Volume I

Bluestone and Stool's

PEDIATRIC OTOLARYNGOLOGY

FIFTH EDITION

Charles D. Bluestone • Jeffrey P. Simons • Gerald B. Healy

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Bluestone and Stool's

Pediatric Otolaryngology

5th Edition, Volume 1

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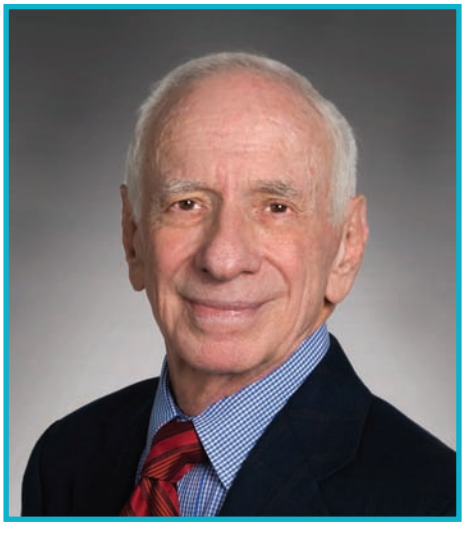

To my wife, Patsy, for 58 years of a wonderful and loving marriage, for her support and patience over the past 30 years during the countless hours of organizing, writing, and editing these five editions; and to our son Jim, his wife Maria (for her expert editing of the otitis media chapters), and our delightful granddaughters, Dane and Elyse. And lastly, to my late colleague and friend Sylvan E. Stool, coeditor for the first four editions, who collaborated with me for a year to develop the initial table of contents as we tried to codify the practice of the new subspeciality of Pediatric Otolaryngology that we had been practicing for many years.

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To my wife, Kate, for her love, support, and encouragement; and to my daughters, Ellie and Lily, who are a constant source of joy and inspiration; and to my parents, Dora and Howard, who have provided me with opportunities for a fine education, instilled in me a love of learning, and influenced me to strive for excellence.

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To my loving wife, Anne, and dear children, Lisa and Laurie. Without their support, love, and wisdom, my life would be empty. Also to my teachers and mentors and the countless patients, students, residents, fellows, and colleagues who allowed me to realize my dream of caregiver and teacher.

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Table of Contents

SECTION 5: THE AIRWAY
David L. Mandell and Reza Rahbar

SECTION 6: THE HEAD AND NECK
Trevor J. McGill and Robert F. Yellon

SECTION 7: COMMUNICATION DISORDERS
Howard C. Shane

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25: Vestibular Evaluation
29: Balance Disorders

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Division of Pediatric Otolaryngology
Children's Hospital of Pittsburgh of UPMC
Pittsburgh, PA
32: Congenital Inner Ear Anomalies
38: Complications and Sequelae of Otitis Media
104: Congenital Cysts and Sinuses of the Head and Neck

Michael J. Cunningham, MD, FACS
Otolaryngologist-in-Chief
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108: Malignant Tumors of the Head and Neck

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27: Otorrhea

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Boston, MA
31: Nongenetic Hearing Loss

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66: Pediatric Sleep Disorders
72: Management of Drooling (Sialorrhea)

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57: Foreign Bodies of the Nose
63: Congenital Malformations of the Mouth and Pharynx: Orofacial Clefts and Related Syndromes

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71: Diseases of the Salivary Glands
78: Trauma to the Mouth, Pharynx, and Esophagus in Children
91: Pediatric Upper Airway Infections

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71: Diseases of the Salivary Glands
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43: Embryology and Anatomy of the Paranasal Sinuses
52: Congenital Malformations of the Nose and Paranasal Sinuses
76: Eosinophilic Esophagitis

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45: Methods of Examination of the Nose, Paranasal Sinuses, Face, and Orbit
94: Pediatric Airway Stenosis: Minimally Invasive Approaches
107: Benign Tumors of the Head and Neck

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35: Surgical Management of Microtia and Congenital Aural Atresia
104: Congenital Cysts and Sinuses of the Head and Neck
106: Head and Neck Space Infections

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95: Airway Surgery: Open Approach

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83: Methods of Examination of the Pediatric Airway

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79: Caustic Injuries and Acquired Strictures of the Esophagus

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87: Aspiration: Etiology and Management

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22: Methods of Clinical Examination: Ear and Related Structures
37: Otitis Media and Eustachian Tube Dysfunction
38: Complications and Sequelae of Otitis Media

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114: Hemangiomas and Vascular Malformations

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95: Airway Surgery: Open Approach
114: Hemangiomas and Vascular Malformations
117: Velopharyngeal Insufficiency

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Director, Pediatric Airway, Voice and Swallowing Center
Chief Quality Officer for Otolaryngology
Massachusetts Eye and Ear Infirmary
Boston, MA
4: Outcomes and Evidence-Based Medicine in Pediatric Otolaryngology

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Children's Hospital of Michigan
Detroit, MI
47: Nasal Obstruction and Rhinorrhea

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Emeritus Healy Chair in Otolaryngology
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Emeritus Surgeon-in-Chief
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Past President, American College of Surgeons
45: Methods of Examination of the Nose, Paranasal Sinuses, Face, and Orbit

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30: Genetic Hearing Loss and Inner Ear Diseases

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St. Louis, MO
20: Embryology and Developmental Anatomy of the Ear

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36: Diseases of the External Ear

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Saint John's Hospital
Budapest, Hungary
86: Stridor: Presentation and Evaluation

Andrew J. Hotaling, MD
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Loyola University Medical Center
Maywood, IL
75: Functional Abnormalities of the Esophagus
85: Cough

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28: Tinnitus in Children

Murad Husein, MD
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30: Genetic Hearing Loss and Inner Ear Diseases

Glenn Isaacson, MD, FACS, FAAP
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Assistant Professor, Pediatrics
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81: Developmental Anatomy and Physiology of the Larynx, Trachea, Esophagus, and Lungs

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Head and Neck Surgical Oncology
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85: Cough

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University of Pennsylvania School of Medicine
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90: Congenital Malformations of the Trachea and Bronchi

Luv R. Javia, MD
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90: Congenital Malformations of the Trachea and Bronchi

Ken Kazahaya, MD, MBA, FACS
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Director, Pediatric Skull Base Surgery
Medical Director, Cochlear Implant Program
Co-Lead Surgeon, Pediatric Thyroid Center
Children's Hospital of Philadelphia
Associate Professor of Clinical Otolaryngology
Department of Otorhinolaryngology/Head &Neck Surgery
University of Pennsylvania Perelman School of Medicine
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46: Imaging of the Paranasal Sinuses in Pediatric Patients With Special Considerations for Endoscopic Sinus Surgery

Melanie A. Kazlas, MD
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Instructor, Harvard Medical School
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13: Pediatric Ophthalmology

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101: Methods of Examination of the Head and Neck

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110: Injuries of the Neck

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101: Methods of Examination of the Head and Neck

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41: Injuries of the Ear and Temporal Bone

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Shahid and Ann Carlson Khan Professor
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Champaign, IL
33: Cochlear Implants in Children

Jerome O. Klein, MD
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Department Pediatric Infectious Disease
Boston University School of Medicine
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22: Methods of Clinical Examination: Ear and Related Structures
37: Otitis Media and Eustachian Tube Dysfunction
38: Complications and Sequelae of Otitis Media

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86: Stridor: Presentation and Evaluation

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115: Pediatric Skull Base Surgery

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92: Acquired Disorders of the Larynx and Trachea

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45: Methods of Examination of the Nose, Paranasal Sinuses, Face, and Orbit

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26: Otalgia

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86: Stridor: Presentation and Evaluation

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80: Neurologic Disorders of the Mouth, Pharynx, and Esophagus

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113: Pediatric Plastic Surgery of the Head and Neck

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54: Surgical Management of Chronic Rhinosinusitis
103: Neck Masses

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56: Allergic Rhinitis

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109: Thyroid

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47: Nasal Obstruction and Rhinorrhea

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59: Tumors of the Nose, Paranasal Sinuses, and Nasopharynx

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48: Epistaxis
77: Foreign Bodies of the Pharynx and Esophagus

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49: Pediatric Headaches

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67: Dental and Gingival Disorders

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8: Psychiatric Disorders in Pediatric Otolaryngology

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111: Craniofacial Development and Congenital Anomaly: A Contemporary Review of Processes and Pathogenesis

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74: Pediatric Dysphagia

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Children's Medical Center
Dallas, TX
65: Tonsillectomy and Adenoidectomy

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34: Congenital Anomalies of the External and Middle Ears

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33: Cochlear Implants in Children

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33: Cochlear Implants in Children

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75: Functional Abnormalities of the Esophagus

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42: Tumors of the Ear and Temporal Bone

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44: Nasal Physiology

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67: Dental and Gingival Disorders

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122: Auditory Access to Language Resulting from Cochlear Implant Technology

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118: Pediatric Voice Disorders: Evaluation and Treatment

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5: Ethical Issues in Pediatric Otolaryngology

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Joe DiMaggio Children's Hospital, Hollywood, Florida
Affiliate Assistant Professor
Florida Atlantic University Charles E. Schmidt College
of Medicine
Boca Raton, Florida
94: Pediatric Airway Stenosis: Minimally Invasive Approaches

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20: Embryology and Developmental Anatomy of the Ear

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80: Neurologic Disorders of the Mouth, Pharynx, and Esophagus

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115: Pediatric Skull Base Surgery

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55: Complications of Nasal and Sinus Infections

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41: Injuries of the Ear and Temporal Bone

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57: Foreign Bodies of the Nose

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64: Inflammatory Disease of the Mouth and Pharynx

