SECOND EDITION

ILLUSTRATED

A Textbook of
ORAL PATHOLOGY

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W. B. SAUNDERS COMPANY

PHILADELPHIA AND LONDON—1963

Press of W. B. Saunders Company. Library of Congress catalog card number: 63:9499.

PREFACE TO THE SECOND EDITION

The increasing responsibilities of the dental profession for the total health care of the patient have necessitated a revision and expansion of this textbook of oral pathology. It seemed important to bring up to date much of the material presented in the first edition as well as to add a considerable amount of new technical data, concepts, bibliographic contributions and illustrations.

Obviously every condition discussed in the present text cannot be considered in detail by the undergraduate dental student. The choice of course content must rest with the teacher. It is our view, however, that a textbook of oral pathology should make available the essential information necessary for an understanding of oral and dental diseases.

The original concept of this textbook has not been altered. Our primary aim has been to present a discerning, objective discussion of oral and paraoral diseases. We hope that the description of the local and systemic aspects of oral disease will enable the dentist to fulfill his obligation to the patient in a more satisfactory professional manner. We have not deviated from our primary concern, which was to explain oral diseases by the application and integration of histologic, chemical and physiologic alterations of basic biologic processes.

The first edition of this book contained its complement of misinterpretations and errors of omission and commission. The critical evaluation of the text by the many friends and colleagues of the authors has resulted in the correction or elimination of many of the errors and misinterpretations. It is the sincere desire of the authors that such critical evaluation will continue in order to provide a text embodying the most generally accepted concepts of oral disease.

THE AUTHORS

ACKNOWLEDGMENTS

Many persons have contributed unselfishly their time, special talents, knowledge and illustrative material to aid in the completion of this book. Those allowing free use of their illustrations have been credited in the legends for the particular figures, and grateful acknowledgment is once again made here. In addition, special acknowledgment must be made to numerous individuals for use of their excellent material in the preparation of the color plates. Of many hundreds of clinical color photographs reviewed, very few were of a quality sufficient to warrant their duplication in color. The authors expressly wish to thank the following individuals for permission to use their material: Dr. Stephen F. Dachi, eight illustrations in Plates III, IV and V; Dr. Warren B. Davis, one illustration in Plate IV; Dr. John R. Mink, one illustration in Plate IV; Dr. David F. Mitchell, four illustrations in Plates I, II and III; Dr. Wilbur C. Moorman, one illustration in Plate VII; Dr. Gil Small, one illustration in Plate II; Dr. S. Miles Standish, two illustrations in Plates I and IV; and Dr. Henry M. Swenson, two illustrations in Plate VII.

Various chapters and sections of the text have been read, criticized and corrected by persons with a particular interest in certain areas of pathology, and to them are extended sincere thanks. These include Drs. Sumter S. Arnim, Boynton H. Booth, Charles Burstone, Martin Lunin, Irwin Mandel, David F. Mitchell, S. Miles Standish and Edward B. Smith. Hours of consultation also were devoted to this project by many members of the staffs of Indiana University School of Dentistry and School of Medicine, including Drs. J. William Adams, G. Thaddeus Gregory, J. Frank Hall, Harry J. Healey, Ralph E. McDonald, Wilbur C. Moorman, Ralph W. Phillips, Ronald S. Ping, Robert J. Rohn, Lewis B. Spear, Henry M. Swenson, Grant Van Huysen, Frank Vellios and Miss Ruhamah Hannah.

Our deep appreciation is due to a number of persons with whom, over a period of years, we have held lengthy discussions of the technical and philosophical aspects of oral pathology which have resulted in the authors' arrival at many of their conclusions and beliefs. This list must include Dr. S. Miles Standish, whose inesti-

mable aid can never be properly or sufficiently acknowledged; Drs. Donald A. Kerr and Robert J. Gorlin, particularly for their critical reading and suggestions; Drs. William G. Sprague and Charles A. Waldron for their continued inspiration through constant discussions, agreement and disagreement; Dr. Frank Vellios, for his willing help and advice; and, finally, the many stimulating graduate students with whom it has been our privilege to be associated over the years.

