Dentition
Genetic Effects

Editor
Ronald J. Jorgenson

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DENTITION: GENETIC EFFECTS

Proceedings of the Fifth Annual Symposium of the Society of Craniofacial Genetics, Held in Birmingham, Alabama, June 13, 1982

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The **March of Dimes Birth Defects Foundation** is dedicated to the goals of preventing birth defects and ameliorating their consequences for patients, families, and society.

As part of our efforts to achieve these goals, we sponsor, or participate in, a variety of scientific meetings where all questions relating to birth defects are freely discussed. Through our professional education program we speed the dissemination of information by publishing the proceedings of these and other meetings. From time to time, we also reprint pertinent journal articles to help achieve our goal. Now and then, in the course of these articles or discussions, individual viewpoints may be expressed which go beyond the purely scientific and into controversial matters. It should be noted, therefore, that personal viewpoints about such matters will not be censored but this does not constitute an endorsement of them by the **March of Dimes Birth Defects Foundation.**

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The boldface number in brackets indicates the opening page of the author's contribution.

Doctor John J. Sharry

Dedication

The proceedings of this symposium are dedicated to the memory of John J. Sharry. Most readers of these proceedings did not know John personally or by reputation: he was not a central figure in genetics. He was a prosthodontist (a plate man, he'd say), a dean, a humanist, and a friend.

John was born on February 11, 1925 in Somerville, Massachusetts. His baccalaureate and dental degrees were earned at Tufts University. He trained as a resident in oral surgery in Birmingham, and as a prosthodontist at Tufts and at the Zoller clinic in Chicago. After his training he began his meteoric and distinguished academic career in Birmingham: in just seven years he rose to the rank of Professor and Head of the Department of Prosthetic Dentistry. During these early years of his career he developed an interest in one of the common craniofacial anomalies—cleft lip and palate. This interest undoubtedly was nourished throughout the years that he was a member and director of the cleft palate program at the University of Alabama, during a fellowship he served in Sweden, and while he was a board member of the Alabama Foundation for Speech and Hearing. Meanwhile his extraordinary talents in his chosen specialty were rewarded by election to offices of its regional and national societies, culminating in his election to the presidency of the American College of Prosthodontics in 1975.

Before 1975, however, he rekindled his interest in craniofacial anomalies through the organization of a clinical genetics program in the College of Dental Medicine at the Medical University of South Carolina, where he was serving as dean of the College. Later, he instituted a similar program at the Dental School of the University of Texas Health Science Center at San Antonio, during his foreshortened service as dean of that school. Two programs in clinical genetics may not seem to be much to some observers, but when one considers that there are only a handful of such programs in dental schools across the country, and what the fiscal climate was during the time he instituted the programs, it represents a remarkable contribution to the field of genetics in dentistry.

No one but John would deny that he was a humanist. His service on the boards of trustees of symphonies, on committees for economic opportunities, community service councils, and the establishment of a humanistic lecture series testify to his involvement with his fellow man. His numerous publications reflect his insight into the human side of dentistry. Included in the titles of his scholarly works are such phrases as: concerning several thoughts, the public weal, research attitudes, prestige for the teachers, notes from a latitudinarian, institutional introspection, balance in universities, whimsy, arguments, and reappraisal.

It is no surprise to those who knew him that he was listed as one of the outstanding educators in America, received numerous awards from his peers, and was chosen shortly before his death as the year's distinguished lecturer by the Greater New York Academy of Prosthodontics. No wonder that many feel a tremendous personal and professional loss at his early demise.

Ronald J. Jorgenson, DDS, PhD

Preface

The Society of Craniofacial Genetics (SCFG) has now sponsored five symposia in conjunction with the annual Birth Defects Conferences. The proceedings of the previous symposia have been published [1–4], and have added substantially to the literature on the special problems of head and neck dysmorphology.

Previous symposia have concentrated on general concepts (the developmental process underlying dysmorphologic features), organ-systems (the eye and the ear), and clinical application. In planning for the symposium that resulted in this text, members of the SCFG felt that there was a need to prepare a series of concise papers on orofacial structures for readers of the **Birth Defects: Original Article Series.** Thus, the papers of the symposium under discussion concentrated on several aspects of the dentition only: embryology, eruption, occlusion, and specific anomalies.

Future symposia are planned on other orofacial structures. In order to enhance the presented papers and further develop the theme that a comprehensive and standard evaluation of the orofacies is important to clinical and nonclinical dysmorphologists, three papers on heterogeneity comprise the second section of this text.

None of the symposia sponsored by the Society would have been possible without the support of the March of Dimes Birth Defects Foundation through its national and local chapters. Special thanks for this symposium are expressed to the San Antonio, New Orleans, and Birmingham chapters; to Mr. Richard Dyer and Dr. Wayne Finley; and to the University of Texas Health Science Center at San Antonio and the University of Alabama in Birmingham.