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97: Diagnosis and Management of Pediatric Laryngotracheal Trauma

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15: Antimicrobial Agents for the Treatment of Pediatric Head and Neck Infections

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102: Imaging of Pediatric Neck Masses

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120: Amplification Selection for Children With Hearing Impairment

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115: Pediatric Skull Base Surgery

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Pediatric Neuroradiologist and Pediatric Radiologist
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24: Methods of Examination: Radiologic Aspects
84: Radiologic Evaluation of the Pediatric Airway

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Director Pediatric Otolaryngology
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16: The Role of Biofilms in Pediatric Otolaryngologic Diseases

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62: Methods of Examination of the Mouth, Pharynx, and Esophagus

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117: Velopharyngeal Insufficiency

Seth M. Pransky, MD
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98: Tumors of the Larynx, Trachea, and Bronchi

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121: Behavorial Intervention and Education of Children With Hearing Loss

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Center for Genetic Medicine Research (CGMR)
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40: Diseases of the Labyrinthine Capsule

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95: Airway Surgery: Open Approach

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17: Pediatric Gastroenterology

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Northwestern University Feinberg School of Medicine
Ann & Robert H. Lurie Children's Hospital of Chicago
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109: Thyroid

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100: The Neck: Embryology and Anatomy

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120: Amplification Selection for Children With Hearing Impairment

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97: Diagnosis and Management of Pediatric Laryngotracheal Trauma

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University of Pittsburgh School of Medicine
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14: Pediatric Hematology: The Coagulation System and Associated Disorders

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106: Head and Neck Space Infections

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43: Embryology and Anatomy of the Paranasal Sinuses

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70: Oral Cavity and Oropharyngeal Manifestations of Systemic Disease

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105: Cervical Adenopathy

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1: Evolution of Pediatric Otolaryngology

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Orlando, FL
19: Pediatric Oral and Maxillofacial Surgery: Craniofacial Growth and Interdisciplinary Surgical Care

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64: Inflammatory Disease of the Mouth and Pharynx

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95: Airway Surgery: Open Approach

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44: Nasal Physiology

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34: Congenital Anomalies of the External and Middle Ears

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47: Nasal Obstruction and Rhinorrhea

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39: Facial Paralysis in Children

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50: Oral and Facial Neuropathic Pain in Children

Rahul K. Shah, MD, FACS, FAAP
Associate Professor, Departments of Pediatrics and Surgery
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Children's National Medical Center
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6: Professionalism, Communication, and Teamwork in Surgery
40: Diseases of the Labyrinthine Capsule

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51: Orbital Swellings

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58: Injuries of the Nose, Facial Bones and Paransal Sinuses

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30: Genetic Hearing Loss and Inner Ear Diseases

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4: Outcomes and Evidence-Based Medicine in Pediatric Otolaryngology

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100: The Neck: Embryology and Anatomy

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70: Oral Cavity and Oropharyngeal Manifestations of Systemic Disease
78: Trauma to the Mouth, Pharynx, and Esophagus in Children
79: Caustic Injuries and Acquired Strictures of the Esophagus

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61: Physiology of the Mouth, Pharynx, and Esophagus

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112: Primary Care of Infants and Children With Cleft Palate

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11: Allergy and Immunology

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30: Genetic Hearing Loss and Inner Ear Diseases

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115: Pediatric Skull Base Surgery

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18: Pediatric Pulmonology

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120: Amplification Selection for Children With Hearing Impairment

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88: Airway and Laryngotracheal Manifestations of Gastroesophageal Reflux Disease

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97: Diagnosis and Management of Pediatric Laryngotracheal Trauma

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62: Methods of Examination of the Mouth, Pharynx, and Esophagus

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60: Embryology and Anatomy of the Mouth, Pharynx, and Esophagus

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2: Phylogenetic Aspects and Embryology
3: Genetics, Syndromology, and Craniofacial Anomalies

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40: Diseases of the Labyrinthine Capsule

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53: Rhinitis and Acute and Chronic Sinusitis

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2: Phylogenetic Aspects and Embryology
3: Genetics, Syndromology, and Craniofacial Anomalies

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107: Benign Tumors of the Head and Neck

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99: Laser Surgery

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93: Pediatric Tracheotomy

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25: Vestibular Evaluation

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108: Malignant Tumors of the Head and Neck

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Professor, Otolaryngology–Head and Neck Surgery
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74: Pediatric Dysphagia
117: Velopharyngeal Insufficiency

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Director, Pulmonary Bronchology
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82: Physiology of the Larynx, Airways, and Lungs

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118: Pediatric Voice Disorders: Evaluation and Treatment

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116: Disorders of Language, Phonology, Fluency, and Voice in Children: Indicators for Referral

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36: Diseases of the External Ear

S. James Zinreich, MD
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Johns Hopkins Hospital
Baltimore, MD
43: Embryology and Anatomy of the Paranasal Sinuses

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9: Munchausen Syndrome by Proxy

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62: Methods of Examination of the Mouth, Pharynx, and Esophagus

Foreword

In 1972, I accepted the position of first academic chairman of the Department of Otolaryngology at the University of Pittsburgh. I realized that the two greatest regional assets that could contribute to developing the most outstanding otolaryngology department in the country were the Eye and Ear Hospital of Pittsburgh and the Children's Hospital of Pittsburgh next door. I was determined to create a Department of Pediatric Otolaryngology, although at that time Children's Hospital in Boston had the only such department. My training at the Massachusetts Eye and Ear Infirmary and rotations at Children's Hospital Boston had shown me the enormous value of developing professionals dedicated to caring for children with diseases of the ear, nose, and throat.

One of the first doctors I recruited to the faculty was Charles D. Bluestone, whose passion and enthusiasm for pediatric otolaryngology was boundless. He promised to make the Department of Otolaryngology at Children's Hospital of Pittsburgh the best in the world. Shortly after his arrival, we recruited Sylvan Stool to join us, and the team of Bluestone and Stool became recognized as the founding fathers of the specialty of pediatric otolaryngology. Our Department of Pediatric Otolaryngology offers a full range of expertise in the field, but there is a special emphasis on Bluestone's particular area of research and interest—the problems of otitis media and middle-ear effusion. He founded the Otitis Media Research Center (generously funded by the National Institutes of Health since 1978), which has a multidisciplinary research team combining the efforts of both basic and clinical scientists to develop new hypotheses which have resulted in improved patient care methods for children with diseases of the middle ear.

The first edition of *Pediatric Otolaryngology* by Bluestone and Stool, published exactly 30 years ago in 1983, was the first book dedicated specifically to pediatric otolaryngology. It was a single-volume text oriented to diseases in this specialty and emphasizing concepts rather than techniques. The authors were the "all-star" team of otolaryngology, since pediatric otolaryngology was not yet recognized as a subspecialty. Colleagues from other specialties wrote outstanding chapters in related fields.

The 5th edition of the book has doubled in size and is now a two-volume text, including a new section devoted to basic science, general pediatric otolaryngology and other pediatric subspecialty areas. Unfortunately, many of the original authors have retired or are in the "big operating room in the sky," but an impressive group of new authors has contributed chapters. Many of these authors are graduates of the two-year Fellowship Training Program established by Bluestone and Stool. They now serve as chiefs of divisions of pediatric otolaryngology and as department chairs. Diagnostic imaging chapters in each subspecialty section present hundreds of new diagnostic CTs and MRIs. Such chapters were nonexistent in the 1st edition, because CT scans and other imaging techniques had not yet come into general use.

Dr. Charles Ferguson, senior otolaryngologist at Children's Hospital in Boston, wrote the foreword to the first edition in 1983. He was a pioneer in the field of otolaryngic care of children, and I had the good fortune to meet him during my residency at Massachusetts Eye and Ear Infirmary. We became good friends and colleagues. In that first foreword, he wrote: "The evolution of Pediatric Otolaryngology as a true subspecialty is a lifetime dream fulfilled. It is most exciting and also gratifying to know that there are now over a score of otolaryngologists who devote over 80% of their professional time to this specialty."

It has been extraordinary to witness the tremendous growth of our specialty with thousands of pediatric otolaryngologists around the world and the growth of knowledge that has greatly improved the care of children with pediatric otolaryngic disorders. This 5th edition reflects the accumulated knowledge and clinical skill acquired over the past 30 years. I congratulate Dr. Bluestone and his team for persevering and producing another outstanding edition of this extraordinary text.

Eugene N. Myers, MD, FACS, FRCS Edin (Hon)
Distinguished Professor and Emeritus Chair
Department of Otolarygnology
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Pittsburgh, Pennsylvania

Preface

It is with great pride and excitement that we publish this fifth edition of *Pediatric Otolaryngology* three decades after the first edition in 1983. This edition reflects the current state of knowledge and practice in pediatric otolaryngology, a subspecialty that has dramatically grown along with our textbook. This edition includes 29 new chapters and 141 new authors (217 total authors), a new section on Basic Science and General Pediatric Otolaryngology, and new addition of color. All authors and the 14 section editors (names listed on the front cover and title page) are authorities in their respective fields. Without the hard work and devotion of these section editors, this fifth edition would not have become a reality. They devoted countless hours recruiting new authors and peer-reviewing and editing each chapter.

We sadly regret that the late Sylvan E. Stool, coeditor of the first four editions, could not witness the amazing maturation of Pediatric Otolaryngology and the publication of this edition. In his honor, we have included his Encomium to remind those who knew him of his tremendous accomplishments and contributions to our subspecialty and to enlighten those too young to have benefitted from his ever-present warm friendship and intellect.

