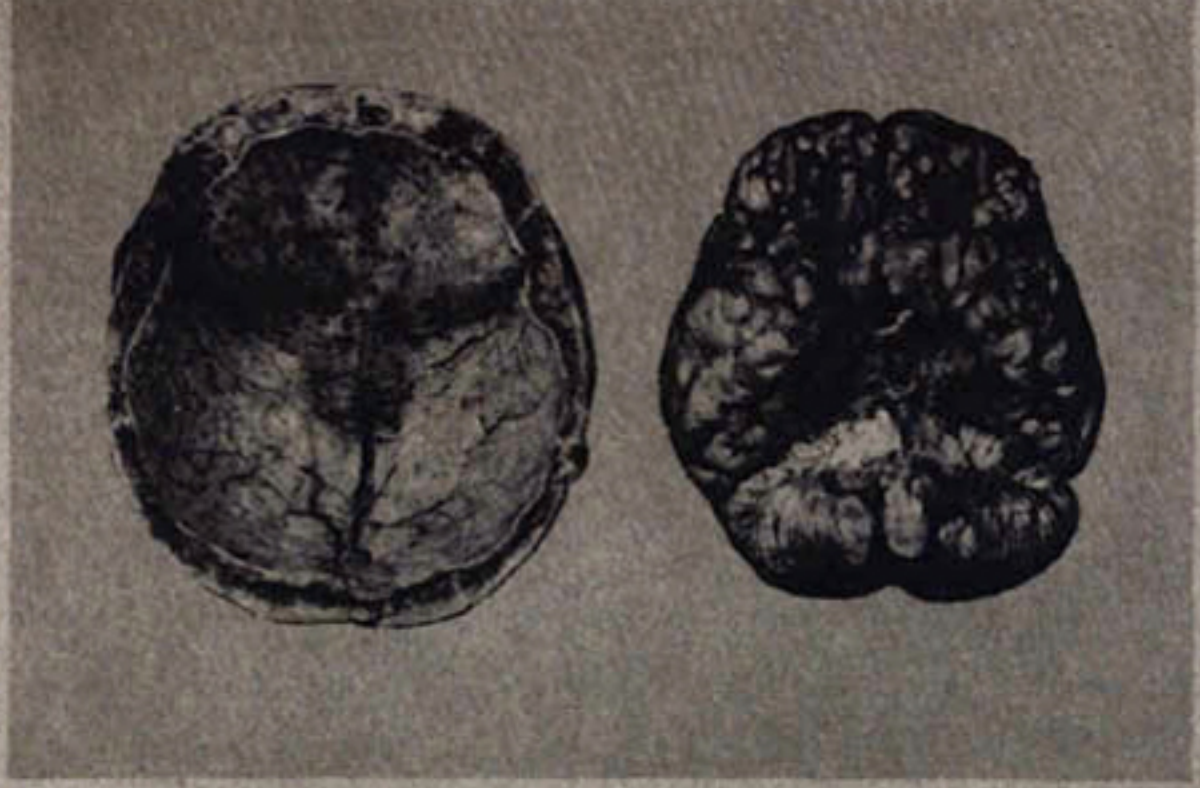

CRANIAL HYPEROSTOSIS

(HYPEROSTOSIS CRANII OR H.C.)

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With a Foreword by

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Director of Research
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Chicago, Illinois

For pathologists and for all physicians with an interest in the human cranium or human brain -- the first original and complete account in English of the pathology of cranial hyperostosis.

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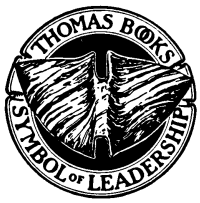

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To my father, my mother and my wife.

FOREWORD

I AM VERY pleased to write a foreword to this careful and scholarly study. After Bayle demonstrated that general paralysis of the insane was always accompanied by a chronic meningitis it was thought that the causes of all types of mental disturbances might be discovered by study of their pathological anatomy and it was considered indispensable that there be a pathologist on the staff of every mental hospital. This hope was not realized and the resultant pessimism plus the rise of equally illogical faith in psychogenetic theories has led to the feeling that the pathologist in a mental hospital, except for the routine clinical examinations of blood, urine and feces, is practically superfluous. This feeling was reinforced by the neglect of pathological anatomy in general pathology in favor of experimental studies. Pathological anatomy has come to be old-fashioned and the younger pathologists are loath to cultivate it. Nevertheless there remains much to be clarified by anatomical studies, especially in the field of mental deficiency, as this monograph demonstrates. For this reason I am especially pleased that a pathologist of the competence and experience of Dr. Perou is willing to undertake such a detailed, careful and exhaustive study of a neglected problem.

