

Contents

1	A Paediatric Overview of Children Seen in the ENT Outpatient Department	1
	<i>Helen Bantock</i>	
	Introduction: Paediatric Consultation	1
	Development in the First Five Years of Life	2
	Children with Special Needs	2
	Speech and Language Delay or Disorder	2
	Autistic Spectrum	2
	Global Delay	3
	Specific Learning Difficulties	3
	Syndromes	3
	<i>Down Syndrome</i>	3
	Behaviour Difficulties	3
	<i>Attention Deficit Hyperactivity Disorder</i>	3
	Social Difficulties	3
	<i>Types of Abuse</i>	3
	<i>Child Abuse in the Context of an ENT Clinic</i>	4
	<i>Alerting Signs of Non-Accidental Injury and Abuse in a Clinic Setting</i> . . .	4
	What to do if Abuse is Suspected	4
	Links with Local Services	4
	Primary Care Team	4
	The Child Development Team	4
	The Education Service	4
	Child and Adolescent Mental Health Services	5
	Social Services	5
	Non-Statutory Organisations	5
2	Nursing Aspects of Paediatric ENT	7
	<i>Rosalind Wilson and Judith Barton</i>	
	Introduction	7
	Communication	7
	Consent for Surgery	8
	Staffing	8
	The Physical Environment	8
	Tracheostomy	8
	Parents	9
	Day Surgery	9
	Pain Control	10

3	Anaesthesia for Paediatric ENT Surgery	11
	<i>Ian Barker</i>	
	Introduction	11
	General Considerations	12
	Anaesthesia for Children	12
	Anaesthesia for the Newborn	12
	Hypoxia	12
	Hypothermia	12
	Hypoglycaemia	12
	Intravenous Fluid Therapy for Children	12
	Specific Conditions	13
	Post-Tonsillectomy Bleeding	13
	Assessment	13
	Resuscitation	13
	Anaesthesia	13
	Airway Endoscopy	13
	Indications	13
	Induction	14
	Maintenance	14
	Laser Surgery	14
	Obstructive Sleep Apnoea	15
	Anaesthetic Equipment	15
	Endotracheal Tubes	15
	Laryngeal Mask Airway	16
	Pulse Oximeter	16
	Limitations	16
	Carbon Dioxide Monitor	16
4	The Evolution of Speech and Language	19
	<i>John F. Stein</i>	
	Introduction	19
	Is Language Innate or Learnt?	20
	Human Evolution	20
	Left-Sided Specialisation	20
	Emotional Vocalisations	21
	Mirror Neurones	21
	Bipedalism and the Aquatic Ape Hypothesis	22
	Larynx Descent	22

	Brain Enlargement	23
	Right-Handedness	23
	Selective Advantages of Speech	23
	Gesture and Speech	24
	Writing	24
	Conclusion	24
5	Evidence-Based Management of Speech and Language Delays	27
	<i>Amy McConkey Robbins</i>	
	Introduction	27
	Milestones for Communication Development	28
	Normal Variation in Language Learning	28
	Normal Variation versus Communication Delay	28
	Communication Milestones in the First Year of Life	29
	Clinical Use of Table 5.1 by Physicians	31
	Speech and Language Intervention is Correlated with Young Age at Identification	32
	Hearing Loss can be Identified and Treated as Early as the Newborn Period	32
	Differential Diagnosis of Several Communication Disorders	33
	Association Between Some Speech/ Language Characteristics and Reading Disability	34
	Evidence-Based Approach to the Management of Children's Speech and Language Development	34
6	Paediatric Voice	37
	<i>Mark E. Boseley and Christopher J. Hartnick</i>	
	Introduction	37
	Anatomy	38
	Gross	38
	Histology	38
	Diagnostic Techniques	39
	Physical Examination	39
	Voice Outcome Surveys	39
	Common Voice Pathologies	39
	Vocal Fold Nodules	39
	Voice Results Following Laryngotracheal Reconstruction	42
	Recurrent Respiratory Papillomatosis	43
	Vocal Fold Paralysis	43