The photographs have been prepared largely by Mr. Richard Scott and Mrs. Gloria Spray of the Illustration Department of Indiana University School of Dentistry. The drawings were made by Dr. Peter Kesling. Assistance for the preparation of illustrative material, an important part of any book, is gratefully acknowledged.

The many rough drafts and the final copy of the manuscript were typed by Mrs. Sondra Hawkins, Mrs. Dorothy Lincoln and Miss Margaret Herron, to whom special thanks are due. Mrs. Ruth Chilton also aided in typing some sections.

The tissue laboratory of the Department of Oral Pathology is in charge of Mrs. Patricia G. Clark, who is responsible for the excellent histologic material used in most of the photomicrographs.

A special debt of gratitude is owed to Sylvia Levy for her part in this undertaking. For both editions of the text, she painstakingly read and reread not only rough and final copies of the manuscript, but also all galley and page proofs, correcting errors in punctuation, spelling, grammar and syntax with an unselfish devotion. The undertaking of this book would have been impossible also without the sacrifices made by the authors' wives in tolerating the long hours spent in writing. To them, special recognition is offered.

THE AUTHORS

PREFACE TO THE FIRST EDITION

ORAL PATHOLOGY represents the confluence of the basic sciences and clinical dentistry. Since it has no methods of its own, knowledge in this field is acquired through the adaptation of methods and disciplines of those sciences basic to dental practice, such as gross and microscopic anatomy, chemistry, microbiology and physiology, and through information obtained by clinical histories and observation of patients. Through the science of oral pathology, an attempt is made to correlate human biology with the signs and symptoms of human disease. The oral pathologist attempts to understand oral disease so that it can be properly diagnosed and adequately treated.

In this text we have attempted to bring the reader to an understanding of the patient and his problems through applied basic science. We have tried to explain clinical signs and symptoms in the light of known histologic, chemical and physiologic alterations. Where possible, the prognosis of each disease is considered as a reflection of the underlying tissue changes and what we know can be done about them today.

In numerous sections of the text we have attempted to integrate information from many of the basic sciences for adequate diagnosis of oral disease. This approach is a departure from that of the usual textbook of oral pathology, representing an effort to place more emphasis on the physiologic and chemical aspects of oral disease.

The references at the end of each chapter are extensive enough to be of value to those interested in additional reading. Only those papers which constitute good review articles or exceptional discussions or which are of historical importance are included. Because the field of oral pathology is large, much of the bibliographic material has had to be curtailed or omitted. The highlights alone have been stressed. It is our hope that this book will prove to be a stimulus to study as well as a guide for undergraduate and graduate students and practitioners of both dentistry and medicine.

THE AUTHORS

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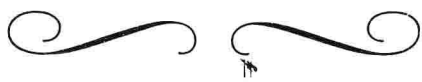
Section I



DISTURBANCES OF DEVELOPMENT AND GROWTH

Chapter 1

Developmental Disturbances of Oral and Paraoral Structures

An understanding of the many disturbances of development and growth which involve the oral and paraoral structures is predicated upon a thorough understanding of the embryology and histology of these structures. Some of the conditions to be discussed here develop *in utero*, are present at birth and persist throughout life. Others may not manifest themselves for many years. The recognition that some abnormalities follow the traditional patterns of inheritance has been of great help to the scientist in explaining many unusual pathologic conditions which affect the living organism. Great care must be taken, however, to distinguish between hereditary and congenital conditions. A congenital disease is one which is present at or before birth, but is not necessarily inherited, i.e. transmitted through the genes. In contrast, many hereditary conditions are apparent at birth, while others do not become evident for a number of years after birth.

The explanation for the tendency of a person to inherit certain features or characteristics from his parents is based upon the monumental observations of Gregor Mendel, who did much to establish the laws of heredity. From his painstaking and carefully recorded work arose the science of genetics.

