone until he was nine months of age, but at eleven months he was able to take a few steps while holding onto someone's hand. Shortly after the onset of these attacks of "bobbing" of the head he was taken to a Pediatrician who made a diagnosis of scaphocephaly.

Examination revealed a well developed, well nourished infant. The skull was scaphocephalic, and on palpation the saggital suture appeared to be closed, with the coronal and lambdoidal sutures open. Both fontanelles were closed. The head circumference of this 9 month

old child was 48 cm. Fundascopic examination revealed nothing abnormal and the only positive neurological finding was a bilateral unsustained ankle clonus. There were no associated congenital deformities.

Roentgenographic examination of the skull revealed synostosis of the saggital suture and also showed that the lower ends of the coronal sutures were narrower than normal but the upper end of the coronal sutures was normal, and the lamboid sutures were normal. There was a slight decalcification of the dorsum sellae and the basalar angle was increased.

A diagnosis of premature closure of the saggital suture was made and the patient was operated on 1-26-44. Under general anesthesia a coronal incision was made, and the scalp was then turned forward and backward so that the superior portion of the skull was completely exposed. Strips of periosteum about 2 cm. in width were then removed from over the coronal and saggital sutures. A strip of bone 1 cm. in width was removed paralleling the saggital suture. The craniectomy was also extended along the coronal sutures so that it extended about three-quarters of the distance from the midline to the pterion. The medial 1.5 cm. of each lamboid suture was removed in the same way. The postoperative course was uneventful and he was discharged from the hospital on 2-4-44.

When seen in the outpatient clinic on 4-4-44 approximately 2 months after the operation his mother said that the bobbing motion of his head was lessening and that he seemed more alert but the head diameter at this time was still 48 cm.

X-rays of the skull on 9-12-44, seven and one half months after operation, showed that bony bridges had grown across the craniectomy so that fusion was nearly complete throughout the length of the craniectomy along the saggital suture but was irregular and incomplete across the defect along the coronal suture.

The child visited the outpatient clinic again on 10-8-46, when, approximately 20 months after operation, X-rays of the skull showed increased digital markings. A Stanford-Binet (form M) test showed that he had an I.Q. of 80. The examiner felt that the low quotient was at least in part due to poor cooperation in doing the tests. The child did poorly on those tests involving motor coordination, but he performed better than average for his age on the picture vocabulary tests. His vocabulary and memory seemed to be good for a child of his age. Thus, it is unlikely that his intelligence is any lower than would be indicated by an I.Q. of 80, and it may be somewhat higher. The final conclusion of the examiner was that the patient had low normal or borderline intelligence.

Because of the regeneration of bone across the craniectomy as shown on X-ray and because of the increase in digital markings seen on the X-rays, a second craniectomy was done on 12-11-46. Except for a small segment at the junction of the coronal and saggital sutures there had been complete regeneration of bone across the previous craniectomy along the saggital suture. At this second operation a 1 cm. strip of bone was again removed paralleling the saggital suture. The bone edge was covered with tantalum foil. The child's postoperative course was uneventful and he was discharged from the hospital one week after the operation.

Case No.2. - a $3\frac{1}{2}$ year old boy was admitted to the hospital on 2-17-44. There was a history that since birth he had prominent eyes, abnormally shaped skull, and thin watery nasal discharge. A few months prior to admission he began to have difficulty in breathing, and the parents noticed that he repeatedly stumbled or he ran into objects as though he was losing vision. The birth history was not unusual. The child held his head up a few months after birth, crawled at eight months, walked at thirteen months, and started to talk at $2\frac{1}{2}$ years. The patient's mother had had an exophthalmos since birth and had impaired vision in one eye. She had also had frequent severe headaches for many years. A maternal uncle of the patient was also

said to have had an abnormally shaped head.