7	Genetics of Non-Syndromic Deafness	47
	<i>Maria Bitner-Glindzicz</i>	
	Introduction	47
	Investigation of the Aetiology of Hearing Loss	48
	Reasons	48
	Protocol	48
	History	48
	Examination	48
	Investigation	48
	Genetic Hearing Loss	49
	Recessive Genes: Common/ Recognizable Types of Non-Syndromic Genetic Hearing Loss	50
	<i>GJB2 (Connexin26)</i>	50
	<i>SLC26A4 (Pendred and Non- Syndromic Enlarged Vestibular Aqueduct)</i>	52
	<i>OTOF (Auditory Neuropathy)</i>	53
	Dominant Genes: Common/ Recognizable Types of Non- Syndromic Genetic Hearing Loss	53
	<i>WFS1 (Dominant Low-Frequency Sensorineural Hearing Loss)</i>	53
	<i>COCH (Menière-Like Symptoms)</i> ...	54
8	ENT-Related Syndromes	57
	<i>David Albert and Fiona Connell</i>	
	Definition	58
	Introduction	58
	Resources	59
	Syndromes of Particular Relevance to the ENT Clinician	59
	Down Syndrome	59
	<i>Genetics</i>	59
	<i>ENT Features</i>	59
	<i>General Features</i>	59
	Pierre Robin Sequence	59
	<i>Genetics</i>	59
	<i>ENT Features</i>	60
	Treacher Collins Syndrome	60
	<i>Genetics</i>	60
	<i>ENT Features</i>	60
	<i>General Features</i>	60
	Goldenhar Syndrome	60
	CHARGE Syndrome	60
	<i>Genetics</i>	60
	<i>ENT Features</i>	60
	<i>General Features</i>	61
	<i>Genetics</i>	61
	<i>ENT Features</i>	61
	<i>General Features</i>	61

Branchio-oto-renal Syndrome	62
<i>Genetics</i>	62
<i>ENT Features</i>	62
<i>General Features</i>	62
22q11.2 Deletion Syndrome	62
<i>Genetics</i>	62
<i>ENT Features</i>	62
<i>General Features</i>	62
Craniosynostosis Syndromes	63
<i>Genetics</i>	63
<i>ENT Features</i>	63
<i>General Features</i>	63
Alport Syndrome	63
<i>Genetics</i>	63
<i>ENT Features</i>	63
<i>General Features</i>	63
Pendred Syndrome	63
Usher Syndrome	63
<i>Genetics</i>	63
Alstrom Syndrome	65
<i>Genetics</i>	65
<i>ENT Features</i>	65
<i>General Features</i>	65
Syndromes Less Commonly Seen by ENT Surgeons	65
Achondroplasia	65
<i>Genetics</i>	65
<i>ENT Features</i>	65
<i>General Features</i>	65
Beckwith-Wiedemann Syndrome	65
<i>Genetics</i>	65
<i>ENT Features</i>	65
<i>General Features</i>	65
Neurofibromatosis Type 2	66
<i>Genetics</i>	66
<i>ENT Features</i>	66
<i>General Features</i>	66
Noonan Syndrome	67
<i>Genetics</i>	67
<i>ENT Features</i>	67
<i>General Features</i>	67
Osteogenesis Imperfecta	67
<i>Genetics</i>	67
<i>ENT Features (Type 1)</i>	67
<i>General Features (Type 1)</i>	67
Prader-Willi Syndrome	67
<i>Genetics</i>	67
<i>ENT Features</i>	67
<i>General Features</i>	67
Stickler Syndrome	68
<i>Genetics</i>	68
<i>ENT Features</i>	68
<i>General Features</i>	68
Turner Syndrome	68
<i>Genetics</i>	68

<i>ENT Features</i>	68
<i>General Features</i>	68
Waardenburg Syndrome	68
<i>Genetics</i>	68
<i>ENT Features</i>	68
<i>General Features</i>	68
Jervell and Lange-Nielsen Syndrome ..	68
<i>Genetics</i>	68
<i>ENT Features</i>	69
<i>General Features</i>	69
Gorlin Syndrome	69
<i>Genetics</i>	69
<i>ENT Features</i>	69
<i>General Features</i>	69
Holoprosencephaly	69
<i>Genetics</i>	69
<i>ENT Features</i>	69
<i>General Features</i>	69
Mucopolysaccharidoses	69
<i>Genetics</i>	69
<i>ENT Features</i>	69
<i>General Features</i>	69
Foetal Alcohol Syndrome	70
<i>Genetics</i>	70
<i>ENT Features</i>	70
<i>General Features</i>	70
Foetal Cytomegalovirus Syndrome	70
<i>Features</i>	70
Congenital rubella syndrome	70
<i>Features</i>	70
Acknowledgements	70

9 EXIT – Antenatal (Pre-natal)

Diagnoses and Management	73
<i>Gavin Morrison</i>	

Introduction	73
Pre-natal Diagnosis	74
Diagnostic Ultrasound Features	74
Congenital High Airway Obstruction Syndrome	75
Foetal MRI Scanning	75
The EXIT Procedure	76
Indications for the EXIT Procedure ...	76
Techniques of the EXIT Procedure	76
Tips and Pitfalls of EXIT	77
<i>Spontaneous Onset of Labour</i>	77
<i>Cystic Hygroma</i>	78
<i>Maintaining Placento-Foetal Circulation</i>	78
<i>Loss of Placento-Foetal Circulation</i> ..	78
<i>Risks to Mother</i>	78
Outcomes of EXIT	78
Counselling and Ethical Issues	78