We give special thanks to Eugene N. Myers who graciously wrote the Foreword as he is uniquely qualified to reflect not only on the progress in the field of pediatric otolaryngology over the past 40 years but also the growth and development of this textbook. We thank our editors at PMPH-USA, Carole Wonsiewicz and Linda Mehta, whose expertise and attention to detail was invaluable. We are indebted to Deborah Buza, administrative assistant, for her dedication and persistence over the past three years tracking all chapters, keeping authors informed, and the production schedule going, and also to Maria B. Bluestone who provided expert and invaluable editorial aid for several chapters.

As editors, we hope the health care of infants and children will be improved by those healthcare professionals who use this textbook as a reference in this 21st century.

Charles D. Bluestone, MD, FACS, FAAP
Jeffrey P. Simons, MD, FACS, FAAP
Gerald B. Healy, MD, FACS

Acknowledgments

I personally want to thank and acknowledge the work and dedication of my two new editors, Gerald Healy and Jeffrey Simons, who accepted my invitation to join in preparing this fifth edition and carry on the role of Sylvan Stool in the first four editions. I consider Gerry a dear friend and colleague and believe I might have some minor influence in his choosing pediatric otolaryngology as a career path as I suggested that to him back in the early 1970s while we were in Boston. He followed me as Chief of Otolaryngology at Boston City Hospital but went on to develop the prestigious Department of Pediatric Otolaryngology at Boston Children's Hospital, become a professor at Harvard, and the only otolaryngologist to serve as President of the American College of Surgeons in its 100-year history. Many of the new authors in this edition are from his former department at Harvard. Jeffrey I consider to be a rising star in the new generation of pediatric otolaryngologists and without his painstaking efforts to organize and recruit section editors and authors, and edit chapters, this edition would never have become a reality. My sincere hope is that he will carry the textbook to new and better editions in the future.

Charles D. Bluestone

We wish to acknowledge the chapter contributions of the distinguished and dedicated authors (217) and the 14 section editors who made this fifth edition possible. Thank you to Eugene N. Myers for his gracious Foreword. We give special thanks to Deborah Buza for her dedication and commitment to the coordination and collation of manuscripts and for her kind and compassionate but persistent reminders to authors and section editors to adhere to our publisher's production schedule. A special thanks also goes to Carole Wonsiewicz, Development Editor at our publisher, PMPH-USA, and Maria B. Bluestone who provided her expert and invaluable editorial expertise for several chapters. Finally, we thank our mentors and our patients and families who allow us to learn from them and pass the knowledge on to our colleagues and students.

Charles D. Bluestone
Gerald B. Healy
Jeffrey P. Simons

Encomium

Sylvan E. Stool, MD (1925–2004)

It is my honor to dedicate this fifth edition of *Pediatric Otolaryngology* to the memory of the late Sylvan E. Stool, MD, pioneer in Pediatric Otolaryngology and my coeditor of the first four editions. Sylvan not only dedicated his career to providing health care to infants and children with ear, nose, and throat diseases and disorders but also was committed to teaching students, residents, and fellows, many of whom are now leaders in our field. He was board-certified in both pediatrics and otolaryngology, a distinction that was, and still is, rare in Pediatric Otolaryngology.

He was born on November 7, 1925, in San Angelo, Texas, where his parents were in the dry goods business and grew up on the dusty plains of west Texas. Sylvan attended high school in Abilene, which was then the University of Texas in Austin. He was accepted into an accelerated program at Southwestern Medical College designed to fill the shortage of physicians during and after World War II and graduated with an MD on June 3, 1947, at the age of 20. This was the first year that the Hippocratic Oath was administered at medical school graduations (*UT Southwestern Medical Center: Commemorating the First Fifty Years*, p.11). After completing a two-year rotating internship and residency in general practice in Dallas and Ft. Worth, Sylvan completed a one-year fellowship in Pediatric Surgery at the Children's Orthopedic Hospital in Seattle. After he could obtain no further training in surgery, he decided to pursue a residency in pediatrics at the University of Utah.

He was a captain in the United States Air Force during the Korean War and stationed in hospitals in Guam and Japan. After the war, he received an appointment as a Fellow in Boston Children's Medical Center. It was there that Sylvan had his first exposure to otolaryngology while filling in for a sick colleague. This position allowed him to work closely with Drs. Charles Ferguson and Carlyle Flake, two of the few early otolaryngologists who worked in Children's Hospital. He was able to live in the house officers' quarters and, as he described it, was allowed "to eat two free meals a day."

After his experience in Boston, Sylvan practiced pediatrics at Denver General Hospital in 1955 during which time he noticed the prevalence of ear and hearing problems in his patients and initiated an informal ENT clinic. He then realized that he needed further formal instruction in otolaryngology. When Dr. Victor Hillyard was appointed Chief of Otolaryngology at the University of Colorado Hospital, Sylvan inquired about training in otolaryngology, and Dr. Hillyard immediately offered him a residency position. (Dr. Hillyard and Sylvan received a grant from NIH and Fitzsimmons Army Hospital to fund his training.)

Sylvan and I first met in 1961 while we were both residents in otolaryngology; he at the University of Colorado and I at the University of Illinois. We met while attending the national meeting of the American Medical Association in Denver, and he expressed such a keen interest in my exhibit on tracheobronchial mechanics that we immediately became life-long friends (Fig. 1).

The late Dr. C. Everett Koop, a pioneer in pediatric surgery and later the celebrated US Surgeon General, was appointed Chief of Surgery and was charged with staffing

FIGURE 1. Sylvan and Charley in Pittsburgh.

Children's Hospital of Philadelphia (CHP) with specialists in pediatrics. Following Sylvan's residency in 1963, he received an inquiry from CHP to help Mary Ames establish a rehabilitation center to serve children with multiple defects. For Sylvan, Philadelphia was a much different environment from the West. He found the East Coast medical community more set in its ways and resistant to changing established medical fields, for example, only the Jackson-trained bronchoesophagologists performed endoscopy and many had difficulty accepting the concept of age-related specialist in otolaryngology. After a few years, the otolaryngology training programs in Philadelphia recognized opportunities in pediatric otolaryngology, and their residents requested rotations with Sylvan at the Children's Hospital where he was the Director of Otolaryngology for 12 years.

Sylvan then realized that unless Pediatric Otolaryngology, similar to Pediatric Surgery as led by Dr. Koop, achieved academic recognition and the ability to train fellows, it could never be established as a true subspecialty. It was then that Sylvan and I discussed the possibility of initiating a Fellowship in Pediatric Otolaryngology in Pittsburgh, since at that time it seemed impossible to achieve such a fellowship in Philadelphia. In 1975, Sylvan accepted an offer from Dr. Eugene N. Myers, the first Academic Chairman of the Department of Otolaryngology at the University of Pittsburgh School Medicine, to join me at the Children's Hospital of Pittsburgh. Sylvan was a tenured Professor of Otolaryngology and Pediatrics at the Medical School and Director of Education in the Division at Children's Hospital of Pittsburgh and remained until 1994.

The first Fellow in Otolaryngology was recruited in 1975, funded by Children's Hospital for one year. In 1985, Sylvan became Principal Investigator for a training grant from the National Institutes of Health to fund a research fellowship year in Pediatric Otolaryngology, and this research year combined with the clinical year initiated our two-year Fellowship in Pediatric Otolaryngology. While Sylvan was in our Department, 40 Fellows were trained in Pittsburgh. Most of the graduates are now in academic medicine, many are Directors of Divisions of Pediatric Otolaryngology, and some chairs of departments. During Sylvan's years in Pittsburgh we invited the only 20 Pediatric Otolaryngologists we knew in the United States and Canada to form the Study Club (Fig. 2).

In 1994, Sylvan returned to Denver invited by one of his Fellows, Kenny Chan to join him at The Children's Hospital where he held the title of Senior Lecturer at the University of Colorado School of Medicine. He worked part-time in the clinic and the operating room and occasionally on-call duties to help out. He continued to engage residents in various research projects with a focus on toy safety. His "Otitis Media Workshop" became well-known all over the Rocky Mountain region and beyond. His Colorado years afforded many opportunities to reacquaint with former pediatric colleagues and friends from the 1960s. One of his memorable presentations, entitled "The Golden Years of Otolaryngology," was delivered

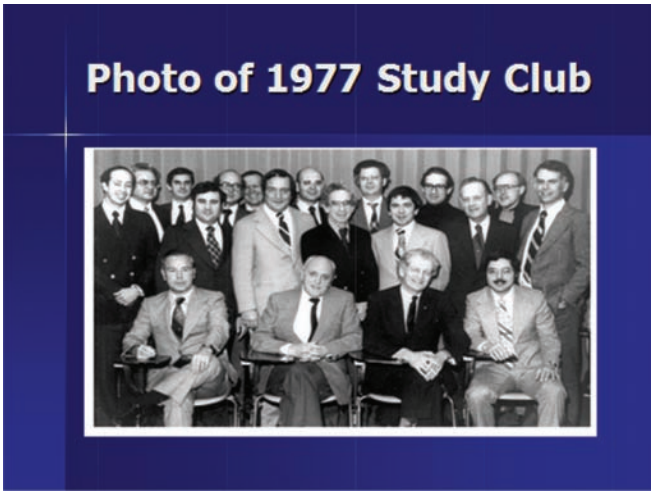


FIGURE 2. Initial 1977 meeting of 20 pediatric otolaryngologists in Pittsburgh. The outcome of this meeting was the formation of the Otolaryngology and Bronchoesophagologic Section of the American Academy of Pediatrics.

in 2002 at the Western Society of Pediatric Otolaryngology Meeting, where there were few dry eyes in the audience. His primary clinical interest was in management of the pediatric airway, and during the last few years of his illustrious career focused on the prevention of obstruction of the airway from foreign objects, primarily potentially dangerous toys. In 1968, Sylvan made his most important and lasting contribution to the health and well-being of children when he introduced the life-saving stay sutures to the tracheotomy procedure that is now the standard of care for children. Sylvan suffered a fatal heart attack in 2004 and "died with his boots on" placing a set of tympanostomy tubes in a cleft palate patient on the day he died. He was the first to report that almost all infants with unrepaired cleft palates had chronic middle-ear effusion.