Ronald J. Jorgenson, DDS, PhD

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Contents

Part I

The Dentist and the Dentition

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This chapter is meant to serve as a brief introduction to this volume. It provides a contemporary description of the dental role as a member of an interdisciplinary health team (a topic covered more thoroughly by the last three papers), and reviews some tenets of dentistry so that those readers who are not dentists may begin to familiarize themselves with the field. Each topic addressed here is covered in significantly more detail by papers that follow.

THE SCOPE OF DENTISTRY AS IT RELATES TO BIRTH DEFECTS

A discussion of the role of the dentist in managing individuals with birth defects was presented in the 1980 "Symposium on Dentistry in the Interdisciplinary Treatment of Genetic Diseases" [1]. In addition to the activities of diagnosing and treating orofacial aspects of genetic disorders, dentists were shown to have roles as anatomists, geneticists, and researchers. Recognition that such roles exist has led to increased emphasis on human genetics in dental curricula. Many dental postgraduate specialty programs include participation in craniofacial anomalies teams as an integral portion of clinical activity.

Dental clinicians have pursued graduate programs in genetics, anatomy, and pathology, which all provide a structured research experience, as well as high competency levels for their respective areas of study.

Dental disciplines that have traditionally been represented on comprehensive health teams include oral pathology, orthodontics, oral surgery, pediatric dentistry, prosthodontics, and general dentistry. In reality, the particular discipline is not nearly as important as the pluralistic interest on the part of the dentist for the total patient and his or her appreciation of the diagnosis and treatment expertise of all team members.

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Diagnosis of dental disease is established in the same manner as that of other diseases—by thorough health and family history, and clinical examination. The dentist must be as precise as other professionals in documenting growth, behavior, and vital signs. In addition, the dentist pays particular attention to extraoral and intraoral findings. Extraoral examination includes assessment of facial profile, facial symmetry, lymphadenopathy, and the state of other structures such as hair, ears, skin, eyes, nose, and lips (Fig. 1). Intraoral examination includes assessment of soft tissues (gingiva, frenula, tongue, palatal mucosa, oral pharynx, and buccal mucosa), assessment of hard tissues (bony exostoses, palate morphology, enamel, dentin, jaws, and teeth), and documentation of dental caries and occlusion (jaw relation, tooth malposition, crossbite, and openbite) (Fig. 2). Assessment of the dentition is the major topic of this text. Such assessment, as described in this volume must include information on tooth structure, size, number, alignment, and interdigitation (Fig. 3).

DENTAL DEVELOPMENT

Embryologically, the maxilla and mandible are derived from the 1st branchial arch. Odontogenic epithelium, which will give rise to the tooth buds, is differentiated from the epithelia of the mandibular, maxillary, and median nasal processes between 28 and 37 days in utero. A single and

Extra Oral:	Initial	Recall	Recall	Recall	Recall
Date					
Ht. In.					
Wt. Lbs.					
Pulse					
B P	/	/	/	/	/
Facial Sym					
Scars					
*Profile					
Lip Length					
T M J					
Nodes					
Hair					
Skin					
Eyes					
Ears					
Nose					

Fig. 1. Checklist used during a comprehensive oral examination.

continuous plate of odontogenic epithelium develops in each arch and gives rise to the dental lamina [2]. Proliferation of dental lamina cells at 20 specific sites occurs at 6 weeks' gestation and represents initiation of the 20 deciduous tooth buds. The 32 permanent tooth buds begin to develop at 5 months in utero, but the last buds appear at 5 years postpartum. The most critical period

Intra Oral:	Initial	Recall	Recall	Recall	Recall
Lips					
Buccal Mucosa					
Palate					
Tonsils					
Oropharynx					
Tongue					
Sublingual Tissue					
Gingiva					
Frenum					
Habits					
Siblents					
Lingual Alveolar					
Bilabial					

*CV - Convex CC - Concave St. - Straight
N = Normal or None
ABN = Abnormal - (Discussed in Progress Notes)

Fig. 2. Checklist used during an intraoral examination.

Occlusion:	Initial		Recall		Recall		Recall		Recall	
Date										
Terminal Plane**	R	L	R	L	R	L	R	L	R	L
6-Year Molar	R	L	R	L	R	L	R	L	R	L
1* Cuspid	R	L	R	L	R	L	R	L	R	L
Overbite (%)										
Overjet (mm)										
Openbite (mm)										
Midline										
Crossbite										
Arch Length	R	L	R	L	R	L	R	L	R	L

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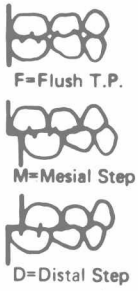

Fig. 3. Checklist used during evaluation of dental occlusion.

for anomalous development of deciduous teeth is between 6 and 8 weeks in utero [3]. Chronologically, tooth initiation occurs after the major organ systems and external structures have been established (Fig. 4).