Termination of Pregnancy	79
Foetal Surgery	79
Intrauterine Treatment of CHD and EXIT	79
Newer Applications and the Future ...	80
10 The Immunocompromised Child	83
<i>Mich Lajeunesse and Adam Finn</i>	
Scope of Chapter	83
Primary Immunodeficiency	83
Natural History if Untreated	84
Prevalence and Incidence	85
Aetiology and Genetics	85
Assessment and Diagnosis	85
<i>Severe</i>	85
<i>Prolonged</i>	85
<i>Unusual</i>	86
<i>Recurrent</i>	86
<i>Examination</i>	86
Classification	86
<i>Antibody Deficiencies</i>	87
<i>Complement Deficiencies</i>	87
<i>Mannose-Binding Lectin Deficiency</i>	87
<i>Neutrophil Disorders</i>	88
Laboratory Tests	88
Treatment	88
<i>Prophylactic Antibiotics</i>	88
<i>Immunoglobulin Replacement</i> <i>Therapy</i>	88
<i>Immunisation</i>	88
<i>Bone Marrow Transplantation</i>	89
<i>Gene Therapy</i>	89
Secondary Immunodeficiency	89
Human Immunodeficiency Virus	89
Description	90
Natural History if Untreated and Complications	90
Prevalence	90
Classification	90
Clinical Assessment	91
Diagnosis	91
Treatment	91
11 Cystic Fibrosis and Primary Ciliary Dyskinesia	95
<i>Andrew Bush and Jonny Harcourt</i>	
Introduction	96
Cystic Fibrosis – The Disease	96
Molecular Defect	96
Pathophysiology	96
<i>Lung Disease</i>	96

<i>Gastrointestinal and Other Disease</i>	
<i>Manifestations</i>	97
Prevalence	97
Diagnostic Tests	97
<i>Clinical Suspicion</i>	97
<i>The Sweat Test</i>	98
<i>Genetic Testing</i>	98
<i>Ancillary Testing</i>	98
Features of the Disease	98
<i>Upper Airway Disease</i>	98
<i>Lower Airway Disease</i>	99
<i>Other Organ Disease</i>	99
Medical Treatment	99
<i>Upper Airway Disease</i>	99
<i>Lower Airway Disease</i>	99
<i>Other Systems</i>	100
When Should an Apparently Healthy Child be Suspected of Having CF? ...	101
PCD – The Disease	102
Molecular Defect	102
Pathophysiology	102
Prevalence	102
Diagnostic Tests	102
<i>In Vivo Tests of Ciliary Function</i> ...	102
<i>Nitric Oxide Studies</i>	102
<i>Measurement of Ciliary Beat Frequency and Pattern</i>	103
<i>Electron Microscopy</i>	103
<i>Culture of Cilia</i>	104
Features of the Disease	104
<i>Upper Airway Disease</i>	104
<i>Lower Airway Disease</i>	104
<i>Other Organ Disease</i>	104
Medical Treatment	105
<i>Upper Airway Disease</i>	105
<i>Lower Airway Disease</i>	105
<i>Other Organ Disease</i>	105
When Should an Apparently Healthy Child be Suspected of Having PCD?	105
The Child Referred for Surgery for an Unrelated Condition	105
ENT Surgery for CF	106
ENT Surgery for PCD	108

12 Head and Neck Masses 111

Ben Hartley

Introduction	112
Part 1. Clinical Assessment of Neck Swelling in Children	112
History	112
Examination of the Neck	112
<i>Site of the Swelling</i>	112
<i>Nature of the Swelling</i>	113

<i>Head and Neck Examination</i>	113
<i>General Examination</i>	113
Investigation	113
<i>Laboratory Tests</i>	113
<i>Radiology</i>	113
<i>Biopsy Sampling versus Fine-Needle Aspiration for Cytology</i>	114
Part 2. Inflammatory Disorders of the Neck and Lymphadenopathy	114
Viral Infections	114
<i>Viral Upper Respiratory Tract Infections</i>	114
<i>Infectious Mononucleosis</i>	115
<i>Human Immunodeficiency Virus</i> ...	115
Bacterial Infections	115
<i>Acute Lymphadenopathy with Suppuration</i>	115
<i>Mycobacterial Neck Infections</i>	115
Non-Infectious Inflammatory Lymphadenopathy	116
<i>Kawasaki Syndrome</i>	116
<i>Sinus Histiocytosis (Rosai Dorfman Disease)</i>	116
<i>Kikuchi-Fujimoto Disease</i>	116
Part 3. Haemangiomas and Vascular Malformations	116
Terminology	116
<i>Haemangiomas</i>	117
<i>Vascular Malformations</i>	117
<i>Capillary Malformations</i>	117
<i>Venous Malformations</i>	118
<i>Arteriovenous Malformations</i>	119
Part 4. Head and Neck Malignancy in Children	119
Hodgkin's Disease	119
Non-Hodgkin Lymphoma	119
Rhabdomyosarcoma	120
Non-Rhabdomyosarcoma Sarcomas	120
Thyroid Carcinoma in Children	120
Salivary Gland Malignancy	120
Neuroblastoma	120
Nasopharyngeal Carcinoma	120
Teratoma	121
Juvenile Nasopharyngeal Angiofibroma	121