One of Mendel's most important discoveries was that of the principle of dominance, which was based upon experiments showing that if the two members of a given pair of individuals with contrasting characters are brought together in a cross, there is a decided difference in the ability of these characters to be expressed in the resulting offspring. Thus one of the characters, but not the other, may manifest itself. Of great importance, however, is the fact that the unexpressed character is not eliminated, for it may be manifested in subsequent generations. The terms "dominant" and "recessive" have been applied, respectively, to those features which do and which do not appear in the first filial generation following the cross. The subsequent separation and reappearance of characters in the offspring of hybrid persons are known as the principle of segregation and are a feature of inheritance.

It is well established that the many characteristics of a person are represented in the genes of the gametes, or reproductive cells. In speaking of or comparing only two characteristics, the possibilities of assortment are relatively limited, but as the number of characteristics increases, it is obvious that the possibilities of combinations become infinitely more complex.

Another important principle of inheritance, discovered by T. H. Morgan, is that of linkage between factors, which offers an explanation for the early observation that two or more characters may remain linked in their passage from one generation to another. An offshoot of this principle was the discovery of sex linkage, or the linkage of certain characters with the factors which determine sex. For example, a man may transmit a sex-linked trait to his grandsons through his daughters, but not to or through his sons. Such a transmission depends on the paired sex chromosomes of an individual.

Many of the developmental and growth disturbances of the oral and paraoral structures, as well as other oral diseases to be discussed, have a definite hereditary background. Other diseases in which the evidence for inheritance is suggestive but not conclusive are sometimes said to present "familial tendencies." Dentistry has much to offer in helping to determine the true etiologic factors in the many disturbances of development and growth of the teeth, bones and various soft tissues. Witkop, writing on the role of genetics in dentistry, has stressed the fact that in some dental diseases, inherited factors can be either decisive or only contributory to the production of a specific illness. As examples of hereditary oral diseases, he has presented the compilation shown in Table 1.

Genetic factors are undoubtedly of importance in the development of many human congenital malformations, although it has been estimated that only about 10 per cent of such malformations can be explained on a genetic basis. A second important factor in the development of such disturbances is that of pathologic environmental conditions, and these have been estimated to account for an additional 10 per cent of developmental anomalies. The remaining 80 per cent are, in light of present knowledge, idiopathic.

A remarkable scientific interest has developed in the possible environmental causes of congenital malformations, and a vast number of both clinical and experimental animal studies have been carried out to clarify this relationship.

Haring and Lewis have reviewed the scientific literature on known etiologic factors in both spontaneous and experimental congenital developmental anomalies and have tabulated all currently known teratogenic factors. These are listed in Table 2. In considering the problem of congenital malformations, Haring and Lewis evolved certain principles, based on scientific evidence, applicable to both animal and human teratogenesis. These principles are as follows: (1) experimentally induced malformations in animals are similar to those occurring spontaneously and sporadically in the animal population, i.e. phenocopies; (2) many different agents can induce the same type of defect; (3) the same agent applied at different stages of development produces different types of defects; (4) the same defect can be induced regularly and at will if a teratogenic agent is applied at the same and proper time during the development in the same strain; (5) specific defects can be induced with greater ease in certain strains of a species than in others; (6) teratogenic agents do not necessarily alter the mother's condition of health.

It is of considerable interest to the dental profession that experimental studies of teratogenic agents have almost invariably revealed a variety of oral malformations, many of which have a human counterpart.

DEVELOPMENTAL DISTURBANCES OF THE JAWS

AGNATHIA

Agnathia is an extremely rare congenital defect characterized by absence of the maxilla or mandible. More commonly, only a portion of one jaw is missing. In the case of the maxilla, this may be one maxillary process or even the premaxilla. Partial absence of the mandible

is even more common. The entire mandible on one side may be missing or, more frequently, only the condyle or the entire ramus, although bilateral agenesis of the condyles and of the rami also has been reported. In cases of unilateral absence of the mandibular ramus, it is not unusual for the ear to be deformed or absent as well.