Examination revealed a small, typically oxycephalic type of skull with a circumference of 44 cm. There was a beak-shaped nose, hypertelorism, divergent strabismus, exophthalmos, a prognathous jaw, a highly arched palate, and narrow maxilla with crowding of the teeth. The child breathed through his mouth and drooled saliva constantly. The exophthalmos measured 26 mm. on the right and 24 mm. on the left. Fundascopic examination showed moderate optic atrophy bilaterally. Vision seemed to be normal, although it could not be tested objectively. The physical examination was otherwise negative.

Psychometric tests using the Stanford-Binet, form L test and the Vineland Social Maturity Scale revealed that the child was of very lowest range of normal intelligence.

The roentgenographic examinations of the skull showed the only significant laboratory findings. These showed a characteristic picture of extreme craniostenosis of the oxycephalic type with greatly increased digital markings and all cranial vault sutures were completely united, but the naso-frontal and the spheno-occipital sutures were open. These were the only sutures at the base of the skull which could be visualized. The posterior fossa was very deep but the middle fossa appeared quite normal. The orbital roofs had a more vertical direction than normal and the orbits were very shallow. The mastoid cells were poorly developed but the ethmoid cells were moderately enlarged.

An operation was performed on 2-21-44 through incision (coronal) with reflexion of the scalp to expose the vault of the skull. A ribbon of periosteum 1 cm. in width was removed from over the coronal suture and similar strip from over the saggital suture extending both anterior and posterior to the coronal suture for a distance of 5 cm. Then a gutter 0.75 cm. in width was ronguered in the bone to remove the site of the denuded coronal and saggital sutures. The patient tolerated the operative procedure well. On 3-11-44 an operation was performed through a coronal incision over the parietal bosses extending from one mastoid process to the other. Strips of bone and periosteum were removed paralleling the lamboid and remaining portion of the saggita; sutures.. Again the patient tolerated the procedure well and the post-operative course was uneventful. He was discharged from the hospital 11 days after the operation. Measurement of the exophthalmos before discharge revealed it to be 21 mm. on each side.

The patient was seen in the outpatient clinic on 7-18-44, four months after the second operation at which time he was more alert and active than he had been preoperatively. Roentgenographic examination of the skull showed some new bone formation along the edges of the operative defects but there were still wide defects and no bridges of bone. The head circumference was $49\frac{1}{2}$ cm., $5\frac{1}{2}$ cm. greater than it was at the time of operation. When seen again three months later the drooling had diminished, but otherwise there had been little change.

On 2-19-46 eleven months after operation his head circumference was 50 cm. and it appeared to be more normal in shape than it was before operation. Psychometric testing on 5-24-46, fourteen months after operation, revealed the boy to be of low normal intelligence, but the examiner felt that he was more nearly normal than he had been before operation. Roentgenologic examination of the skull on 9-11-46 revealed that the operative bony defects were stil present, and they did not seem to be filling in with new bone. The only change was a considerable increase in the size of the skull. On 11-8-46 the patient was again given the Stanford-Binet intelligence test which showed an I.Q. of 74. The Goodenough test showed an I.Q. of 71. These results would indicate that the patient's intelligence is "very low normal". The child is cooperative and is able to follow directions.

Case 3. ____ - This 28 year old female, white, is the mother of Case 2. Very little history could be obtained and no examination could be done, because the patient was very sen-

sitive about the deformity of her skull. She insisted that there was no abnormality in the shape of her head. The abnormality in the shape of the skull and a moderate exophthalmos had been present since early childhood. The patient had a rather typical cranial-facial dysostosis with a beak shaped nose, prognathous jaw, and crowded teeth. She did consent to have an X-ray of the skull taken and it showed a moderately severe scaphocephaly. No sutures could be seen, the basalor angle was increased, and the ethmoid sinuses were larger than normal.

Case No. 4

A 1900 gm. premature baby boy was delivered by Caesarean section on Nov. 9, 1944 after the mother had been hospitalized for three months because of rheumatic heart disease and subacute bacterial endocarditis. Routine examination of the newborn infant on Nov. 10, 1944 revealed that the fontanelles were open and soft. The anterior fontanelle was 1 cm. in diameter. The impression at this time was that this was a normal but premature male infant and he was discharged from the premature nursery on Dec. 14, 1944 when he weighed 2710 gms. The head circumference at this time was 36 cm. and the fontanelles were open and soft.