13 Paediatric ENT

in Developing Countries	123
<i>Neville P. Shine and Christopher A. J. Prescott</i>	

Introduction	123
Ear Disease	124
Chronic Suppurative Otitis Media ...	125

Tympanomastoid Surgery	125
Hearing	126
Adenotonsillar Surgery	127
HIV/AIDS and Tuberculosis	127
Tracheotomy	128
ENT Aspects of Accidents and Trauma	128
Paediatric Head and Neck Tumours	128

14 Tonsils and Adenoids 131

Anne Pitkäranta and Pekka Karma

Waldeyer's Ring	131
Tonsillitis	132
Acute Tonsillitis	132
Viral Tonsillitis	132
Bacterial Tonsillitis	133
Chronic Tonsillitis	134
Complications of Tonsillitis	134
<i>Peritonsillar Abscess (Quinsy)</i>	134
<i>Adenotonsillar Hyperplasia</i>	134
<i>Mouth Breathing</i>	134
Tonsillectomy	135
Indications	135
Techniques	135
Complications	136
Evaluation of the Adenoids	137
Adenoidectomy	137
Indications	137
Techniques	138
Complications	138
Neoplasms of the Tonsils and the Adenoids	138

15 The Causes and Effects of Obstructive Sleep Apnoea in Children 141

Ray W. Clarke

Pathophysiology of Childhood Obstructive Sleep Apnoea	142
Introduction	142
Prevalence	142
Aetiology	142
Mechanisms	143
Diagnosis of OSA	143
The Spectrum of Airway Obstruction	143
Diagnostic Criteria	144
Presentation	144
<i>Clinical Features</i>	144
Differential Diagnosis	144
Effects of OSA	144
Management	145
Investigations	145
Sleep Studies (Polysomnography)	145

<i>Pre-Operative Investigations</i>	146
<i>Endoscopy (Sleep Nasendoscopy)</i> ...	146
<i>Paediatric Evaluation</i>	147
Treatment	147
<i>General Principles</i>	147
<i>Pharmacological Treatment</i>	147
<i>Dental Devices</i>	147
<i>Continuous Positive Airway Pressure</i>	147
<i>Postural Therapy</i>	147
<i>Nasopharyngeal Airway</i>	147
<i>Adenotonsillectomy</i>	147
<i>Nasal Surgery</i>	148
<i>Maxillomandibular Surgery</i>	148
<i>Palatal Surgery</i>	148
<i>Tongue-Base Surgery</i>	149
<i>Tracheotomy</i>	149
Challenges in OSA Management	149
OSA and Obesity	149
OSA in Special Needs Children	149
16 Cleft Lip and Palate	153
<i>John Boorman</i>	
Embryology	153
Aetiology	153
Prevalence	153
Presentation	154
Robin Sequence	154
Feeding	155
Surgery for Cleft Lip and Palate	155
Velopharyngeal Insufficiency	156
Sub-mucous Cleft Palate and Bifid	
Uvula	156
Assessment of VPI	157
Treatment of VPI	157
Velocardiofacial Syndrome	157
17 Salivary Gland Disease in Childhood	159
<i>Peter D. Bull</i>	
Introduction	159
History, Investigations	159
History	159
Examination	160
Investigation	160
Congenital Disorders	160
Congenital Cysts and Fistulae	160
Inflammatory Disorders	
of the Salivary Glands	160
Viral Infections	160
Bacterial Disease	160
Atypical Mycobacterial Disease	
of Salivary the Glands	160

<i>Features</i>	160
<i>Investigations</i>	161
<i>Treatment</i>	161
Recurrent Parotitis of Childhood	161
Sjogren's Syndrome	161
Human Immunodeficiency Virus/ Acquired Immune Deficiency Syndrome	162
Sarcoidosis	162
Obstructive Disorders	162
Salivary Gland Calculi	162
<i>Presentation</i>	162
<i>Treatment</i>	162
<i>Ranula</i>	162
<i>Treatment</i>	162
Salivary Gland Tumours in Childhood	162
Benign	162
Malignant	163
<i>Presentation</i>	163
<i>Pathology</i>	163
<i>Treatment</i>	163
<i>Complications of Parotid Surgery</i> ...	163