MICROGNATHIA

Micrognathia literally means a small jaw, and either the maxilla or the mandible may be affected. Many cases of apparent micrognathia are due not to an abnormally small jaw in terms of absolute size, but rather to an abnormal positioning or an abnormal relation of one

Table 1. Hereditary Oral Diseases

ORAL DISEASE	MODE OF INHERITANCE D—DOMINANT R—RECESSIVE S—SEX-LINKED IS—INTERMEDIATE SEX-LINKED	ACCURACY OF GENETIC PROGNOSIS ...—ACCURATE ..—APPROXIMATE .—QUESTIONABLE
Heritable defects in dentition without generalized defects:		
Hypoplasia of enamel	SD	..
Hypocalcification of enamel	D	...
Hypomaturation of enamel	SR	...
Pigmented hypomaturation of enamel	R	.
Local hypoplasia of enamel	D (with incomp. penetrance)	..
Dentin dysplasia	D	...
Dentinogenesis imperfecta	D	...
Missing or peg laterals	D	...
Missing maxillary incisors and cuspids	D or R	..
Missing premolars	D	..
Missing third molars	D	..
Gigantism of maxillary central incisors	D	..
Fused primary mandibular incisors	D?	..
Familial dentigerous cysts	D	..
Heritable defects in dentition with generalized defects:		
Dentinogenesis imperfecta with osteogenesis imperfecta	D	..
Enamel hypoplasia in vitamin D-resistant rickets	D (irregular)	..
Enamel hypoplasia with epidermolysis bullosa dystrophia	R	..
Local hypoplasia of enamel with Fanconi syndrome	R?	.
Missing teeth with ectodermal dysplasia	IS or D	..
Missing premolars with premature whitening of hair	D	...
Missing lateral incisors with ptosis of eyelid	D	..
Retarded eruption with cleidocranial dysostosis	D	..
Heritable defects of oral structures without generalized defects:		
Ankyloglossia	D	..
Elephantiasis gingivae	D	..
Harelip and harelip with cleft palate	R?	.
Heritable defects of oral structures with generalized diseases:		
Gangrenous stomatitis with acatalasemia	R	..
Periodontitis with agammaglobulinemia	SR	..
Periodontitis and osteoporosis of jaw bones with thalassemia major	R	...
Alveolar bone changes in sickle cell disease	R	..
Gingival and postoperative hemorrhage in hemophilia and Christmas disease	SR	...
Mucosal telangiectasia in hemorrhagic telangiectasia (Osler)	D	..
Facial angiomatosis with Sturge-Weber disease	D (irregular)	.

Table 1. Hereditary Oral Diseases—*Continued.*

ORAL DISEASE	MODE OF INHERITANCE D—DOMINANT R—RECESSIVE S—SEX-LINKED IS—INTERMEDIATE SEX-LINKED	ACCURACY OF GENETIC PROGNOSIS ...—ACCURATE ..—APPROXIMATE .—QUESTIONABLE
Oral hematomas with Ehlers-Danlos syndrome	D	..
Facial deformity in gargoylism	R (RS)	..
Facial deformity with mandibulofacial dysostosis (Franceschetti)	D (irregular)	..
Facial deformity with craniofacial dysostosis (Crouzon)	D (irregular)	..
Micrognathia with Pierre Robin syndrome	R (incomp. dom.)	.
Hypoplasia of maxilla with achondroplasia	D	..
Multilocular cystic fibrous dysplasia of the jaws and face (Jones)	D	...
Osteosclerosis in Albers-Schönberg disease	D (irregular)	..
Hyperostosis of jaws in generalized hyperostotic bone disease (Witkop)	R	...
Hypercementosis and bone changes in osteitis deformans (Paget)	D (incomp.)	.
Neurofibroma and pigmentation in neurofibromatosis (von Recklinghausen)	D	..
Circumoral pigmentation with gastrointestinal polyposis (Peutz-Jeghers)	D	..
Facial pigmentation and carcinomas of lip in xeroderma pigmentosum	ISR or R	..
Gingival and lingual amyloid deposits in familial amyloidosis	D	..
White spongy nevus of mucous membranes	D	...