After his discharge from the hospital and when he was 8 weeks of age he came under the observation of Doctor Northrop Beach who noted that the infant was doliocephalic. The infant was seen by Dr. Beach on 3-5-45 and he noted that the head circumference was 39.5 cm. The fontanelles were closed and therefore synostosis cranii was thought to be developing. On 5-21-45 ($6\frac{1}{2}$ months) the head circumference was only 42 cm. and on 7-25-45 (8 months of age) the head circumference was 43 cm. and there was a very definite scaphocephaly. At this time the child could not yet sit up unassisted and because of this suggestion of developmental retardation he was again admitted to the hospital on 8-13-45.

The only significant physical findings at the time of admission were those related to the skull. There was a marked scaphocephaly, the head circumference was 44 cm. and the fontanelles were closed. Roentgenogram of the skull showed a synostosis of the saggital and coronal sutures. The lamdoid, basiphenoid and squamousal sutures were still open. The orbital roofs were more vertical in direction than normal. All other laboratory findings were within normal limits.

The child was operated on 9-5-45 when 10 months of age. A coronal incision was made and the scalp was reflected from the superior surface of the skull so that the saggital and coronal sutures were exposed. A strip of bone about 1 cm. in width was removed paralleling the saggital and coronal sutures. Strips of tantalum foil were interposed between the bony margins of these grooves The patient tolerated the operative procedure well. The postoperative course was uneventful and the patient was discharged from the hospital on 9-12-45.

The child was seen in the outpatient clinic on 3-5-46 six months after operation and at this time his development was apparently normal. The head circumference was 48 cm. (an increase of four centimeters) and, although the head was still scaphocephalic, its shape was more nearly normal than it had been previous to operation. The child was again seen in the outpatient clinic on 9-10-46, one year after operation and this time the head diameter was 49 cm., an increase of five centimeters since operation. There was a moderate scaphocephaly.

The patient was given a Stanford-Binet test (flrm L) on 10-14-46. This test indicated an I.Q. of 100. The Cattell test of infant intelligence given on the same day also showed an I.Q. of 100. The child was cooperative during the testing. His attention span was long and he followed directions well. Although intelligence tests given to children as young as this patient are not apt to be as valid as when they are given to older children, it seems that, on the basis of these tests in addition to observations of his general behavior, that this child has a normal mental development.

Case No. 5 - a 29 year old woman was first admitted to the hospital on 7-23-41 with a complaint of persistent severe frontal headaches. She had had headaches as long as she could remember, but in the few years just previous to her admission they had become much worse. In addition, she gave a history of having had an abnormally shaped head, bilateral exophthalmos, and poor vision since early childhood.

There was no history of a similar abnormality in any other member of the family.

Examinatiin revealed that the head was shortened in its anterio-posterior diameter. There was prominence of the bregma. There was a bilateral exophthalmos with an exophthalmometer reading of 23 mm. on the right and 21 mm. on the left. There was a divergent strabismus of 20. There was slight pallor of the optic disks, but visual acuity was only slightly below normal. The visual fields were normal. The palate was high and arched, but the teeth were normal.

Roentgenographic examination of the skull revealed a complete closure of all the sutures. The skull was short in its anterior posterior diameter and there was bulging in the region of the anterior fontanelle. The sella turcica was deep and moderately enlarged. There were digital markings suggestive of increased intracranial pressure.

On 7-28-41 a right myoplastic subtemporal decompressive craniectomy was performed. The patient's postoperative course was uneventful. She had relief of headaches on the right side following this procedure, but left sided headaches still persisted.

On 11-1-41 a similar subtemporal decompression was done on the left side. The postoperative course was complicated by a wound infection, and a localized osteomyelitis. Following this operation the patient continued to have occasional headaches, but they were much less severe and less frequent than they had been previous to operation.