This monograph will appeal to many people because of its thorough documentation, its extensive analysis of the related literature and its beautiful illustrations. Not only will those who are interested in mental deficiency consult it but also students of the pathology of bone, roentgenologists, brain surgeons, orthopedic surgeons and all those who have to deal with diseases of the head. These meticulous descriptive studies are indispensable to all such people however old-fashioned they are felt to be. It is still necessary to establish accurate diagnoses on the basis of clearly distinguished clinical and anatomo-pathological entities.

Dr. Perou has shown also that it is possible to do original work along these lines. Perhaps the most useful feature of this study is

the clear distinction he draws between Hyperostosis Calvariae Interna and idiopathic Hyperostosis Calvariae Diffusa, the former being a specific entity characterized by an inherited imperfection of the bony tissue of the cranium, the latter being the end-product of several pathological processes. But this is not only a descriptive anatomical study; explanation of the pathological processes is given where possible even though often conjectural.

Although use is made of the studies of his predecessors the personal experience and extensive material studied by the author is evident from the tables. It is comforting to know that our mental institutions are being used not only to care for our unfortunate children but also to advance our knowledge of the maladies from which they suffer. It is to be hoped that this splendid study by Dr. Perou is but the beginning.

PERCIVAL BAILEY, M.D.

INTRODUCTION

ANCIENT medicine paid great attention to the skull and the term Phrenology was coined to characterize the study of the mind and character in relation to the shape of the skull. Phrenology has long ago been discredited and is now a dead science. However, the study of the cranium remains a fascinating and yet neglected subject. Anthropologists and anatomists have shown greater interest in the human cranium than physicians as a whole, with the exception of radiologists. Even neurologists, psychiatrists and neurosurgeons have shown only casual interest in the study of the cranium, which they treat more as an obstacle thrown around the brain than as an entity, the function of which is to lodge and protect the brain. Pathologists, with only a few exceptions, have greatly underestimated the human cranium. In countless laboratories and departments of pathology throughout the world, the emphasis is on the brain, always. The opening of the cranium and, for that matter its examination, are often left to morgue attendants, medical students and apprentice physicians.

The present study was started simply because this observer could not recognize and classify to his satisfaction the numerous cranial lesions and anomalies encountered during the routine performance of autopsies at the Dixon State School, for mentally retarded children and adults. At first it covered the cranium as a whole. However it was deemed necessary, for the time being, to limit it to the problem of hyperostosis or thickening of the human cranium, which is the most common and most misunderstood condition of this structure. A preliminary report of this work was presented at the joint annual meeting of the American Society of Clinical Pathologists and College of American Pathologists (Chicago, Illinois, October, 1962).

It is paradoxical that little is known concerning a subject about which so much has been written. Over the years, starting with the important work of Morgagni and continuing with the outstanding monographs of Beadles, Naito, Morel, Henschen, Campos, Calame

and Moore, there have been excellent discussions of the cranial hyperostoses. For some unknown reason much of the discussion has been centered on one specific type of cranial hyperostosis, namely the so-called "Morgagni-Stewart-Morel Syndrome," with some attention being given also to the hyperostosis of the anemias of childhood. Only Naito considered the cranial hyperostoses as manifestations of multiple disease processes and based his monograph on these premises. Subsequently, Schüller and Babäiantz made brief and comprehensive reviews of the cranial hyperostoses, mostly from the clinical and radiological viewpoints.

This monograph follows in the footsteps of Naito's original work. Its purpose is to try to bring again into focus, in the light and context of general pathology, a subject which has remained both obscure and complex. It is simply a *mise-au-point* which is both a follow-up and an attempt at rehabilitation.

It is primarily an anatomical study, and its ultimate purpose is to define, classify, describe, explain and, if possible, clarify, the different varieties of cranial hyperostosis. Emphasis is placed on the so-called "idiopathic" varieties of calvarial hyperostosis. Statistical data are kept to a minimum. Likewise, clinical and radiological data are sparingly used. Explanation of pathological processes is attempted and, of necessity, is often conjectural.