His accomplishments and contributions to medicine are numerous. He published more than 150 peer-reviewed articles and 70 publications. Sylvan was past president of the Society for Ear, Nose and Throat Advances in Children (SENTAC), and in 1995, the Society established the Sylvan E. Stool Lectureship. Also in 1995, the Department of Otolaryngology established the Sylvan E. Stool Lectureship in the Carol F. Reynolds History of Medicine Society at the University of Pittsburgh School of Medicine in honor of his contributions to the history of medicine. Sylvan received the Humanitarian Award (2000) from the American Academy of Otolaryngology–Head & Neck Surgery for his commitment to teaching pneumatic otoscopy, as part of Otitis Media Workshops, not only in the United States but in many other countries, primarily those in Latin America. He made more than 30 trips to teach about otitis media with didactic and "hands-on" teaching styles. The Latin American efforts were recognized by both the World Health Organization and the Pan American Health Organization as the model of how to train local care providers.[1] Just in Mexico alone these efforts

led to (1) placement of otoscopes in all public health clinics in the country, (2) making otitis media a reportable disease in Mexico, and (3) focusing on national vaccination policy that included ear disease. This work also led to the creation of the vibrant educational organization—the Inter-American Association of Pediatric Otolaryngology—of which Sylvan was the first President.

His personal life was dynamic and full as well. His eldest daughter, Evelyn, described her father's interests as very diverse and extending well beyond medicine but always tangentially associated with it. He became fascinated in Western art through an unexpected discovery of an old book, *Shut Your Mouth and Save Your Life* by George Catlin. Catlin was a painter who had observed the breathing habits of the Plains Indians while traveling with them and painting their portraits. Sylvan became an expert on George Catlin's own health issues and well respected in the Western art community as one of few Catlin experts. Much of Sylvan's joy and passion resulted from teaching, whether in the clinic, the operating room, an airstrip in the south Pacific or a hospital in South America. He traveled all over the world teaching doctors how to diagnose otitis media and practice airway safety. His wife, June Keil, traveled the world with him. They met in 1955 at a music appreciation class and married later that year. They had a wonderful marriage of 48 years produced four children, Evelyn, Daniel, Laura, and Karen, two grandsons, Lloyd and Sander, and much happiness and lots and lots of music.

Charles D. Bluestone
April 5, 2013

1. Eavey RD, Santos JI, Arriaga MA, et al. An educational model for otitis media field-tested in Latin America. *Otolaryngol Head Neck Surg* 1993;109:895-898.

Acknowledgments

I am indebted to Evelyn Stool Waldren, for her contribution to this, her father's, biography, Kenny Chan for his memory of Sylvan's 10 years in Denver, Roland Eavey's remembrances of his teaching in Latin America, Eugene N. Myers for his important additions and editing, and to Sylvan's own reflections in the fourth edition of *Pediatric Otolaryngology* (pp. 62–63).

SECTION 1

Basic Science/General Pediatric Otolaryngology

Michael J. Cunningham and Joseph E. Dohar

1

CHAPTER

Evolution of Pediatric Otolaryngology

Robert J. Ruben

Pediatric otolaryngology (ORL) evolved through a synergy of perceived societal needs and availability of acceptable interventions, relating to the economic, social, and philosophical concept of childhood.[1,2] The history and conceptualization of childhood can be usefully divided into four overlapping but distinct ideological periods.[3] The first, from 1600 to the 1750s, is the end of the Reformation and the period of the Counter-Reformation; the second, from the 1750s to the 1850s, is the Enlightenment; the third, from the 1860s to the 1920s, is the Romantic period; and the last from the 1920s to the present is the period of Entitlement.

REFORMATION

The Reformation and Counter-Reformation are characterized by concern with the child's soul, exemplified in the later writings of John Locke,[4] which was considered as either sinful (Reformation) or pure (Counter-Reformation). As this was a period of great economic disparities, the limited medical knowledge available was applied for the benefit of only a very few. The child, at this time and until the end of the Romantic period, was viewed economically as a producer, not as a consumer; that is, in all classes, the child was expected to augment the family economically. For this reason, boys were favored over girls. If the male child was high born, then he was trained to be a ruler and/or a warrior. A female was valued in terms of her potential economic benefit to the family through marriage and was trained to optimize "her chances" and her usefulness to the family after her marriage. When born to peasant, males and females soon became productive workers and ultimately served as the old-age security for parent(s). In this context, most infants with otolaryngic diseases and disorders died or were abandoned.[5]

Two instances during the 17th century, each involving a child of a family of substantial wealth and political power, are illuminating. The first concerns the infant Dauphin of France, who became Louis XIII.[6] Dr. Jean Héroard, his physician, kept a daily diary concerning the care of this very special patient, slated to rule. In 1601, at 1 day of age, his condition was noted:

> *September 28th. His nurse was demoiselle Marguerite Hotman and as he seemed to have some difficulty in sucking his mouth was examined and it was found that he was tongue-tie; so at five o'clock in the evening M. Guillemeau, the Kings surgeon, cut the tendon three times.*[6]

Two weeks later, there is an outcome report to this surgical procedure for questionable pathology:

> *when he sucks it is in great gulps so that he swallows as much in one gulp as other babies in three. His nurse never has enough for him.*[6]

The second case, published in 1620,[7] concerns the education of Luis, the congenitally deaf son of the Captain of Castile, in whose interbred family there were numerous deaf relatives. A significant motivation for pursuing the child's education was the need for Luis to take communion, so that he could be a "legal person; then his mother, Doña Juana de Córdoba, Duchess of Frias, could be regent until Luis came of age and thereby control the patrimony. The treatment of Luis, described in Bonet's 1620 publication, was based on an earlier lost manuscript, *Doctrina para los mudos sordo*, attributed to the Benedictine, Pedro Ponce de León (d. 1584), who had educated a number of the deaf Spanish nobility.

These 17th-century pediatric otolaryngic interventions, one surgical and the other habilitative, reflect the status of the child. First, both had been baptized so as to be a full member of the Church. Then, and foremost, the young child was an economic producer. It was important for these highborn children to survive—for the Dauphin, to rule, and for the Spanish Luis, to keep the fortune in the family. They are characteristic of the times in that these interventions were confined to the very wealthy. For most children of the 17th century, there were neither surgeons nor deaf educators.[8]

ENLIGHTENMENT

The Enlightenment saw a radical change in the concept of childhood, from one that considered the child as a little adult with either a sinful or a pure soul to one in which the child was essentially different from the adult in that each child was thought to come into this world with the mind as a blank slate, the famous "tabula rasa." In a sense, "the child" was born during the period of the Enlightenment. Rousseau comments on the residual old view, and points the way to the new one, in his preface to Émile[9]:

> *The wisest writers devote themselves to what a man ought to know, without asking what a child is capable of learning. They are always looking for the man in the child without considering what he is before he becomes a man.*

Childhood in the Enlightenment is seen as a unique condition of life, and the child's own "natural" course of

development should be the basis for education. This new view leads to a new invention, the children's book, one of the first being published by John Newberry in 1744.[10] Economically, however, this conceptual change did not alter the child's fundamental role as a producer, a role that becomes even more onerous with the advent of the industrial revolution. On the contrary, the view that the value of a child rests on his or her productivity was somewhat mitigated by the political and social revolutions at the end of the 18th century, and at this time, otolaryngic care of children became available to a somewhat larger segment of the population.

Important changes in the otolaryngic care of the child began with the Abbé Charles Michel de L'Épée who undertook the teaching of two middle-class deaf sisters by means of signs to enable them to take communion.[11] He expanded his teaching, at his own expense, to include a number of poor, possibly abandoned, deaf Parisian children. In 1791, Louis the XVI, the descendent of the possibly tongue-tied Dauphin, established the first state school for the deaf, open to all[12] (Fig. 1-1). Its development was furthered by the Abbé Sicard who persuaded the revolutionary National Assembly that aid for the handicapped was part of the "natural duties" encompassed by the "rights of man."[13] It is interesting to consider the way this view was congruent with those of Danton and Robespierre, who believed that children belonged to the state before they belonged to their families.[3]

The responsibility of the state to care for the deaf rapidly spread throughout Europe and North America. Deaf children, by the end of the Enlightenment, were cared for, normatively if not in all cases, regardless of their social status.

The Connecticut Asylum for the Education of Deaf and Dumb Persons—now the American School for the Deaf in Hartford, CT—was opened on April 15, 1817, the first such institution in the United States. The second was the New York Institution for the Instruction of the Deaf and Dumb; this free school for all deaf children of the state over five years old was incorporated on April 15, 1817, and opened on May 20, 1818, in a room in an almshouse. Before opening such a school in New York, the organizing committee needed to determine the number of deaf children in the city and chose Dr. Samuel Mitchell to assess this. His pamphlet[14,15] showed the current number of deaf children as 63 in New York City with 8 more in the vicinity and provided a reasonable prediction for the near future of more than 100. In part of this basis, the committee then obtained funding from the government and philanthropists and enrolled the first four pupils in 1818.