From C. J. Witkop, Jr.: Genetics and Dentistry. Eugenics Quart., *5*:15, 1958.

Table 1 is a partial classification of hereditary oral diseases listing the common mode of inheritance and the accuracy of prognosis based on the oral findings. Some traits, such as dentinogenesis imperfecta, are known to be inherited only in the manner listed, while others, such as peg or missing laterals, may be associated with a number of genetic entities or may be phenocopies; that is, due to a non-genetic cause. If good evidence exists that a trait does occur in a hereditary form, then that trait has been included. Thus, a specific trait such as premature whitening of the hair and missing premolars is known to be inherited in an autosomal dominant manner in some kindreds. This does not mean that all cases of premature whitening of the hair will be associated with missing premolars or that all cases of missing premolars will be inherited with premature whitening of the hair.

A single trait may show two forms of inheritance. Thus, missing teeth may be found in two forms of ectodermal dysplasia. One form shows an autosomal dominant mode of inheritance in some families, while in other families the gene apparently is passed on the X-chromosome. In this latter form, males are severely affected, but females with the gene may show only a few missing teeth.

This list should be used as a guide only and should not be relied upon as the sole evidence for making a genetic prognosis in any individual case. A genetic prognosis in any individual case should be made on the basis of a complete family history of how the trait in question is inherited in that particular family.—Carl J. Witkop, Jr.

jaw to the other or to the skull, which produces the illusion of micrognathia.

True micrognathia may be classified as either (1) congenital or (2) acquired. The etiology of the *congenital* type is unknown, although in many instances it is associated with other skeletal abnormalities. It occasionally follows a hereditary pattern. Micrognathia of the maxilla is frequently due to a deficiency in the premaxillary area, and patients with this deformity appear to have the middle third of the face retracted. Although it has been suggested that mouth-breathing is a cause of maxillary micrognathia, it is more likely that the micrognathia may be one of the predisposing factors in the mouth-breathing, owing to associated

Table 2. Experimental and Clinical Causes of Congenital Developmental Anomalies

Genetic factors	Inherited
	Mutagenetic
Environmental factors:	
1. Infections	Rubella
	Influenza A
2. Physical injuries	Pressure
	Temperature changes
	Radiation
3. Hormones	Diabetes mellitus
	Alloxan diabetes
	Hyperthyroidism
	Hypothyroidism
	Antuitrin G
	ACTH
	Cortisone
	Steroid hormones
	Insulin
4. Nutrition	Deficiencies of
	Vitamin A
	Vitamin B_1
	Vitamin B_2
	Vitamin B_6
	Niacin
	Folic acid
	Vitamin B_{12}
	Vitamin D
	Vitamin E
	Vitamin K
	Proteins
	Amino acids
	Unsaturated fatty acids
	Potassium
	Excess of vitamin A
5. Respiration	Hypoxia
	Carbon dioxide excess
	Carbon monoxide
	Anesthesia with ether-gas-oxygen
6. Miscellaneous drugs and chemicals	
	Antimetabolites
	Aminopterin
	6-Mercaptopurine
	Azaserine
	5-Fluorodeoxyuridine
	6-Aminonicotinamide
	Thiadiazole
	6-Chloropurine
	Thioguanine
	2-6-Diamonopurine
	Polyfunctional alkylating agents:
	HN_2
	CB 1348
	TEM
	ThioTEPA
	Myleran and others
	Triazene
	Quinine
	Urethane
	Colchicine
	Trypan blue
	Pilocarpine
	Eserine
	Boric acid
	Thallium
	Selenium
	Nicotine
	Sulfonamides
	Antibiotics
	Salicylates
	Malachite green
7. Maternal diseases and defects	Uterine tumors
	Uterine inflammation
	Uterine malformation
	Defects in implantation
	Age
	Emotional disturbances
	Stress
	Multiple pregnancies
8. Embryonic defects	Abnormalities of the ovum
	Abnormalities of the semen
	Antigen-antibody reactions

Courtesy of Drs. Olga M. Haring and F. John Lewis: Surg., Gynec. & Obst., *113*:1, 1961.

maldevelopment of the nasal and nasopharyngeal structures.