In order to facilitate comprehension of the text, a short summary of essential techniques and anatomical data as well as a glossary are submitted. The pompous latin words used here and throughout the text are now—unhappily—part of the jargon which has become the trademark of anyone writing on the subject of hyperostosis of the cranium, better known as "Hyperostosis Cranii" or "H.C."

Finally, the author wishes to apologize for a rather free use of many classical textbooks and treatises. This was necessary, particularly for the brief summing up of certain rare diseases and conditions, some of which he has yet to encounter.

Some might rightly criticize this text as lacking in originality and would have preferred to have it shortened by sacrificing all the already well-known anatomical and pathological data. However, the gain in conciseness would probably not balance the loss in completeness and intelligibility. The author's own material

forms the backbone of this communication. It is his sincere hope that, by blending his own observations with those of his predecessors, he will perhaps help bring to the cranial hyperostoses the modest recognition that they well deserve.

Dixon, Illinois MAURICE L. PEROU, M.D.

ACKNOWLEDGMENT

I GRATEFULLY acknowledge the generous assistance and kind advice of my friends and colleagues. They gave so willingly of their time and knowledge that I will always be indebted to them. I cannot possibly thank everyone in particular, but would like to give special recognition to those whose help and cooperation made this study possible.

Drs. Percival Bailey, Julius Kolis, Saul Parks, Thomas Flynn and Catherine Haberland made invaluable suggestions during the preparation of this monograph. I am most indebted to Dr. Percival Bailey for his unfailing moral support, patience and understanding. Drs. Julius Kolis and Saul Parks also helped with the radiological interpretations and provided a number of cases of cranial hyperostosis, some of which are included in this report. To them, I am also greatly indebted.

Drs. Ernest Aegerter and Paul Bucy not only made pertinent and useful suggestions, but also were generous enough to authorize the incorporation in this publication of some of their own material. For this, I am very grateful.

Drs. John Lindsay and Raul Hinojosa were most helpful in granting me free use of the extensive material of the "Temporal Bone Banks Center" of the University of Chicago. Dr. Hinojosa was particularly generous of his time and without his help the paragraph of Otosclerosis could not have been written. However, he might be relieved to know that I am taking full responsibility for the final text.

Dr. Gerritt Bras graciously opened to me the doors of the pathological museum of the University College of the West Indies, Kingston, Jamaica. He also kindly furnished two photographs of his valuable specimens for use in this monograph.

Dr. Norman Johnson helped considerably in matters dealing with polarized light microscopy.

Dr. John Levitsky gave invaluable criticism pertinent to the genetic aspect of cranial hyperostosis.

Drs. Orville Bailey, Otto Saphir and Jacob Shanberge, Cecil Krakower and Ruth Wong, generously gave me access to the pathological files of the Illinois Neuro-psychiatric Institute, the Michael Reese Hospital and the University of Illinois Hospitals.

Norma Conrad and Mildred Tress shared the burden of preparing the bone sections and gave selflessly of their time. My deepest appreciation goes to them.

Dr. Frank Wagner, Mrs. Frances Rink, Mr. Hershel McKee, Mr. Harold Nelson and Mrs. Sue Wise gave useful assistance in many ways. The selfless, patient and competent cooperation of Mrs. Pat Mangler, who typed and retyped the manuscript so many times, deserves special praise and special thanks.

To Mr. Zalkin Mandelstam goes most of the credit for the excellent photographic work. Mr. Bert Nordgren helped also with the photographs of the patients. A few unworthy photographs, taken by me, will be found easily in the text. They are kept only because each illustrates an important lesion, a better likeness of which I do not have. Several x-ray photographs were made by the Hospital Picture Service, Red Bank, New Jersey.

This study was supported by "Mental Health Fund No. 1805" and I am most indebted to the Department of Mental Health for its generosity. I specifically would like to express my deep appreciation to Drs. Harold Visotsky, Percival Bailey, V. Karr McKee and Paul Tillman, as well as to Mr. Leon White and Mr. David Edelson from the Department of Mental Health and the Dixon State School. Without their selfless cooperation and generosity this monograph could not have been published.

I wish also to express my gratitude and appreciation to the medical staffs of the Dixon State School, the Community General Hospital of Sterling and the Morrison Community Hospital. Their cooperation was heart-warming.

I am indebted to the following publishers for authorizing the use in this publication of the figures stated below:

W. B. Saunders Company: Figures 2 and 38.

Charles C Thomas, Publisher: Figure 54.