Summary

The only satisfactory result following surgical intervention in these cases was Case No. 4. Although this child showed definite signs of mental retardation before operation, he has had satisfactory skull growth following the operation. He is symptom free and appears to be of normal intelligence. There appear to be two principal reasons for the success of this operation. First, he was operated upon early in life and second, it was possible to prevent regeneration of bone across the craniectomy defect by the use of tantalum foil.

Case No. 1 was probably improved by the operation but certainly the result can not be considered satisfactory. That the result was not completely satisfactory was due in large measure, we are sure, to the rapid regeneration of bone across the craniectomy defect.

The result in Case No. 2 similarly is far from satisfactory. This child was $3\frac{1}{2}$ years old and had signs of increased intracranial pressure when he was operated upon. Undoubtedly this child had already had a considerable amount of brain damage by this time so that a completely satisfactory result would be impossible. In this case it should have been possible to arrige at a correct diagnosis at a very early age because the mother showed typical signs of a craniofacial dysostosis. It seems quite possible that the marcellation procedure of King would be the operation of choice in this case rather than a linear craniectomy. In another similar case we would choose the marcellation operation because the skull is rigid at this age and a linear craniectomy may not allow for adequate expansion of the skull.

In Case No. 5 there was little to offer except the relief of her headaches. Subtemporal decompression seems to be an adequate procedure for this purpose. Some might prefer the marcellation operation because it allows for more expansion. The choice between these two procedures would seem to be largely one of personal preference after evaluation of the needs of the individual case.

References

1. Virchow,
Uber den cretinismus namentlich in franken, und uber pathologische schadelformen.
Verhandlungen d. Physik Med. Gesellschaft in Wurzburg. 2:230, 1851.

2. MacKenzie, W.
A practical treatise on the diseases of the eye.
Blanchard and Lea, Phila., p.116, 1855.

3. von Graefe, A.
Ueber Neuroretinitis und geivisse falle fulminirender erblindung,
von Graefe's Archiv., 12:114, 1866.

4. Lannelongue,
De la craniectomie dans la microcephalie.
Compt. rend.Acad.,d.sc.,110:1382, 1890.

5. Lane, L. C.
Pioneer craniectomy for relief of mental imbecility due to premature sutural closure and microcephalis,
J.A.M.A., 18:49, 1892.

6. Jacobi, A.
Address delivered before the eleventh international congress.
M.Rec., 45:609, 1844.

7. Scammon, R. E. and Dunn, H. L.
Empirical formulae for the postnatal growth of the human brain and its major divisions.
Proc.Soc.Exper.Biol.and Med.,
22:114, '22.

8. Giblin, N. and Alley, A.
A method of measuring bone growth in the skull.
Anat.Rec., 83:381, '42.

9. Idem., Studies in skull growth.
Coronal suture fixation.
Anat.Rec., 88:143, '44.

10. Bolk, L.
On the premature obliteration of sutures in the human skull.
Am.J.Anat., 17:495, '15.

11. Greig, D. M.
Oxycephaly.
Edinburgh M.J., 33:189, '26.

12. Sear, H. R.
Some notes on craniostenosis.
Brit.J.Radiol., 10:445, '37.

13. Bertolotti,
Le syndrome oxycephalique on syndrome craniosynostose pathologique.
Press med. 1:332, '14.

14. Young, F. W.
Acrocephaly.
Arch.Pediat.,39:629, '22.

15. Park, E. A. and Powers, G. F.
Acrocephaly and scaphocephaly with symmetrically distributed malformations of the extremities.
Am.J.Dis.Child, 20:235, '20.

16. Ogilive, A. G. and Posel, M. M.
Scaphocephaly, oxycephaly and hypertelorism.
Arch.Dis.Child.,2:146, '27.

17. Mann, I.
A theory of the embryology of oxycephaly.
Tr.ophth.Soc.U.Kingdom, 55:279,'35.

18. Marselli, M.
Sur la scaphacephalie.
Bull.de la Soc.d'Anthropologie de Paris, 10:445, 1875.

19. Rieping, A.
Zur Pathogenese des Turmschadels.
Deutsche Ztschr.f.Chir.,148:1,'19.

20. Bedell, A. J.
Oxycephalus.
J.A.M.A.,68:1979,'17.

21. Carpenter, G.
Specimen of oxycephaly.
Tr.Ophth.Soc.U.Kingdom, 29:157,'09.

22. Carr, A. D.
Oxycephaly.
J.Missouri M.A., 26:398, '29.

23. Crouzon, M. O.
Dysostose cranio-faciale heredetaire,
Arch.de med.d.enf., 18:529, '15.

@$. Fletcher, H. M.
On oxycephaly.
Quart.J.Med.,4:385, '10.