DÉPARTEMENT DU VAR.

LOI

Relative à M. l'Abbé de l'Epée, & à son établissement en faveur des Sourds & Muets.

FIGURE 1-1. Loi Relative à M. l'Abbé de l'Épée, & à son établissement en faveur des Sours & Muets, passed by the National Assembly, Paris, July 29, 1791. Département du Varennes. Original document of the enacted legislation authorizing the establishment of a school for the deaf and appropriating 12,700 livres for expenses. This was dedicated to L'Épée, who died in 1789, and sanctioned by Louis XVI, at the time a constitutional monarch and a virtual prisoner in the Tuilleries. This was the first state-sponsored school for the deaf and was open to all.

Conversely, the industrial revolution, taking hold in the later years of the Enlightenment, increased the need for economic productivity for many children. It is in this period that 18th-century ideas, rooted in John Locke, were instituted as noted by Jonas Hanway as early as 1766:

> *That poor children should be put to work at age 3 with daily bread and in cold weather, if thought to be needed, a little warm gruel*. (p. 138)[3]

The importance of child labor in economic development can be seen, for example, in the employment records of the Manchester cotton mills in the 1830s; 76% of females working in these mills were girls under 14 years, and 61% of males were boys under 14 years.[3] These children did not have access to medical, let alone otolaryngic, care.

ROMANTIC

During the Romantic period of the 19th and early 20th centuries, both negative and positive experiences of childhood moved hand in hand. Some societal forces were working to better the child's lot; for the first time, many children become in part consumers, while also maintaining their role as economic providers in most families. A great advance witnessed by this period was that orphan asylums were supplanted by children's hospitals.[3] In 1802, for example, the Hôpital des Enfants-Malades opened in Paris in the former Maison de l'Enfant-Jésus that had been founded in 1724 as an orphan asylum for abandoned girls. The middle of the 19th century saw an increased pace in the establishment of children's hospitals, such as Great Ormond Street Hospital for Children opened in 1852 and the Children's Hospital of Philadelphia opened in 1854, and subsequently several other major North American institutions were established including Boston Children's Hospital in 1869, The Hospital for Sick Children, Toronto, in 1875, and the Children's Hospital Los Angeles that was incorporated as the Children's Hospital Society of Los Angeles in 1901.[16]

John Snow of London demonstrated that anesthesia could be used in children; by 1857, he had anesthetized 186 children under the age of 1 year with chloroform.[17]

Wilhelm Meyer discovered the disease process of the adenoid[18] in 1868 and its relationship not only to otitis but also to mouth breathing, sleep disturbance, sluggish facial expression, and fatigue. Meyer's work provided a rationale for innovations directed at improving and optimizing

the health and appearance of the child; these ameliorations are described often in pediatric and otolaryngic literature from the end of the 19th century into the first half of the 20th century. Hypertrophy of the tonsils and adenoid with incomplete and faulty ventilation, or acid secretions of the tonsil and adenoid as a cause of decreased appetite and subsequent malnutrition, were standard diagnoses during the first decades of the 20th century. Malnutrition, which is a very unusual indication today, was a common rationale for tonsillectomy and adenoidectomy up to the end of the 1920s.

The perfectionist ideology that was to an extent a result of the romantic mind-set played a significant role in the quest for "normalcy." Deviance was disparaged, and the normal was thought to be ultimately achievable through eugenics. One of the more benign examples of this attitude was the state-sponsored "better baby contests" held in the midwestern United States from 1920 to 1935 (Fig. 1-2).[19,20] It became incumbent for the parent to do all that could be done for their child to be as normal as possible, and tonsillectomy and adenoidectomy were recommended by health providers, physicians, and public health workers to encourage full physical and mental development. Consequently, any child whose growth and development was not at "normal" became a potential candidate for this operation.

A significant and efficacious advance in the ORL care of children in North America came about from the need for intervention for children with diphtheria, the most deadly pediatric otolaryngic condition of the 19th and early 20th centuries. The diphtheritic child would either suffocate or undergo myocarditis with cardiac arrest until Joseph O'Dwyer published his method of intubation in "Two cases of croup treated by tubage of the glottis"[21] in 1885; he followed up this landmark description with the publication of an additional 50 cases[22] in 1888.

O'Dwyer's work facilitated the acceptance of peroral endoscopy. The North American leader in this respect was Chevalier Jackson, whose attention was drawn to numerous laryngeal, tracheal, bronchial, and esophageal foreign bodies in children, culminated in the publication of his monograph on foreign bodies[23] in 1936. Jackson's work with peroral endoscopy exposed him to a large number of esophageal strictures from lye (sodium hydroxide) ingestion. He became a children's advocate and was instrumental in the passage of correct product labeling for containers with lye and other poisons[24]—the Federal Caustic Labeling Act of 1927 (Fig. 1-3).

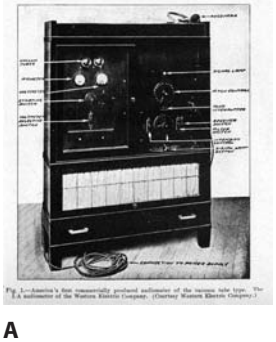
Although concern for the hearing of schoolchildren existed from the beginning of the 20th century, there was no accurate method for hearing assessment. The major advance in the diagnosis and care of children's hearing was the development of the first commercial vacuum tube audiometer, the Western Electric 1A, by Harvey Flecther[25] and introduced as a clinical tool by E. P. Fowler Jr. and R. L. Wengel[26,27] in 1922. The use of this device to objectively test the hearing of school children resulted in the 1928 Fowler article entitled "Three million deafened school children."[28] This article was a major factor in the establishment of childhood hearing screening programs in the public school system (Fig. 1-4A and B).

ENTITLEMENT

The political and philosophical ideals and aspirations of the mid-19th century, unsuccessfully expressed in the revolutions of 1848, but sustained and refined up to the present, have resulted in our present day principle of entitlement. The view

FIGURE 1-3. "From a photographic of a child fatally burned by swallowing Red Star Lye. The lower part of the illustration shows the inadequacy of the warning common to all labels of lye containers sold in groceries and used in kitchens. Parents are not aware of the danger of leaving the lye-preparations in the reach of children. This label is removed to get at the directions on the back, and removal usually destroys or removes the tiny, inconspicuous vertical cautionary wording."[24]

FIGURE 1-2. Spectators watching the various testing and measurement tables at the 1930 contest. (Photo courtesy of the Indiana State Archives, Indiana Commission on Public Records.[19])



A

B

FIGURE 1-4. *A*, America's first commercially produced audiometer of the vacuum tube type. The I-A audiometer of the Western Eclectic Company.[63] *B*, School testing with the Western Electric 4-A audiometer.[63]

has been generalized that all children are entitled to life and to the fulfillment of their potential. Otolaryngic care is accessible to all children throughout much of the industrialized and postindustrialized world through various private and public state plans. From the early role as producer, the child has now become a consumer. For a glimpse of this trend beyond medicine, Disney products sold $10 million in 1933 and $3 billion in 1990 representing a 40-fold increase when corrected for inflation.[3] In matters of health, there is a tension between parental control and state control, often resolved in favor of the state.

A consequence of entitlement in synergy with medical advance can be found in the care of the premature infant.[20,29] The first reported use of an incubator to care for a premature infant was by Carl Credé in Germany in 1837. This use was rediscovered in 1880 by Stéphane Tarnier[3] who, observing chicken incubators in the Paris Zoo, applied the process tothe premature infants in the Paris Maternity hospital (Fig. 1-5). These first incubators held, like those for the chickens, multiple infants. The mortality of prematurity, despite the use of warmth, still remained quite high. The highest cause of mortality occurring in infants born before the seventh month of gestation was found by Avery in 1959 to be lack of surfactant resulting in hyaline membrane disease.[30] The solution was to increase either ventilation by mechanical means or the amount of oxygen available. The latter resulted in the ophthalmologic condition retrolental fibroplasia and its associated blindness, furthering the need for a localized delivery system. In the 1940s and the 1950s, most assisted ventilation was delivered by tank-type negative pressure ventilators such as the drinker respirator used for polio victims. The European polio epidemic of 1952 overwhelmed the supply of the iron lung-type negative respirators. To meet this problem, the physicians of Copenhagen performed tracheotomies, and the medical students hand ventilated the patients with positive pressure. This was successful, leading to the development of positive pressure respirators for the intubated premature infant.[29] The infants survived but could not be extubated as they had acquired subglottic stenosis and required tracheostomies that had their own associated morbidity and mortality.

This new morbidity—acquired subglottic stenosis of infancy—required and received an effective intervention, laryngotracheal reconstruction, pioneered by two pediatric otolaryngologists Blair Fearon and Robin Cotton.[31,32] Thus, the entitlement of the premature baby created demand for specialized care of the infant airway; this was a salient factor that resulted in the formation of the specialty of pediatric ORL throughout the world,[33] because while there may have been a need heretofore, there was little or no demand.

NORTH AMERICA

In the late 1940s, three of the pioneering children's hospitals had physicians who concentrated their practices in pediatric ORL. Drs. Charles Ferguson and Carlyle Flake worked full time at Boston's Children's, with their wards dedicated to the

FIGURE 1-5. Trainer's incubators in use at the Maternité Hospital, Paris, 1884. (From the *Illustrated London News*, March 8, 1885: 228.)

treatment of croup and operating rooms on the same floor as their offices.[34] Dr. Seymour Cohen, whose major interest was pediatric endoscopy, practiced at the Los Angeles Children's Hospital. Dr. Blair Fearon, at Toronto's Sick Children's Hospital, practiced pediatric ORL and also, critically, undertook basic research with Dr. Robin Cotton in the reconstruction and repair of the infant airway. Their research resulted in the landmark paper entitled "Surgical correction of subglottic stenosis of the larynx: Preliminary report of an experimental surgical technique"[31] in 1972.