Finally, to my publishers, particularly to Mr. Payne Thomas, I am thankful for the utmost courtesy and for the invaluable help given in the preparation of this monograph.

M.L.P.

CONTENTS

CRANIAL HYPEROSTOSIS

(Hyperostosis Cranii or H.C.)

MATERIAL AND METHODS

IT IS NOT within the scope of this monograph to give a detailed abstract of each individual case. Rather, the aggregate cases (Table I), will be used for interpretation and illustration of the total pathological picture of the cranial hyperostoses. These cases were collected over a six year span from a total of 1,010 necropsies performed at the Dixon State School, Dixon, Illinois; the Community General Hospital of Sterling, Illinois and the Morrison Community Hospital, Morrison, Illinois. However, this project was not started in earnest until the early part of 1961; cases of cranial hyperostosis were systematically collected from this date. When needed, comparative studies of the autopsy material were done with live patients of the Dixon State School, which has a population of about 5,000 inmates. Of these live patients, 175 with idiopathic cranial hyperostosis detected by routine x-ray examination of the skull are being studied. In twenty-eight cases done in the early part of this study—including ten with idiopathic cranial hyperostosis—the parathyroids, other bones and, occasionally, the gonads were not available for microscopic study. In all the other cases a complete and thorough autopsy was performed, including removal and microscopic examination of all the endocrine glands and several other bones of the body, namely, ribs and vertebrae. The brain, in all instances, was examined at the Illinois State Psychiatric Institute by various neuropathologists, namely Drs. Ben W. Lichtenstein, Percival Bailey, and Catherine Haberland. They were also studied by the author, who performed all the postmortem examinations, except one.

Technique of Skull Opening and Closure: A simple technique of skull opening and closure was devised in connection with this study. The five basic purposes of this technique are: to arrive at a standard skull cap, to permit easy removal and replacement of the latter, as well as good exposure of the cranial contents, to ensure secure closure of the cranial box and finally, to allow the

TABLE I

List of Cases

	Disease	*Author*	*Autopsy*	*Clinical*	*Total*
I.	Osteopetrosis	Aegerter	0	1	1
	Osteogenesis imperfecta	Perou	0	3	3
II.	Tori, occipital and frontal	Perou	2	3	5
	Occipital spur	Perou	3	4	7
	Oxycephaly	Perou	0	5	5
	Scaphocephaly	Perou	0	2	2
	Plagiocephaly	Perou	10	0	10
	Trigonocephaly	Perou	0	1	1
	Microcephaly vera	Perou	10	0	10
	Secondary microcephaly	Perou	16	0	16
	Hemi-microcephaly	Perou	15	5	20
	Hydrocephaly	Perou	33	0	33
III.	Syphilis	Bras	1	0	1
	Non-specific infection	Perou	1	4	5
IV.	Erythroblastic anemia	Perou	1	7	8
	Spherocytic anemia	Perou	0	2	2
	Sickle cell anemia	Perou	4	8	12
	Others	Perou	4	0	4
V.	Cephalhematoma and variants	Perou	5	7	12
	Fracture	Perou	6	6	12
VI.	Scurvy	Perou	0	2	2
VII.	Acromegaly	Perou	3	2	5
	Fröhlich's Syndrome	Perou	2	0	2
	Pregnancy	Perou	1	0	1
VIII.	H.C.I.	Perou	25	85	110
	H.C.D. (idiopathic)	Perou	32	90	122
	Otosclerosis	Lindsay and Hinojosa	20	0	20
	Paget's Disease	Perou	0	2	2
		Aegerter	1	0	1
		Saphir and Shanberge	2	0	2
	Fibrous dysplasia	Perou	0	6	6
IX.	Meningioma	Perou	2	3	5
		Bras	1	0	1
	Breast carcinoma	Perou	3	0	3
	Prostatic carcinoma	Perou	1	0	1
	Cylindroma	Perou	1	0	1
	Osteoma	Perou	15	0	15
	Hemangioma	Bucy and Capp	1	0	1
	Reticulum cell sarcoma	Bucy and Ullrich	1	0	1
X.	Storage disease	Perou	6	0	6
	Meningeal ossification	Perou	18	0	18
	Wormian bone	Perou	30	10	40