18 Drooling – Salivary Incontinence

(Sialorrhoea)	165
<i>Peter D. Bull</i>	

Introduction	165
Innervation	165
Assessment	165
Non-Surgical Management	166
<i>Posture Control</i>	166
<i>Drug Treatment</i>	166
Surgical Management	167

19 Congenital Cysts, Sinuses

and Fistulae	169
<i>Fiona B. MacGregor</i>	

Introduction	170
Developmental Anatomy	170
Classification of Congenital Branchial Abnormalities in the Neck	170
Pre-Auricular Sinuses	170
<i>Presentation</i>	171
<i>Investigations</i>	171
<i>Management</i>	171
<i>Complications</i>	171
First Branchial Arch Anomalies	171
<i>Presentation</i>	171
<i>Investigations</i>	172
<i>Treatment</i>	172
<i>Complications</i>	172

Second Branchial Arch Anomalies ...	172
<i>Presentation</i>	172
<i>Investigations</i>	172
<i>Treatment</i>	172
Third and Fourth Arch Anomalies ...	172
<i>Presentation</i>	172
<i>Investigations</i>	173
<i>Management</i>	173
<i>Complications</i>	173
Branchio-Oto-Renal Syndrome	173
Thyroglossal Duct Cyst	173
<i>Presentation</i>	173
<i>Investigations</i>	174
<i>Treatment</i>	174
<i>Complications</i>	174
Lingual Thyroid	174
<i>Presentation</i>	174
<i>Investigations</i>	174
<i>Management</i>	174
<i>Complications</i>	174
Dermoid Cyst	174
<i>Presentation</i>	175
<i>Investigations</i>	175
<i>Treatment</i>	175
Nasal Dermoid Cysts	175
<i>Presentation</i>	175
<i>Investigations</i>	175
<i>Management</i>	175
 20 Airway Endoscopy and Assessment in Children	 177
<i>Sonia Ayari-Khalfallah and Patrick Froehlich</i>	
Introduction	177
Rigid Endoscopy	178
Supplementary Examinations	181
 21 Emergency Management of the Paediatric Airway	 183
<i>Michael Kuo and Michael Rothera</i>	
Anatomical and Physiological Considerations	183
Clinical and Radiological Assessment of the Acutely Compromised Upper Airway	183
Resuscitation and Pre-Operating-Room Management	186
Endoscopy Under General Anaesthesia	186
Surgical Management of the Acutely Obstructed Paediatric Airway	187

22	Congenital Disorders of the Larynx, Trachea and Bronchi	189
	<i>Martin Bailey</i>	
	Introduction	189
	Larynx	189
	Supraglottis	190
	Laryngomalacia	190
	Saccular Cysts and Laryngocoeles	190
	Lymphangioma	191
	Bifid Epiglottis	191
	Glottis	191
	Laryngeal Webs	191
	Laryngeal Atresia	191
	Cri-du-Chat Syndrome	191
	Vocal Cord Paralysis	191
	Subglottis	192
	Congenital Subglottic Stenosis	192
	Subglottic Haemangioma	193
	Laryngeal and Laryngotracheo- Oesophageal Cleft	193
	Trachea and Bronchi	194
	Agenesis	194
	Stenosis	194
	Tracheomalacia and Bronchomalacia	194
	Tracheo-Oesophageal Fistula	194
	Vascular Compression	194
	<i>Vascular Ring</i>	195
	<i>Vascular Sling</i>	195
	Anomalous Bifurcations	195
	Congenital Cysts and Tumours	195
23	Acquired Disorders of the Larynx in Children	197
	<i>Neville P. Shine and Christopher Prescott</i>	
	Infections	197
	Laryngitis	197
	Laryngotracheobronchitis (Croup)	198
	Bacterial Laryngitis/Tracheitis	199
	Supraglottitis (Epiglottitis)	200
	Unusual Laryngeal Infections	200
	<i>Herpetic Laryngitis</i>	200
	<i>Candidiasis</i>	201
	<i>Tuberculosis</i>	201
	<i>Diphtheria</i>	202
	Gastro-Oesophageal Reflux Disease – Laryngeal Manifestations	202
	Laryngeal Trauma	203
	External Trauma	203
	Internal Trauma	204
	<i>Inhalation Burns</i>	204
	<i>Corrosive Ingestion</i>	205
	<i>Intubation Injury</i>	205