True mandibular micrognathia of the congenital type is often difficult to explain. Some patients appear clinically to have a severe retrusion of the chin, but, by actual measurements, the mandible may be found to be within the normal limits of variation. Such cases may be due to a posterior positioning of the mandible with regard to the skull or to a steep mandibular angle resulting in an apparent retrusion of the jaw. Agenesis of the condyles also results in a true mandibular micrognathia.

The *acquired* type of micrognathia is of postnatal origin and usually results from a disturbance in the area of the temporomandibular joint. Ankylosis of the joint, for example, may be caused by trauma or by infection of the mastoid, of the middle ear or of the joint itself. Since the normal growth of the mandible depends to a considerable extent on

normally developing condyles as well as on muscle function, it is not difficult to understand how condylar ankylosis may result in a deficient mandible.

The clinical appearance of mandibular micrognathia is characterized by severe retrusion of the chin, a steep mandibular angle and a deficient chin button (Fig. 1).

MACROGNATHIA

Macrognathia refers to the condition of abnormally large jaws. An increase in size of both jaws is frequently proportional to a generalized increase in size of the entire skeleton, e.g. in pituitary giantism. More commonly only the jaws are affected, but macrognathia may be associated with certain other conditions, such as (*a*) Paget's disease of bone, in which overgrowth of the cranium and maxilla or occasionally the mandible occurs; (*b*) acromegaly, in which there is progressive enlargement of the mandible owing to hyperpituitarism in the adult; or (*c*) leontiasis ossea, a form of fibrous dysplasia in which there is enlargement of the maxilla.

Cases of mandibular protrusion or prognathism, uncomplicated by any systemic condition, are a rather common clinical occurrence (Fig. 2). The etiology of this protrusion is unknown, although some cases follow hereditary patterns. In many instances the prognathism is due to a disparity in the size of the maxilla in relation to the mandible. In other cases the mandible is measurably larger than normal. The angle between the ramus and the body also appears to influence the relation of the mandible to the maxilla, as does the actual height of the ramus. Thus prognathic patients tend to have long rami which form a less steep angle with the body of the mandible. The length of the ramus, in turn, may be associated with the growth of the condyle. It may be reasoned, therefore, that excessive condylar growth predisposes to mandibular prognathism.

General factors which conceivably would influence and tend to favor mandibular prognathism are as follows: (1) increased height of the ramus, (2) increased mandibular body length, (3) increased gonial angle, (4) anterior positioning of the glenoid fossa, (5) decreased maxillary length, (6) posterior positioning of the maxilla in relation to the cranium, (7) prominent chin button, and (8) varying soft tissue contours.

Surgical correction of such cases is feasible. Ostectomy, or resection of a portion of the mandible to decrease its length, is now an established procedure,

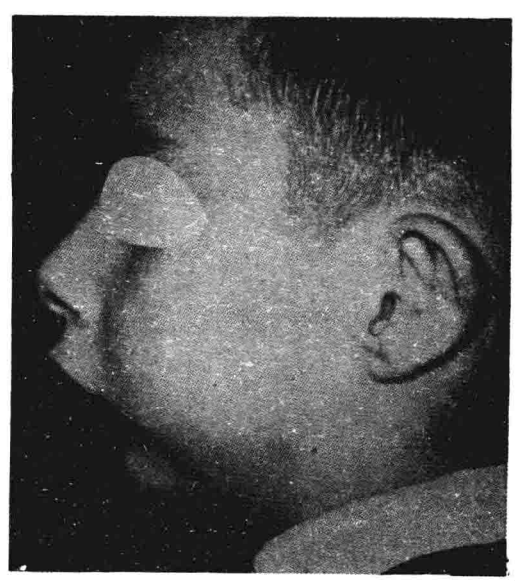


Figure 1. ***Mandibular micrognathia.*** (Courtesy of Drs. G. Thaddeus Gregory and J. William Adams.)