Total Autopsy Cases: 276 Author: 249 Others: 27
Total Clinical Cases: 258 Author: 257 Others: 1
Total autopsy cases pertinent to true hyperostosis: 222; Author: 195; Other: 27
Author cases of true hyperostosis: 154*
Total autopsy cases of Pseudo-hyperostosis: 54; Author: same

CONTROL CASES

Name	*Author*	*Autopsy*	*Clinical*	*Total*
Various convulsive disorders without hyperostosis	Perou	55	0	55
Mongolism	Perou	38	0	38
Leukemia	Perou	10	0	10
Multiple myeloma	Perou	1	1	2
Normal	Perou	230	100	330

Total Control Autopsy Cases: 334 Normal: 230 Abnormal: 104
Total Control Clinical Cases: 101 Normal: 100 Abnormal: 1

*Balance of forty-one cases represent instances where hyperostosis, although theoretically expected, was not found.

prosector to remove and keep part or parts of the cranium for special studies.

A circular cut is made, starting 2 to 2½ cms. above the supraorbital arches, running laterally through the midportion of the temporal muscles and posteriorly just above the external occipital protuberance (Fig. 1). Comparison between this technique and two other well-known techniques[1, 2] is outlined in the drawing (see Fig. 1). The proposed technique offers the following advantages: the line of removal of the skull cap corresponds to fixed and easily recognized anatomical landmarks; a standard skull cap corresponding closely to the calvaria itself is obtained; standard measurements can be taken pertinent to weight of the skull cap, thickness of calvarial bones and measurements of the head diameters; the greater part of the squama of the frontal bone, the site of various fairly common and important pathological changes is removed; finally, the frontal sinuses can be easily examined.

For closing the skull, the skull cap is replaced in its original position and secured by interrupted or uninterrupted sutures running through the temporal muscles and adjacent fascia. A fresh, moistened, plaster-bandage, preferably three inches wide, is passed

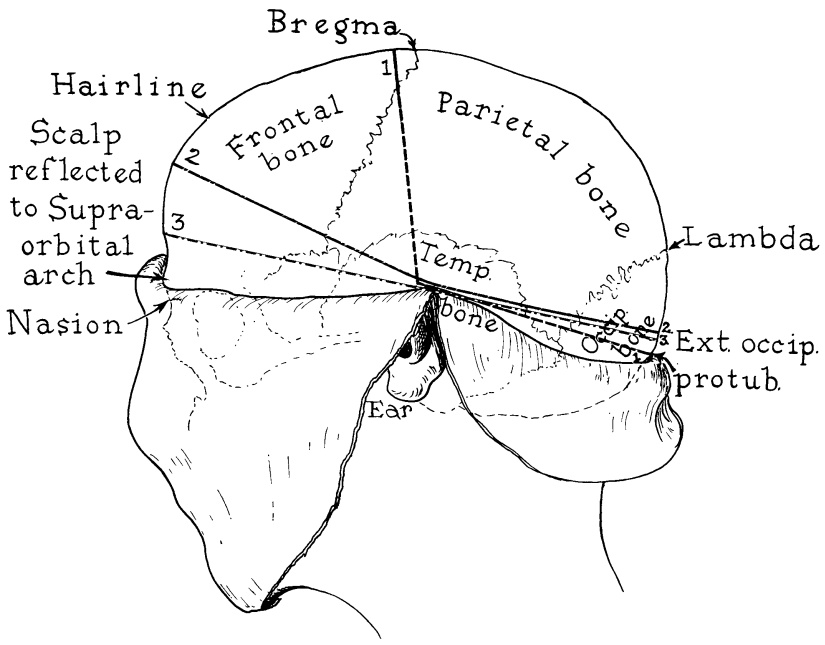

Fig. 1. Lines of skull opening. 1) From *A Manual for Pathologists; The Technic of Necropsy.*[1] 2) From: *Autopsy Diagnosis and Technic.*[2] 3) Author.

around the lines of sawing three or four times. It is allowed to dry and subsequently the scalp is closed in the ordinary fashion. When large chunks of calvarial bones or even the entire calvaria are removed and kept for special scientific studies, they can be easily replaced by an appropriate plaster of Paris mold.

Technique of Bone Preparation:

1. Decalcification in 5 per cent nitric acid for about three to four days. This process was verified for the larger and harder specimens by x-ray and/or chemical means.
2. Celloidin embedding. Duration: 2 to 2½ months.
3. Sectioning of block at about 14 microns.
4. Staining with hematoxylin and eosin using the flotation technique.
5. Mounting of sections in thickened synthetic mounting medium.
6. Flattening of sections with weights of lead or mercury. Duration: two to three days.