25. Klein, O. L. and Childe, A. E.
Oxycephaly with the report of two cases in a brother and sister.
Canad.M.A.J. 34:397, '36.

26. Sheldon, W.
Hereditary and familial oxycephaly.
Proc.Ray.Soc.Med., 24:574, '31.

27. Skipper, E.
Oxycephaly - with report of three cases in one family.
Quart.J.Med., 3:579, '34.

28. von Vershuer, O.
Die vererbungsbiologische zwillingsfarschung,
Ergebn. D.Inn.Med.U.Kinderh., 31:35, '27.

29. Gunther, H.
Der Turmschadel als Konstitutionsanomalie und als klinischer Symptom.
Ergbn.d.Inn.Med.u.Kinderh., 40:40,'37.

30. Scharpe, W.
The cranial deformity of oxycephaly; its operative treatment with a report of cases.
Am.J.M d. Sc., 151:840, '16.

31. Greig, D. M.
Hypertelorism
Edinburgh.M.J., 31:560, '24.

32. Braithwaite, J. V. C.
The relation of hypertelorism to mongolism.
Arch.Dis. Child.,1:369, '26.

33. Wheaton, S. W.
Two specimens of congenital cranial deformity in infants associated with fusion of the fingers and toes.
Tran.Path.Soc.London, 45:238, 1894.

34. Apert, E.
De L'acrocephalosyndactylie.
Bull.er mem.Soc.med.d.hop.de Paris, 23: 1310, '06.

35. Davis, B. F.
Acrocephalosyndactylism.
Am.J.Dis.Child.,9:446, '15.

36. Schloffer, H.
Ueber die Grundlagen und methoden der operativen behandlung der sehstaryngen beim turmschadel.
Beitrage z. Klin. Chir., 86:265,'13.

37. Van der Hoeve, J.
Roentgenography of the optic foramen in tumors and diseases of the optic nerve.
Am.J.Ophth.,8:101, '25.

38. Behr,
Changes in the region of the asseous canal in steeple skull.
Arch.Ophth., 13:926, '35.

39. Bequmont, W. M.
Oxycephaly.
Tr.Ophth.Soc.U.Kingdom, 30:44, '10.

40. Bennett, A. E., Keegan, J. J., and Hunt, H. B.
Oxycephalus: Premature synostosis of the cranial sutures - prevention of blindness by craniectomy and decompression.
J.Nerv.and Ment.Dis., 84:274, '36.

41. Faber, H. K and Towne, E. B.
Early craniectomy as a preventive measure in oxycephaly and allied conditions.
Am.J.Med.Sc.,173:701, '27.

42. Idem, Faber, H. K. and Towne, E. B.
Early operation in premature cranial synostosis for the prevention of blindness and other sequelae.
J.Pediat.,22:286, '43.

43. Dorfmann, R.
Uber Pathogenese und Therapie des Turmschadels.
Arch.F.ophth., 68:412, '08.

44. Watts, S. H.
Oxycephaly: report of two cases.
Ann.Surg.,71:113, '20.

45. Hildebrand, O.
Eine neue operationsmethode zur Behandlung der durch Turmschadel bedingten Schnervenatrophie.
Arch.f.klin.Chir.,124:199, '23.

46. Elschnig, A.
Resektion des Canalis Opticus bei Turmschadel.
Med.Klin.,20:1281, '24.

47. Bauer, K. H.
Die zirkulare Kraniotomie als Entlastungstrepanation bei drahender Turmschadelerblindung und bei nicht-lokalisierbaren Hirngeschwulsten.
Deutsch.Ztschr.f.Chir.,237:204, '32.