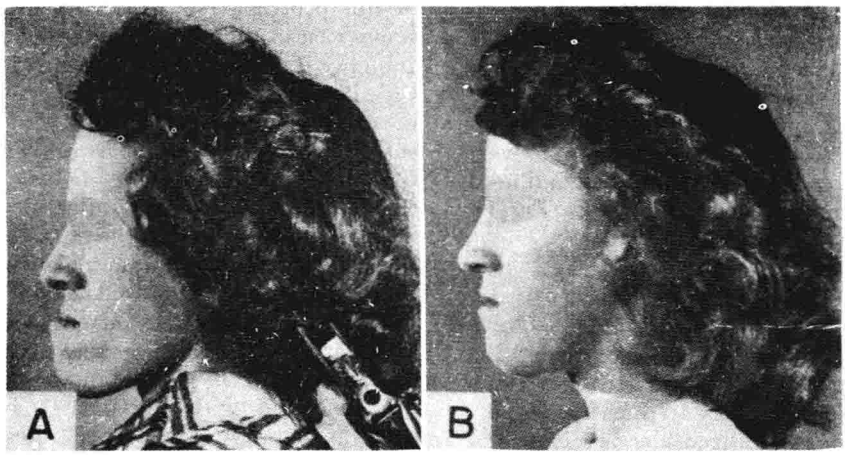


Figure 2. ***Macrognathia (prognathia) of the mandible.***
A, The protrusion of the mandible is obvious. *B,* The same patient after surgical correction (ostectomy). (Courtesy of Drs. G. Thaddeus Gregory and J. William Adams.)

and the results are usually excellent from both a functional and a cosmetic standpoint.

FACIAL HEMIHYPERTROPHY

A very mild degree of facial asymmetry is present in nearly all persons, and this is often imperceptible to even close observation. Occasionally, however, a congenital hemihypertrophy may occur, involving (1) the entire half of the body, (2) one or both limbs or (3) the face, head and associated structures. Although the unilateral facial hypertrophy is the most gross striking feature in patients with this disturbance, the unusual hemihypertrophy of the jaws and teeth is the most significant finding to the dentist.

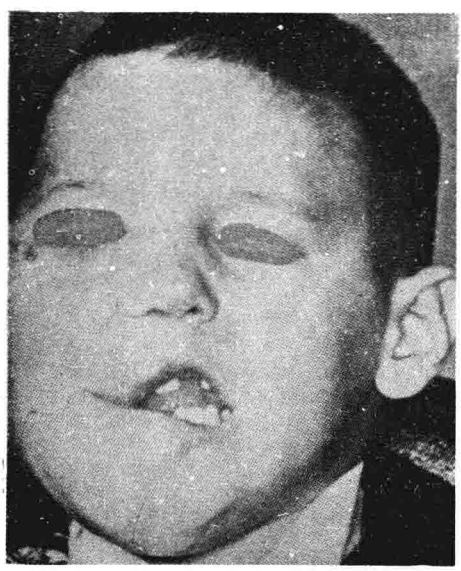


Figure 3. ***Facial hemihypertrophy.***
The asymmetric disfigurement is obvious despite several attempts at surgical correction.

Etiology. The cause is unknown, but the condition has been variously ascribed to (1) hormonal imbalance, (2) incomplete twinning, (3) chromosomal abnormalities, (4) localized alteration of intrauterine development, (5) lymphatic abnormalities, (6) vascular abnormalities, and (7) neurogenic abnormalities. Of all of these, the last two, vascular and neural abnormalities, seem the most plausible to explain the clinical findings.

Clinical Features. Patients afflicted by facial hemihypertrophy exhibit enlargement of one half of the head (Fig. 3). In some cases this is obvious even at birth. The enlarged side grows at a rate proportional to the uninvolved side, so that the disproportion is maintained throughout life, although growth of the entire face generally ceases by the age of twenty years. Familial occurrence has been reported on a few occasions, according to the excellent review of the