Additional Techniques: In twenty-five cases, bone pieces were ground on sandpaper to a thickness of about 30 microns. For the same number of cases, unstained but decalcified bone sections, prepared according to the above-described technique, were also made.

Examination of Sections: All hematoxylin and eosin stained sections were studied with a standard microscope, both in ordinary light and in polarized light. The ground sections and the unstained preparations were also studied in polarized light. For the purpose of this study, enough valuable information was gained by simply studying the hematoxylin and eosin sections in polarized light. Not only the architecture of the bone and the bone matrix could be well visualized, but the sections were cleaner and thinner than with the other preparations. There was some color variation, mostly in the background, due to the quality and amount of the stains used. However, this was not found to be a serious handicap.

ESSENTIAL ANATOMICAL DATA—NOMENCLATURE AND ABBREVIATIONS

ANATOMICAL DATA

IN ORDER to better evaluate the abnormal and borderline, a thorough knowledge of the normal is necessary. The following paragraph consists of a succinct summary of the data obtained partly from the literature but primarily from the study of normal skulls done in connection with this monograph. Normal and abnormal values are listed in Table II.

There are great individual and probably also racial, variations as to size, shape and weight of the skull. The calvaria is in general thicker in the midline where there is less pressure from the brain and thinner on the sides where there is more pressure from the brain. Consequently, the mid-frontal, mid-parietal and mid-occipital regions are generally thicker than their lateral counterpart. The thickest portion of the skull is often the external occipital protuberance. In this region a thickness of 15 mm. is not unusual and entirely normal. Likewise, the mid-frontal and mid-parietal regions may measure up to 9 mm. and 9.5 mm. respectively.

These figures were obtained by studying the skulls of average adults of an average mid-western United States population. The majority of the patients were white.

The bones of the calvaria have three layers: an external layer or outer table, a mid-layer or diploe, and an internal layer or inner table. The diploe is generally twice as thick as the inner or outer table and generally equals the thickness of these two layers together. The outer table is generally slightly thicker than the inner table. The diploe is not present at birth. It appears generally at the seventh year of life and, occasionally, is not well developed until puberty. In the adult the diploe is reddish and spongious; with advancing age it may lose some of its reddish tinge and be-

TABLE II

		Normal Values*			Abnormal Values	
		Min.	*Max.*	*Mean*		
1) Head circumference	Male	54 cms.	60 cms.	57 cms.	< 54 - small	> 60 = large
	Female	51 cms.	58.5 cms.	54.75 cms.	< 51 = small	> 58.5 = large
2) Diameters:						
Antero-posterior (AP)	Male	16.5 cms.	19.5 cms.	18 cms.	< 16.5 = short	> 19.5 = long
Biparietal (BP)	Male	13.5 cms.	15.5 cms.	14 cms.	< 13.5 = narrow	> 15.5 = broad
Antero-posterior (AP)	Female	14.5 cms.	18 cms.	16.25 cms.	< 14.5 = short	> 18 = long
Biparietal (BP)	Female	12.5 cms.	14.5 cms.	13 cms.	< 12.5 = narrow	> 14.5 - broad
3) Weight of skull cap	Male	350 gms.	525 gms.	437.5 gms.	<350 = light	>525 = heavy
	Female	300 gms.	450 gms.	375 gms.	<300 = light	>450 = heavy
4) Skull thickness:						
Frontal bone	Middle	6 mm.	9 mm.	7.5 mm.	> 9 mm. - hyperostosis	
	Lateral	3 mm.	7 mm.	5 mm.	> 7 mm. - hyperostosis	
Parietal bone	Vertex	5 mm.	9.5 mm.	7.25 mm.	> 9.5 mm. = hyperostosis	
	Lateral	3 mm.	8 mm.	5.50 mm.	> 8 mm. hyperostosis	
Occipital bone	Middle	7 mm.	9.5 mm.	8.25 mm.	> 9.5 mm. = hyperostosis	
	Lateral	5 mm.	8 mm.	6.50 mm.	> 8 mm. = hyperostosis	
Temporal bone	Squama	1.5 mm.	5 mm.	3.25 mm.	> 5 mm. = hyperostosis	
Orbital plate		1 mm.	2.5 mm.	1.75 mm.	> 2.5 mm. = hyperostosis	
					4- 7 mm. = Average calvarial thickness	
					7- 9 mm. = 1+ hyperostosis	
					9-11 mm. 2+ hyperostosis	
					11-13 mm. = 3+ hyperostosis	
					>13 mm. - 4+ hyperostosis	