	<i>Vocal Abuse</i>	207
	Foreign Bodies	207
	Neurological Conditions	208
	Recurrent Laryngeal Nerve Palsy	208
24	Laryngeal Webs and Subglottic Hemangiomas	211
	<i>Michael J. Rutter</i>	
	Introduction	211
	Congenital Anterior Glottic Webs	211
	Diagnostic Guidelines	212
	Management Strategies	213
	Acquired Anterior Glottic Webs	214
	Management Strategies	215
	Hemangiomas of Infancy	216
	Natural History	216
	Subglottic Hemangiomas	217
	Diagnostic Guidelines	218
	Management Strategies	220
	Observation	220
	Pharmacotherapy	220
	Endoscopic Surgical Management ...	220
	Tracheotomy	221
	Open Surgical Excision	221
25	Post-Intubation Laryngotracheal Stenosis	223
	<i>David Albert</i>	
	Introduction	224
	Definition	224
	Pathology and Aetiology	224
	Incidence	224
	Prevention	224
	Assessment of LTS	224
	History	224
	Examination	224
	Pre-Endoscopy Investigations	224
	Endoscopy	224
	Grading	225
	Scenarios	225
	Failed Extubation	225
	Existing Tracheostomy	226
	Recurrent Croup/Progressive Stridor	226
	Management of Early Soft Stenosis	226
	Laryngeal Rest	226
	Removal of Granulations	226
	Avoidance of the Laser	226
	Endoscopic Procedures	226
	Radial Dilatation	226
	Endoscopic Decompression/Cricoid Split	226
	Mitomycin	227

Conventional External Cricoid Split	227
Management of Firm Established	
Stenosis	227
Expectant Waiting/Natural Resolution	227
Augmentation Grafting	227
Resection	227
Prevention of Post-Operative	
Restenosis – Salvage Techniques	227

26 Congenital and Acquired Tracheal Stenosis in Children 229

*Emmanuel Le Bret
and E. Noel Garabédian*

Congenital Stenosis	230
Assessment of the Lesion	230
<i>Presenting Symptoms and Management</i>	230
<i>Investigation, Imaging</i>	230
Description of the Lesions	230
<i>The Narrowness of the Trachea</i>	231
<i>The Extent of Tracheal Involvement</i>	231
<i>The Anatomy of the Bronchi</i>	232
<i>Associated Lesions</i>	232
Surgical Treatment	232
<i>Gas Exchange During Reconstruction</i>	232
<i>Very-Short-Segment Tracheal Stenosis</i>	233
<i>Medium-Length Stenoses (up to Two-Thirds of Tracheal Length)</i>	233
<i>Long and Very Long Tracheal Stenoses</i>	234
<i>Post-Operative Complications and Their Management</i>	234
Acquired Stenosis	235
Aetiologies and Localisation of Acquired Tracheal Stenosis in Children	235
Patient Assessment	235
Treatment	236
<i>Endoscopic Procedures</i>	236
<i>Open Surgical Procedure</i>	236

27 Tracheomalacia in Children 241

*Jean-Michel Triglia, Richard Nicollas
and Stephane Roman*

Introduction	241
Clinical Manifestations	242
Investigations	242
Aetiology of Tracheomalacia	243
Primary Tracheomalacia	243

Secondary Tracheomalacia	243
Oesophageal Atresia and TOF	243
Tracheal Compression due to Cardiovascular Malformations	243
Tracheal Compression due to Cervical and Mediastinal Tumours ..	244
28 Tracheostomy: an Ancient Life Saver Due for Retirement, or Vital Aid in Modern Airway Surgery?	247
<i>Hans L. J. Hoeve</i>	
Introduction	247
Definitions	247
Indications	247
Alternatives to Tracheotomy	248
The Technique of Tracheotomy	248
Complications	249
Complications of a Tracheotomy	249
Complications of a Tracheostomy ...	250
Types and Sizes of Tracheostomy Tube	250
Tracheostomy Care in the Hospital, an Institution or at Home	251
Decannulation	252
29 Recurrent Respiratory Papillomatosis	255
<i>Brian J. Wiatrak</i>	
Introduction	255
Historical Perspective	256
Pathogenesis of RRP in Children	256
Epidemiology	258
Diagnosis and Staging of RRP in Children	258
Disease Progression and Complications	259
Managing RRP in Children	260
Principles of Surgical Management	260
Anesthesia/Airway Management	
Techniques	261
<i>Spontaneous Ventilation Technique</i>	261
<i>Intermittent Apnea Technique</i>	261
<i>Jet Ventilation Technique</i>	261
Endoscopy Techniques	261
Laser Techniques	261
Microdebridement	261
Adjuvant Medical Therapy	262
Indol-3 Carbinol	262
Alpha Interferon	262
Cidofovir	262
Photodynamic Therapy	263
Other Agents	263
Investigational Agents	263
Voice Outcome	263