More North American physicians began to concentrate their practice to children, and there was a need to bring attention of this development to the otolaryngic community. Sylvan Stool (Fig. 1-6)[35] posted a notice at the 1971 meeting of the American Academy of Ophthalmology and Otolaryngology (AAOO) for all those interested in pediatric ORL to meet informally. Approximately 20 physicians attended this initial meeting, and it was decided to convene again at the AAOO 1972 meeting in Dallas. A decision was made at the1972 meeting to form a new society focused on pediatric ORL, and a small group was formed to write a set of bylaws and incorporate this new venture.

The new society was called The Society for Ear, Nose and Throat Advances in Children, Inc. (SENTAC) and was founded in 1973 as a nonprofit interdisciplinary professional organization. Its members were and continue to be otolaryngologists, pediatricians, surgeons, pediatric otolaryngologists, speech pathologists, audiologists, nurses, and basic scientists, all of whom are interested in enhancing the care of children with acquired or congenital disorders of the ear, nose, and throat. Dr. Robert Ruben was its first president. SENTAC continues to be an interdisciplinary forum for new ideas; it is one of the few medical societies in which membership is determined solely by interest, not by professional association, facilitating the successful interchange of information between many different professional and lay groups.

One year later in 1975, Dr. Basharat Jazbi organized the "First International Symposium on Pediatric Otorhinolaryngology," held in Kansas City, Missouri.[36,37] Following this was a course given at the Armed Forces of Institute Pathology in Washington, DC, in 1976 entitled "Pediatric Otolaryngic Pathology" organized by Captain Vincent J. Hyams, MC, USN. This, so far as can be determined, was the first such course ever given that systematically reviewed all that was known about the cellular pathology concerning pediatric otorhinolaryngology.[38]

FIGURE 1-6. Sylvan Stool, 1925–2004.

The Pediatric Otolaryngic Study Group began in 1977 with a meeting at the Pittsburgh Children's Hospital[39] hosted by Dr. Charles Bluestone and Sylvan Stool. There were 22 attendees at this meeting, and it was decided to organize a session on Pediatric Otolaryngology and Bronchoesophagology of the American Academy of Pediatrics, which resulted in the writing of a set of bylaws. This new organization would increase the recognition of ORL and bronchoesophagology by the pediatric medical community and provide a platform for the education of both pediatricians and otolaryngologists.

The study group continued to meet at different medical centers for the next few years. These meetings were informal and provided opportunities for participants to learn from their colleagues, which accelerated the dissemination of knowledge of advances in surgical techniques, instrumentation, diagnostic procedures, and effective interventions. It also allowed participants to see various clinical and administrative arrangements. Some examples of these meetings were as follows: Boston Children's Hospital,[40] hosted by Dr. Gerald Healy, in 1978 where new laser techniques were explored; Children's Memorial Hospital, Chicago,[41] hosted by Dr. Gabriel Tucker, Jr., in 1978 where pediatric endoscopy was demonstrated; Children's Hospital of Cincinnati,[42] hosted by Dr. Robin Cotton, in 1979; Children's Hospital of Philadelphia, hosted by Dr. William Potsic, in 1980[43]; and at the Albert Einstein College of Medicine (AECOM),[44] in the Bronx, New York, hosted by Dr. Robert Ruben, in 1981, where there was an emphasis on both communication disorders, including language, and cell biology. At the AECOM meeting, there was a special session to discuss the design of a cooperative study of medical therapy for respiratory papilloma. This resulted in the 1988 article "Treatment of recurrent respiratory papillomatosis with human leukocyte interferon. Results of a multicenter randomized clinical trial."[45]

A number of the members of the American Academy of Pediatrics Section of Otolaryngology and Bronchoesophagology perceived the need for a society that would be limited to those otolaryngologists who predominately practiced pediatric ORL and who demonstrated proficiency in this area. Their conception was a society modeled after the traditional ORL specialty societies such as the American Otological Society and the American Laryngological Society. At the 1979 San Francisco business meeting of the AAP Section of Otolaryngology and Bronchoesophagology, a committee was formed, chaired by Dr. Mark Richardson, to further explore this idea, formulate a set of bylaws, and incorporate the entity.[46] The new society was called the American Society of Pediatric Otolaryngology and held its first meeting in Bermuda in 1985 with Dr. Seymour Cohen as its first president (Fig. 1-7).

FIGURE 1-7. American Society of Pediatric Otolaryngology first meeting in Bermuda, 1985.

As pediatric ORL developed in North America, there was the desire and need to properly train physicians to become competent pediatric otolaryngologists. In 1975, Dr. Charles Bluestone, the first full-time director of the Department of Pediatric Otolaryngology at Children's Hospital of Pittsburgh, with Dr. Sylvan Stool, created the first pediatric ORL training fellowship program; this fellowship program has been funded by the National Institutes of Health since 1985. Several other programs were initiated, including that by Dr. Gerald Healy at the Boston Children's Hospital and by Dr. Robin Cotton at the Cincinnati Children's Hospital Medical Center. As more fellowships were established, there was a need for quality control and also a need for certification. This initially resulted in petitioning the American Council for Medical Education (ACGME) to standardize criteria for the training of a pediatric otolaryngologist and a process of accreditation of the training program by the ACGME. Currently, there are seven ACGME-accredited fellowship programs in pediatric ORL, including those at the University of Colorado in Denver, the George Washington University/Children's National Medical Center Program in Washington, DC, the Pediatric University of Iowa Hospitals and Clinics Program in Iowa City, the Cincinnati Children's Hospital Medical Center/University of Cincinnati College of Medicine Program, the Children's Hospital of Philadelphia Program in Philadelphia, the University of Pittsburgh Medical Center Medical Education Program in Pittsburgh, and the Baylor College of Medicine Program.[47] There are an additional 22 fellowship programs in North America.[48] As the number of applicants and fellowships increased, there was a need for a matching program. This was established in 1999. In 2000, there were 14 positions offered to 31 applicants, and in 2008, there were 35 positions offered to 48 applicants.[49] This more than twofold increase in positions is consistent with the overall shifting of physicians to specialization in American medicine.[50]

Pediatric ORL has evolved during the past three decades as a major medical discipline in North America. Almost every major pediatric hospital is now staffed by otolaryngologists well trained and experienced in the ear, nose, throat, head, and neck diseases and disorders of children.

EUROPE

Poland was the leader in the development of pediatric ORL in Europe, with the formation of specialty beginning there in the late 1940s, after the end of World War II.[51,52] Associate Professor Jan Danielewicz (Fig. 1-8), the father of pediatric ORL in Poland and one of its cofounders in Europe, established the first modern Department of Pediatric Laryngology at the hospital of Mother and Child in 1947, followed in 1956 at the Warsaw University Hospital. Danielewicz created both a training program and a specialty examination in and for pediatric ORL. The first pediatric ORL examination was held in 1961.

An initial "Days of Pediatric Laryngology" was held in 1958 in Zakopane, Poland, and has continued to be given every two years. Profs. Ewa Kossowska and Danielewicz went on to organize jointly the First European Congress of Pediatric Laryngologists in Warsaw in 1979.

On Prof. Danielewicz's retirement in 1973, Prof. Kossowska succeeded him. Under her leadership, the department focused on endoscopic surgery of the trachea and esophagus, laryngeal reconstructive surgery, sinonasal surgery, and the physiopathology of the upper respiratory tract and tonsillar infections. Prof. Kossowska retired in 1993 and was succeeded by Prof. Mieczyslaw Chmielik. Currently, Poland has four established clinical pediatric ORL departments: in Łódź, Poznań, Lublin, Białystok, and one that is in formation at Śląsk. The Polish Society of Pediatric Otorhinolaryngology is a registered organization. Specialized pediatric ORL training is open to all doctors who have completed their education in laryngology for two years and who pass an examination in pediatric ORL.[51]

In the context of the growing awareness of pediatric ORL as a significant specialty, Dr. Carlo Gatti Manacini, head of the Pediatric ORL Department in Brescia, initiated the idea of holding a World Pediatric Otolaryngology Congress,[53] and the concept was a great success from the start. He, together with Drs. Renato Fior and Giulioand Giulio Pestalozza, organized the landmark First World Congress of Pediatric Otolaryngology in 1977 in Sirmione, Italy. There were round tables, instructional courses, and 150 free papers from 29 countries in 5 continents, with an attendance of more than

FIGURE 1-8. Jan Danielewicz, 1903–1982.