* Because of the great individual variation in cranial measurements, rough figures are given preference over mathematically accurate but meaningless ones.

come somewhat yellow as a result of fatty degeneration of the marrow. The inner and outer tables are whitish-gray or whitish-yellow bands of compact bone. The bones of the vault of the skull arise chiefly from the membranous cranium while those of the base arise mainly from the chondrocranium and are preformed in cartilage.

NOMENCLATURE

Skull. The skull is the skeleton of the head and face. It comprises the cranium and the bones of the face.

Cranium. The cranium is the brain case. It is composed of eight bones whose function is to contain and protect the brain.

Calvaria or Calvarium. The calvaria is that portion of the cranium which is situated above the supra-orbital ridges anteriorly and the superior nuchal lines of the occipital bone posteriorly.

Skull Cap. It is that portion of calvaria removed at autopsy following a circumferential line starting 2 to 2½ cm. above the supra-orbital ridges, running laterally through the mid-portion of the temporal muscles and posteriorly just above the external occipital protuberance (Fig. 1).

Hyperostosis (Hyper = over, osteon = bone, and osis = condition). The simplest definition of hyperostosis is: *an increase or excess of bone.* On that basis it is analogous to hypercementosis, hyperadiposis, hypertrichosis, etc. The prefix "hyper" means "in excess of, increase of, overgrowth or overproduction of" and not "on top of." This is why the osseous overgrowth may be within as well as over and under a given bone. This definition encompasses not only increase in thickness, but also increase in surface or area of a given bone. However, as applied to the skull, the word hyperostosis has generally a more limited sense. It means: *an indigenous increase, overproduction or overgrowth of bony tissue causing increase in volume or thickening of one or many skull bones.*

Dysplasia (Dys = bad and plasia = molding). Literally, defective molding or development. Speaking of the skull, it means in its broad sense, *abnormal development* and in a restricted sense, *abnormal tissue development,* generally with emphasis on proliferation rather than degeneration. On that basis from the histopathologic viewpoint, a dysplastic process is primarily and often

a hyperplastic one. Calvarial dysplasia is then analogous to mammary dyplasia or fibrous dysplasia of bone.

Dystrophy (Dys = bad and trophe = nourishment). Literally, defective nutrition resulting in defective development. Speaking of the skull, it means simply *abnormal development* and specifically *abnormal tissue development,* with emphasis on degeneration. On that basis a dystrophic process is primarily and often a degenerative one. Calvarial dystrophy has the same meaning as muscular dystrophy. A perusal of the literature reveals that the words dysplasia and dystrophy are often used interchangeably, just as hyperplasia and hypertrophy. However, at present there is greater unanimity of opinion concerning the meaning of hyperplasia and hypertrophy than there is concerning that of dysplasia and dystrophy. Nevertheless an attempt is made to differentiate both processes on the basis of etymology and usage.

ABBREVIATIONS

H.C.	=	Hyperostosis Cranii
H.C. Syndrome	=	Hyperostosis Cranii Syndrome
H.C.I.	=	Hyperostosis Calvariae Interna
H.Cr.I.	=	Hyperostosis Cranii Interna
Hfi	=	Hyperostosis frontalis interna
Hpi	=	Hyperostosis parietalis interna
Hfpi	=	Hyperostosis fronto-parietalis interna
Hfti	=	Hyperostosis fronto-temporalis interna
Hfpoi	=	Hyperostosis fronto-parieto-occipitalis interna
H.C.D.	=	Hyperostosis Calvariae Diffusa
H.Cr.D.	=	Hyperostosis Cranii Diffusa
Hfd	=	Hyperostosis frontalis diffusa
Hfpd	=	Hyperostosis fronto-parietalis diffusa

These abbreviations will be used throughout the main text. Specifically, except on occasion, cranial hyperostosis will be henceforth referred to as simply H.C.

CLASSIFICATION

THE FOLLOWING classification of the cranial hyperostoses is offered (Table III). It is partly based on the classifications of Naito,[3] Schüller,[4] and Babäiantz,[5] but primarily follows the classical outlines of general pathology.