30	Pharynx and Oesophagus	267
	<i>David Rawat and Mike Thomson</i>	
	Introduction	267
	Ontogeny of Oesophageal Motor Function	269
	Swallowing in the Normal Infant and Child	269
	The Pharyngeal Phase	269
	The Oesophageal Phase	270
	<i>Oesophageal Sphincter Physiology</i> ..	271
	Sphincter Dysfunction	271
	Mucosal Immunology and Inflammation	271
	Gastro-Oesophageal Reflux	271
	Pathophysiology of GOR	272
	GOR-Related Apparent Life- Threatening Events and Sudden Infant Death Syndrome	273
	GOR and Asthma	273
	Recurrent Pneumonia	273
	Upper Airway Symptoms	273
	Chronic Tubotympanic Disorders ...	273
	Diagnostic Options	274
	Intraluminal Impedance	274
	Endoscopy and Biopsy Sampling	274
	Oesophageal Dysmotility, Extrinsic Compression and Mechanical Obstruction	274
31	Paediatric Thyroid Disease	277
	<i>Michael Kuo and Tim Barrett</i>	
	Introduction	277
	Embryology, Physiology and Anatomy	277
	Embryology	277
	Physiology	278
	Anatomy	278
	Investigation of the Thyroid Gland	279
	Thyroid Function Tests	279
	Thyroid Immunology	279
	Genetic Testing	279
	Ultrasonography/Computed Tomography/Magnetic Resonance Imaging	280
	Radionucleotide Scan	280
	Fine-Needle Aspiration	280
	Congenital Abnormalities	280
	Thyroglossal Duct Cysts and Fistulae, and Ectopic Thyroid	280
	Congenital Hypothyroidism	280
	Syndromes Associated with Thyroid Abnormalities	281
	Functional Abnormalities of the Thyroid Gland	281

Hypothyroidism	281
Hyperthyroidism	281
Neoplasms of the Thyroid Gland	282
Approach to the Solitary Thyroid	
Nodule in Children	282
Sporadic Differentiated Thyroid	
Cancer	282
MTC and the Management	
of the Thyroid in MEN-2	283
 32 Nasal Foreign Bodies, Epistaxis	
and Nasal Trauma	285
<i>Sean Carrie</i>	
Introduction	285
Foreign Bodies	285
Types	285
Presentation	285
Signs and Symptoms	286
Management	286
Rhinolith	286
Epistaxis	286
Prevalence	286
Blood Supply to the Nose	286
Aetiology	287
Trauma	287
Assessment and Management	287
Nasal Trauma	288
Assessment	288
Management	288
 33 Management of Choanal Atresia	291
<i>Patrick Froehlich</i>	
<i>and Sonia Ayari-Khalfallah</i>	
Introduction	291
Anatomical and Physiological	
Background	291
Clinical Presentation	292
Management is by Surgery	292
Intranasal Approach	292
Transpalatine Approach	293
Indications and Timing of Surgery	293
Contraindications	293
Risks and Side Effects	293
 34 Allergic Rhinitis	295
<i>Alexander N. Greiner</i>	
Introduction	295
Epidemiology	295
Etiology of AR	297
AR and its Associated Conditions	
and Comorbidities	297

Quality of Life	298
Classification	298
Treatment Modalities	300
Environmental Control Measures	300
Overview of Pharmacological	
Treatment	300
<i>H1-Antihistamines</i>	300
<i>Intranasal Corticosteroids</i>	302
<i>Decongestants</i>	302
<i>Cromolyn</i>	303
<i>Antileukotrienes</i>	303
<i>Omalizumab (Anti-IgE)</i>	304
Allergen Immunotherapy	304
Other Treatment Modalities	304

35 Rhinosinusitis in Children 307

Peter A. R. Clement

Introduction	307
Morphogenesis, Epidemiology	
and Pathophysiology	308
Classification and Definitions	310
Acute Rhinosinusitis	310
Chronic Rhinosinusitis	311
Diagnosis	312
Presenting Symptoms	
of Rhinosinusitis in Children	312
Clinical Examination	312
Microbiology	312
Imaging	312
Additional Investigations	312
<i>Respiratory Allergy</i>	313
<i>Immune Deficiency</i>	313
<i>Cystic Fibrosis</i>	313
<i>Primary Ciliary Dyskinesia</i>	313
<i>Gastro-Oesophageal Reflux</i>	313
<i>Mechanical Obstruction</i>	313
<i>Miscellaneous</i>	313
Therapeutic Management	314
Medical Treatment	314
<i>Antimicrobial Therapy</i>	314
<i>Additional Medical Therapy</i>	316
Surgical Treatment	317
Adenoidectomy	317
Antral Lavage, Nasal Antral Window	
Endoscopic Sinus Surgery	317
<i>Absolute Indications</i>	318
<i>Possible Indications</i>	318
Complications of Rhinosinusitis	318
Osteomyelitis of the Maxilla	318
Odontogenic Sinusitis	318
Orbital and Intracranial	
Complications	318
<i>Pott's Puffy Tumour</i>	318
<i>Osteomyelitis of the Posterior Wall</i>	
Orbital Complications	319
	320