400 delegates. This Congress was a major catalyst for the initiation of focused pediatric ORL in many countries and its advancement worldwide by bringing together physicians from throughout the world for the first time, including many from Eastern Europe in the period of the "Iron Curtain," who met for the first time as individuals on a social basis and exchanged information. Among the fruitful outcomes of this first Congress were plans for a second Congress, which was subsequently held in Bath, Great Britain, in 1982 under the organizational leadership of Mr. Robert Pracy (Fig. 1-9). The First World Congress also led to the establishment of a European Working Group in Pediatric ORL that later became the European Society of Pediatric Otolaryngology (ESPO) and to the initial planning for the creation of *The International Journal of Pediatric Otorhinolaryngology.*[54]

Renato Fior, head of the Department of Otorhinolaryngology at the Istituto Per L'Infanzia in Trieste, organized the first European Course of Pediatric Otorhinolaryngology in Trieste in 1978.[55] Dr. Manacini, president of the Società Italiana di Otorinolaringologia Pediatrica, opened the meeting with the history of pediatric otorhinolaryngology in Italy. There were 20 lectures covering all the major areas of pediatric ORL. At this meeting, the bylaws for the newly formed European Working Group of Pediatric Otolaryngology (EWGPO) were established. These bylaws, which follow, demonstrate the successful cooperation of physicians coming together from many nations and exemplify the ideals of pediatric ORL:

a. To foster clinical and research work in the field of medicine, functional and plastic surgery, and rehabilitation of diseases of the ear, nose, throat, and bronchoesophagology in infants and children.
b. To coordinate cooperative work on a national and international basis between otolaryngologists, pediatricians, audiologists, and speech pathologists.
c. To promote and maintain cooperation with other societies International Federation of Oto-Rhino-Laryngological Societies (IFOS), agencies, health departments, and organizations having a role in health planning within the countries and generally in Europe with the aim of carrying out the purposes of this working group.
d. To organize an information service to provide the public and the national and international health organizations with relevant data or significant events and research findings in the field of pediatric ORL and on the availability of health services, preventive measures, and means for early detections of disease and of rehabilitation.
e. To maintain this working group as a nonprofit organization whose activity shall be strictly scientific and charitable.

FIGURE 1-9. Second World Congress of Pediatric Otolaryngology held in Bath, 1982.

Subsequent EWGPO conferences were held in Warsaw (Poland), Bath (United Kingdom), Sèvres (France), Eger (Hungary), Nijmegen (The Netherlands), and Ghent (Belgium). After the second EWGPO Congress in Sirmione in 1992 and a pediatric ORL conference in Jerusalem (1993), the VIth International Congress of Pediatric Otorhinolaryngology was held in 1994 in Rotterdam under the leadership of Profs. Carl Verwoerd and Jetty Verwoerd-Verhoef, where the Board of the EWGPO founded the ESPO; this organization gained official legal status in 1997 with a set of bylaws, signed by Renato Fior, Paul van Cauwenberge, Pekka Karma, Cor Cremers, Carel Verwoerd, and Jetty Verwoerd-Verhoef.

The International Congresses have continued to be organized every four years (1998 Helsinki, Finland; 2002 Oxford, United Kingdom; and 2006 Paris, France), with the ESPO Conferences being held at the intervening two-year periods (1996 Siena, Italy; 2001 Graz, Austria; 2004 Athens, Greece; and 2008 Budapest, Hungary).

Several European countries have established national societies for pediatric ORL. These include the Association Française d'Otorhinolaryngologie Pédiatrique, the British Association for Paediatric Otorhinolaryngology, the Dutch/Flemish Working Group for Pediatric Otorhinolaryngology, the Hungarian Society of Otorhinolaryngologists Section on Pediatric Otorhinolaryngology, the Italian Society of Pediatric Otorhinolaryngology, and the Polish Society of Pediatric Otolaryngology.

ASIA AND AUSTRALASIA

The Japan Society for Pediatric Otorhinolaryngology was founded in 1979. In composition, it is similar to SENTAC in that the society members consist of both otolaryngologists (approximately 80%) and pediatricians (approximately 20%).[56] The society was initiated by Prof. Junichi Suzuki of Teikyo University and Dr. Keijiro Koga of The National Children's Hospital, both in Tokyo, for the purpose of developing education, practice, and science in pediatric otorhinolaryngology. Conferences were held twice a year in July and December from 1980 to 2005 with the July conference in Osaka and the December conference in Tokyo; each conference focused on a particular theme of pediatric ORL, and presented papers were published twice a year in the *Pediatric Otorhinolaryngology Japan*. In 2006, the Japan Society for Pediatric Otorhinolaryngology changed the organization from holding two conferences a year to an annual meeting and to publish three issues of the society's journal each year. Prof. Shinsaku Horiuchi of the Tokyo Medical and Dental University was the first president of the society, serving from 1979 until 1990, and Dr. Yoshiharu Niino was the first editor-in-chief of the

Pediatric Otorhinolaryngology Japan, from 1980 until 1992. Currently, the society has approximately 600 members.

Australia and New Zealand have established the Australasian Society of Paediatric Otorhinolaryngology,[57] which was formed with the purpose of enabling pediatric ear, nose, and throat—head and neck surgical specialists to engage in meaningful discussion and clinical information sharing. The society promotes research in pediatric ORL and has an annual discussion forum. Membership is for ear, nose, and throat—head and neck surgical specialists who devote a substantial portion of their clinical work to pediatric care. Membership in the society, by application, is open to Australian and New Zealand surgeons.

SOUTH AMERICA[58]

Alexandre Médicis da Silveira in 1960 established a pediatric ORL service at the Hospital Infantil Meñino Jesus in São Paulo, Brazil. Several clinically focused conferences were held subsequently, and by 1977, these became more frequent, aided by the University of São Paulo Medical School together with that at the University of Rio Grande do Sul. Dr. Tania Sih of University of São Paulo Medical School published the first book on pediatric ORL written in Portuguese in 1998,[59] followed by an edition in Spanish[60] in 1999. Dr. Alberto Chinski founded the Asociación Argentina de Otorrinolaringologia y Fonoaudiologia Pediátrica, which has organized several meetings in Argentina.

In 1994, Drs. Sih from Sao Palo, Brazil, Chinski from Buenos Aires, Argentina, and Roland Eavey from Boston, United States, initiated the formation of a pan-Latin American organization devoted to pediatric ORL to promote diffusion of knowledge in this area. A year later, the Interamerican Association of Pediatric Otorhinolaryngology (IAPO) was established officially in Argentina. One hundred and fifty practitioners in ORL joined the organization at that time, and Dr. Sylvan Stool became its first president. Since 1996, IAPO has promoted several Congresses held in Brazil, Ecuador, United States, Chile, Colombia, Argentina, and Panama and an international symposium every other year in São Paulo, Brazil. Thus, the organization has indeed succeeded in bringing together practitioners from much of Latin America. While focused on Latin America, its geographical inclusiveness has brought in physicians from well beyond that region, and it has enjoyed great growth in membership. Today, IAPO has over 6000 members from 85 countries across 5 continents.

CONCLUSION

Pediatric otolaryngic care developed from 1600 to 2000 as a result of medical advances related to evolving conceptions of the philosophical, sociological, and economic status of the child. In the earlier periods, while from our current perspective, there was need for pediatric otolaryngic care, the demand, the economic resources, and the knowledge to effect it were not existent. Today the story is quite different: the needs are better understood, knowledge has advanced greatly, and almost all children in the developed nations can receive effective pediatric otolaryngic care. A substantial need remains for the effective care of children in the developing nations of the world.[61] This need must be fulfilled with the deployment of appropriately educated health care providers and the fiscal resources to enable timely, effective, and efficacious prevention and care of otolaryngologic pathologies in children. This is more essential than ever because in the 21st century, the world's economy is based fundamentally on communication, the diseases and disorders of which are the province of pediatric otorhinolaryngology.[62]

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2 CHAPTER

Phylogenetic Aspects and Embryology

Anne Chun-Hui Tsai and Carol Walton

Knowledge of the embryology, growth, and development of the face and craniofacial complex, and of the various factors involved in normal variations and anomalies of this region, facilitates understanding of the many otorhinolaryngologic disorders affecting infants and children.

The face is the first region that the clinician and, indeed, the layperson inspect on encountering another person. An evaluation of facial type and facial expression are usually made instantly; thereafter, the general body type and posture are noted. This immediate composite impression provides important nonverbal clues to feelings, affect, and communication.

Any observer can appreciate that there is great variation in the appearance of the normal face. In addition, there are certain characteristics that we associate with facial types almost on an instinctive basis. These variations and expectations in facial types can be appreciated by examining Fig. 2-1, which is a sketch of a group of children of different ethnic backgrounds. The variations in facial configuration are obvious: there are round, oval, long, and triangular faces. Individual characteristics of the eyes and the nose also show tremendous variation. The diagnosis of certain conditions based on facial configuration may be difficult to make unless the observer knows the hereditary background of the individual. For example, although the craniofacial features of Down syndrome are readily recognizable, an Asian newborn with a flat nasal bridge and bilateral single palmar creases could be mistaken as having Down syndrome if one is not keeping the ethnic background in mind. Increased intercanthal distance and epicanthic folds are relatively common in the general population among some Asian population.[2] Thus, although we recognize great variations in facial type as being normal, we also instinctively recognize other features as being abnormal in a particular individual on the basis of our ability to assess facial patterns in the context of age, race, and ancestry.

It is notable that the structures of the human craniofacial complex, which required 500 million years of natural selection to evolve, take shape embryologically in incredibly rapid sequence. The embryogenesis of the craniofacial complex is indeed an amazing phenomenon; form and function must relate to each other with an almost unbelievable precision and at exactly the right points in time. Any interference with this process, particularly in the early embryonic stages, may have catastrophic consequences. An abbreviated review of the normal embryogenesis of the human craniofacial complex follows in order for the pediatric otolaryngologist to appreciate why the anatomical and physiological development of the ears, nose, and throat structures occasionally goes awry.

Cellular and molecular advances that have contributed to a better understanding of the embryology of facial configuration and the occurrence of craniofacial anomalies of interest to the physician are also discussed. This method of presentation parallels the way in which the clinician usually views patients with anomalies of this region.