According to the previously given definition of hyperostosis, the degenerative hyperostoses which often are osteoporotic or "hypo-ostotic," should not be included in this study. However, they are discussed here and are kept in the hyperostotic group for several reasons. Not only are they closely linked with this group, but they also cause the most severe calvarial thickening and, although predominantly degenerative and osteoporotic, are fairly often either focally hyperostotic or have gone through transitory hyperostotic phases. Finally, the marrow, which in these cases goes also through phases of hyperactivity or hyperplasia, is an intrinsic part of the diploe, that is, of the bone. This leads us to the hyperostosis of the anemias of childhood and their proper classification. Many of them could be classified with the hereditary hyperostoses or the dystrophic hyperostoses but, because of their distinct features and because of the lack of agreement on their pathogenesis, they are placed in a class of their own.

Physiological congenital overgrowths such as occipital spur and elongated styloid process are briefly considered here because they do represent "excess bone" and fall in the broad category of hyperostosis.

Otosclerosis has never been included in the cranial hyperostoses. Yet it fits perfectly well in this group. Some objected to its inclusion, claiming that the lesion is primarily one of otospongiosis. This is not quite true. Furthermore, such an arbitrary elimination, to be consistent, should also embrace all the other truly spongious hyperostoses.

Finally, for comparative purposes the pseudo-hyperostoses are also briefly considered.

In the ensuing paragraphs the diverse varieties of H.C. will be discussed, using the following classification (see Table III) as an outline.

Classification of Cranial Hyperostosis

I. Hyperostosis due to hereditary factors affecting bones in general.
II. Hyperostosis due to congenital malformations and anomalies of development of skull.
III. Hyperostosis due to infectious and parasitic agents.
IV. Hyperostosis due to blood dyscrasia.
V. Hyperostosis due to physical agents and mechanical factors.
VI. Hyperostosis due to dietary factors.
VII. Hyperostosis due to known metabolic endocrine disturbances.
VIII. Hyperostosis due to dysplastic or dystrophic changes of unknown or complex etiology.
IX. Hyperostosis due to neoplasms and neoplasm-like lesions.
X. Pseudo-hyperostosis caused by lesions or conditions mimicking hyperostosis.

Subclassification

I. A) Racial factors.
B) Hereditary pathologic conditions (Ex. Osteopoikilosis; osteopetrosis; osteogenesis imperfecta).
II. A) Congenital, pathological and physiological overgrowths [Ex. Tori; occipital spur; elongated styloid process; natiform skull (?)].
B) Craniostenosis (Ex. Oxycephaly; scaphocephaly; plagiocephaly; trigonocephaly).
C) Maldevelopment (Ex. Macrocephaly; microcephaly; hemi-microcephaly; synergistic hyperostosis).
III. A) Infectious (Ex. Syphilis; tuberculosis; non-specific; miscellaneous).
B) Parasitic (Ex. Malaria; Kala-azar; toxoplasmosis).
IV. A) Erythroblastic anemias of childhood (Ex. Cooley's anemia).
B) Hereditary spherocytosis.
C) Sickle cell anemia.
D) Miscellaneous.
V. A) Traumatic agents (Ex. Cephalhematoma; old fracture).
B) Miscellaneous.
VI. A) Scurvy.
B) Rickets.
VII. A) Acromegaly—gigantism.
B) Fröhlich's syndrome.
C) Pregnancy.
D) Osteitis fibrosa cystica generalisata.
VIII. A) Predominantly dysplastic and hyperplastic [Ex. H.C.I. and variants; Paget's disease of bone; fibrous dysplasia and variants; leontiasis ossea (?); otosclerosis; infantile cortical hyperostosis (?); progressive diaphyseal dysplasia].
B) Predominantly dystrophic or degenerative (Ex. H.C.D. and variants).
IX. A) Neoplasm (Ex. Meningioma; metastatic tumor; primary malignant tumor).
B) Neoplasm-like lesion (Ex. Osteoma and variants; hemangioma).
X. A) Pseudo-hyperostosis proper (Ex. Macrocrania without hyperostosis).
B) So-called intercalated or wormian bones.
C) Heterotopic or heteroplastic hyperostosis (Ex. Meningeal ossification).

PART I

In this part the first six varieties of cranial hyperostoses are grouped together in order to set them apart from the so-called "idiopathic" hyperostoses and the pseudo-hyperostoses which will be the subjects of Parts II and III.