48. King, J. E. J.
Oxycephaly - a new operation and its results.
Arch.Neurol.and Psychiat.,40:1205,'38.

49. Idem.
Osycephaly.
N.Y.State J.M d.,49:2119,'41.

50. Idem.
Oxycephaly: Tr.South.S.A.,54:8,'41.

51. Woodhall, B.
Oxycephaly - results of treatment by the King "morcellation" method.
J.Pediat.,20:585, '42.

52. Dandy, W. E.
An operation for scaphocephaly.
Arch.Surg.,47:246, '43.

53. Brai, A.
Oxycephaly and optic atrophy.
Ann.Ophth.,21:1, '12.

III. GOSSIP

Commercial radio enjoyed unusual popularity during the war because of the timeliness of its news bulletins. The public appreciated reports from around the world but with peace, the character of the news changed, and the balance of the programs on the radio began to suffer by comparison. Commercial radio had tasted high incomes in recent years as advertisers lavished money upon it, for it was either giving it to them or turning it over to the government. During the past year, advertisers have started to use the printed page, and radio is suffering from dwindling incomes. This has caused commercial radio to turn to the psychologists for help in an attempt to gauge public opinion and to rememdy the situation. But the psychologists are trying to do a job of public relations through convincing the public that radio knows best and the public really likes the commercials and they should be grateful to radio for its public service features. Local radio broadcasts are subject to the whims of the chains as local station managers are unable to guarantee uninterrupted schedules to local sustaining or commercial accounts. The future of local broadcasts is at stake for there is a definite advantage which is lost by constant change of times, just to make another commercial possible. The public is getting fed up with the situation and radio will have to clean house or else. But apparently chain management is more concerned with the ready dollar which they can pick up at this time, than in a long time program of public service. Advertising has grown more objectionable as unwarranted claims occupy more and more of radio time. ...A good friend of mine was visited by a representative of the telephone company not long ago in the interest of their program to raise their rates. My friend felt their rates should not be raised in view of the poor service they were rendering. He calculated how much he had spent over the years in telephone charges, and he still had his old style telephone and not the modern French type. The company representative explained that because of shortages, etc., they were not able to get the new type, but my friend found they had been giving them to new subscribers and not old. The upshot of the whole thing was my friend got his new telephone, and the public relations department of the telephone company is nursing its wounds from failure of its sure-fire technique to win a supporter to their side...I tried a new recording machine the other day in which the voice is registered on a strip of paper dusted with a chemical compound. Reproduction was excellent. When you are through with the recording, it can be wiped off with a rag and used over again. Paper discs also are available for correspondence. Record your letter, fold the paper, put it in an envelop and mail it. The only joker is that a special recording device is needed at the other end....One day follows the other in rapid succession as the winter quarter gets under way. Enrolment is off a few hundred students, but it is so close to 27,000 that little difference can be noted on the campus. The temporary wooden buildings are arriving. The first to be placed is just south of Folwell Hall. They are coming from the Navy Base and we will have several near the hospital in due time. There is nothing quite so permanent as a temporary structure, and years from now, they probably will still be here....Everyone is hopeful the Mayo Memorial Building will receive the additional funds needed to start work on the structure. Medical education is concerned with training, service, and research. More physicians are needed, but building new schools is costly, and supporting them requires more funds than most people appreciate. Average school needs a budget of half a million dollars a year to operate, and this does not permit use of these funds for hospital beds and patient care. There is plenty of money for support of research projects but practically none for buildings in which to carry on this activity. Large sums of money will be collected in the next few years for special projects such as polio, crippling in childhood, eye defects, hearing loss, cancer, heart disease, leukemia, disseminated sclerosis, arthritis, and many others. Will the medical schools be able to handle so many different kinds of research and instruct students at the same time?.... I wrote a column the other day on clinical research in which I followed the American Medical Associations Council on Pharmacy and Chemistry in asking for more support for investigation of drugs and treatments of patients.....