PRENATAL DEVELOPMENT OF THE FACE

The development of the face from midembryologic through midfetal life is illustrated in Fig. 2-2. At approximately 3–4 weeks of age (Fig. 2-2A), the embryo does not have a face, the head is composed of a brain covered with a membrane, and the anterior neuropore is still present. The eyes, which are represented by optic vesicles, are on the lateral aspects of the head, as in fish, and the future mouth is represented by a stomodeum. The nasal pits develop only in the latter part of this period of embryonic growth. At the embryonic age of 5–6 weeks (Fig. 2-2B), the general shape of the face has begun to develop. The frontonasal process is prominent; the nasal pits are forming laterally; and with the increase in size of the first and second branchial arches, there is the suggestion of a mouth. In the subsequent weeks of embryonic life (Fig. 2-2C), the structures that we associate with the human face—jaws, nose, eyes, ears, and mouth—take on human form.[3]

During this period of rapid growth and expansion, there is also tremendous *differential* growth. Thus, the development of a human baby is not merely the enlargement or rearrangement of a previous form but, by differential growth, the development of a new configuration. This is a concept that has been difficult for students to comprehend, perhaps because of the tendency for different stages of embryonic development to be illustrated with drawings of equal size. These illustrating techniques have been used because minute structures are difficult to demonstrate without magnification. It is important, however, to view human embryologic development in both spatial and temporal perspectives to appreciate both its similarities to phylogenetic development and its unique course in humans.

The embryonic period ends at about eight weeks, at which time the embryo has achieved sufficient size and form so that facial characteristics can be recognized and photographed at actual size (Fig. 2-2D). At this stage of late embryonic or early fetal development, the facial features are characterized by the appearance of hypertelorism. During subsequent growth, it will appear as though the eyes are moving closer together. This is, however, not the case; the eyes actually continue to move farther apart, but the remainder of the face is growing at a much more rapid rate, and thus it appears that the eyes are moving closer together. These observations may

FIGURE 2-1. Children from a sixth-grade class. Note the variation of facial types, even though all the children are the same age and race.

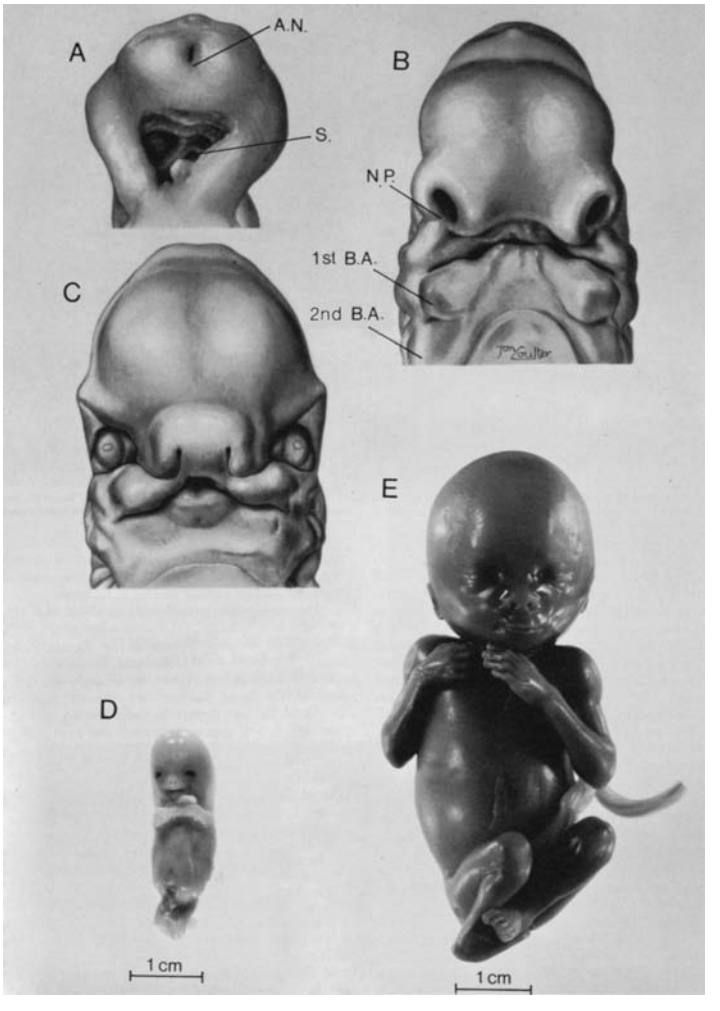

FIGURE 2-2. Prenatal facial development. *A*, An embryo of 3–4 weeks. A.N., anterior neuropore; S., stomodeum. *B*, An embryo of 5–6 weeks. N.P., nasal pit; 1st B.A., first branchial arch; 2nd B.A., second branchial arch. *C*, An embryo of 7–8 weeks. *D*, A fetus of 8–9 weeks. *E*, A fetus of 3–4 months. Fetal specimens are from the Krause Collection, the Cleft Palate Center, University of Pittsburgh.

be of importance in understanding some of the craniofacial syndromes in which hypertelorism is a prominent feature.

The rapid growth and change in configuration, not only of the face, but also of the extremities and body, continue during the next few months (Fig. 2-2E). The fetus now has facial features that are easily recognized and construed as human. The ears, nares, and lips are well developed, and the head constitutes a large portion of the body mass, a relationship that exists at birth and gradually changes during postnatal life.

This concept of differential growth is vital to the comprehension of both prenatal and postnatal development. Although this concept is difficult to grasp when the student must view development of structures of different ages magnified to the same size and when illustrations are in two dimensions, it is important to visualize the process in three dimensions *as well as* in the fourth dimension of time.

FORMATION OF THE CRANIOFACIAL COMPLEX

The factors that influence the formation of the craniofacial complex have been the subject of investigation by embryologists for many years. Among the most interesting studies has been the research of Johnston[4] into the development and migration of cells in the neural crest. These cells of ectodermal origin are found around the anterior neuropore, as demonstrated in Fig. 2-3A, whereas in most of the body, the embryonic tissue is derived from mesoderm. In the craniofacial complex, it is these neural crest cells that give rise to a large variety of the connective and neural tissue structures of the skull, face, and branchial arches. Therefore such ectodermal tissue constitutes the majority of the pluripotent tissue of the face. The sequence of events after the initial formation of the neural crest cells is illustrated in Fig. 2-3B. The differentiation, proliferation, and migration of these cells are critical in the formation of the face.

Neural crest cell migration occurs at different rates. For instance, the cells that form the frontonasal process are derived from the forebrain fold, and their migration is over a relatively short distance as they pass into the nasal region. However, the cells that form the mesenchyme of the maxillary processes have a considerably longer distance to migrate, because they must move into the branchial arches, where they surround the core-like mesodermal muscle plates.[5] In Fig. 2-3C, the ultimate distribution of neural crest cells from the frontonasal process and from the branchial arches is illustrated. Because this mesenchymal tissue contributes to the majority of the soft tissues and bones of the face, failure of neural crest cell proliferation or migration may be responsible for a number of craniofacial abnormalities such as orofacial clefting.[6] An example of a severe facial abnormality due to failure of neural crest cell migration is illustrated in Fig. 2-3D.

Several recognizable craniofacial phenotypes and syndromes result from errors during such embryologic formation of the key structures of the craniofacial complex. Many of these syndromes can now be explained based on recent molecular advances that have contributed to the understanding of the genes involved (Table 2-1).

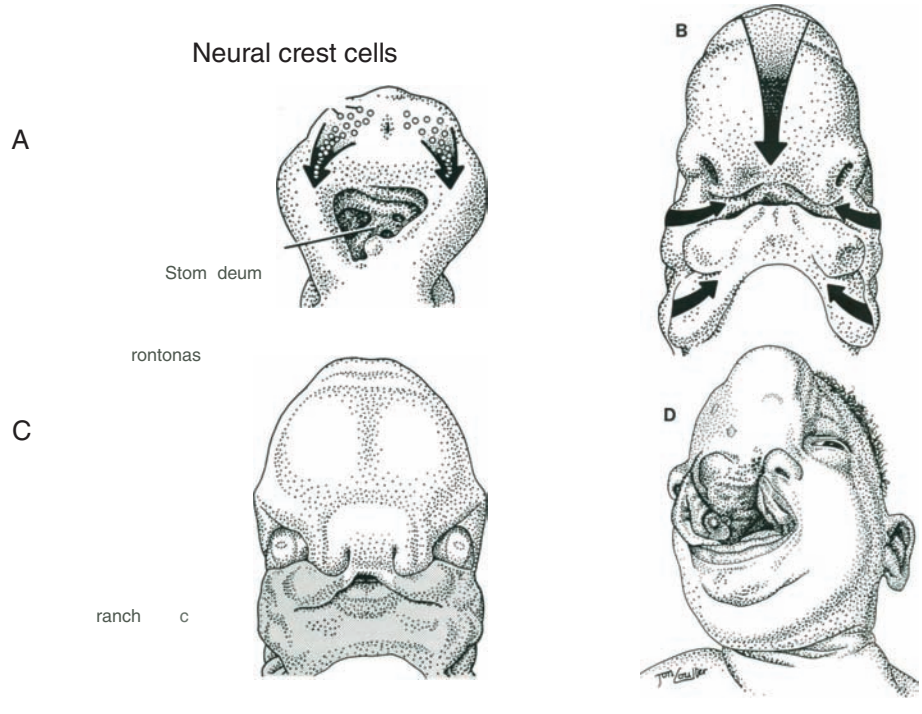

FIGURE 2-3. Formation of the craniofacial complex. *A*, An embryo of 3–4 weeks showing development and beginning migration of neural crest cells. *B*, Migration of neural crest cells to the forebrain and the branchial arches. *C*, Contributions to the face of the frontonasal process and branchial arches. *D*, Deformity caused by failure of neural crest cell migration.