Teeth and bone are hard tissues, but differ in several respects (Fig. 5). Teeth do not have the regenerative ability of bone; damaged dental tissues cannot recuperate. Unlike bone, the only changes in teeth that occur after calcification completion are degenerative. Embryologically, tooth formation precedes bone formation in the jaws. In contrast to bone, teeth have ectodermal as well as mesodermal derivatives. Teeth do not have the capacity to remodel and are formed only by apposition on their surfaces. Defects resulting from systemic disturbances are produced only during the developmental period of teeth.

Dental development involves complex processes which result in the formation of enamel, dentin, cementum, and pulp in the characteristic shapes of crowns and roots. Developmental insults are manifested as abnormalities of number, size, shape, and structure.

Abnormalities in tooth number result from problems during the initiation stage of dental development [4]. Hyperdontia may occur in the deciduous and permanent dentitions. Such teeth may be supplemental or rudimentary and occur in cleidocranial dysplasia, Gardner syndrome, orofaciodigital syndrome, and cleft lip and palate. Hypodontia also occurs in the deciduous

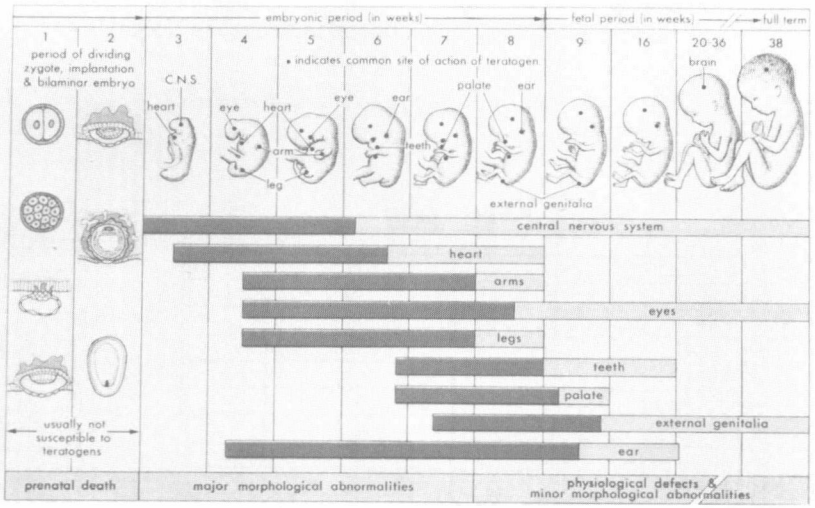

Fig. 4. Developmental table showing periods during which various organs are susceptible to teratogens.

Differences Between Teeth and Bone

1. Recuperative power
2. Post-calcification changes
3. Timing of formation
4. Embryologic tissue origins
5. Remodeling vs. apposition
6. Uniqueness of tissues
7. Timing of systemic influences

Fig. 5. List of differences between teeth and bones.

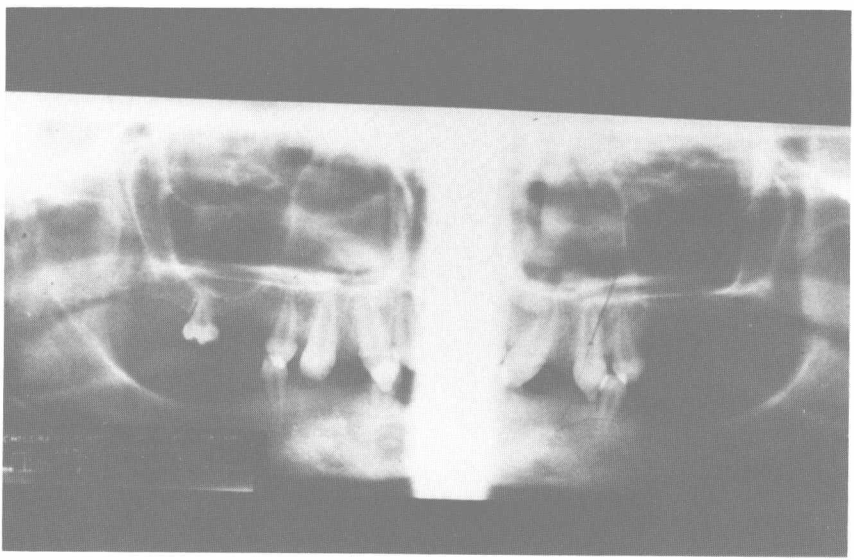

Fig. 6. Radiographs of the dentition of a patient with ectodermal dysplasia.