Mucocoeles	323
36 Management of the Deaf Child	327
<i>Kevin Patrick Gibbin</i>	
Introduction	327
Aetiology and Pathology of Childhood Deafness	328
Pre-natal Causes	328
Perinatal Causes	328
Post-natal Causes	329
Epidemiology	329
Risk Factors for Hearing Loss in Children	330
Identification of Hearing Loss	330
Screening – Targeted versus Universal Early Childhood/Pre-school Programmes	330
Monitoring of Special Groups	331
<i>Post-Meningitis Screening</i>	331
Reactive Testing	331
Investigation	331
<i>Clinical History Taking</i>	331
<i>Clinical Examination</i>	332
<i>Special Investigations</i>	332
Audiological Assessment	333
Treatment of Children with Hearing Loss	333
37 Audiometric Testing of Children	337
<i>John Graham, Milanka Drenovak and William Hellier</i>	
Introduction	338
Neonatal Tests Used From Soon After Birth Until Six Months	339
Transient Otoacoustic Emission Testing	339
<i>Benefits</i>	339
<i>Limitations</i>	339
<i>Use</i>	339
Distortion Product Otoacoustic Emissions	339
Auditory Brainstem Response	339
<i>Benefits</i>	341
<i>Limitations</i>	341
<i>Use</i>	341
Electrocochleography	342
<i>Benefits</i>	342
<i>Limitation</i>	342
Behavioural Testing: Ages Six Months to Three Years	342
Distraction Testing: Age Six to Eighteen Months	342
<i>Benefits</i>	343

<i>Limitations</i>	343
<i>Use</i>	343
Visual Reinforcement Audiometry:	
Age Six Months to Three Years	344
<i>Benefits</i>	344
<i>Limitations</i>	344
<i>Use</i>	344
Conditioned Audiometry:	
Two to Three Years and Upwards	344
Play Audiometry:	
Age Two to Three Years and Upwards	344
<i>Benefits</i>	345
<i>Limitations</i>	345
Speech Audiometry:	
Age Two to Six Years and Upwards ..	345
<i>Benefits</i>	346
<i>Limitations</i>	346
Cortical Electric Response Audiometry	346
Audiometric Steady-State Response	347
<i>Benefits</i>	348
<i>Limitations</i>	348
<i>Use</i>	348
Tympanometry and Acoustic Reflexes	348
Special Situations	349

38 Otoplasty and Common Auricular

Deformities	351
<i>Werner J. Heppt</i>	
Introduction	351
Basics	352
Development of the Auricle	352
Clinical Anatomy	352
Classification of Congenital Deformities	353
Protruding Ears (Prominent Ears, Bat Ears)	353
Causes	353
Conservative Treatment Options	353
Surgical Correction (Pinnaplasty) ...	354
<i>Goals</i>	354
<i>Choice of Appropriate Method</i>	354
<i>Antihelix</i>	354
<i>Conchal Hyperplasia, Increased</i>	
<i>Conchomastoid Angle</i>	356
<i>Prominent Lobule</i>	357
<i>Severe Deformities, Revision Surgery</i>	357
Operative Management	357
<i>Timing</i>	357
<i>Medical Consultation</i>	357
<i>Peri- and Post-operative Care</i>	357
<i>Results</i>	357
Trauma	358
Abrasion	358
Haematoma	358
Laceration	358
Thermal Injuries, Frostbite	359

39	Diagnosis and Management Strategies in Congenital Middle and External Ear Anomalies	361
	<i>Frank Declau, Paul Van De Heyning and Cor Cremers</i>	
	Introduction	362
	Epidemiology	362
	Classification	363
	Ossicular Malformation	363
	Atresia Classification	363
	Microtia Classification	364
	Patient Evaluation	364
	History and Physical Examination	364
	Audiometric Evaluation	365
	Radiological Assessment	365
	Medical Management	365
	Unilateral Atresia	366
	Bilateral Atresia	366
	Surgical Management	366
	Ossicular Malformation Surgery	366
	Atresia Repair Surgery	366
	<i>Surgical Planning</i>	367
	<i>Timing of Surgery</i>	368
	<i>Surgical Techniques</i>	368
	<i>Surgical Results</i>	369
	<i>Complications</i>	370
	Bone Conduction Implant Surgery	370
	Total Auricular Reconstruction	
	Surgery	370
	<i>Surgical Planning</i>	370
	<i>Surgical Techniques</i>	370
	<i>Comparison of the Two Techniques</i>	372
	<i>Other Techniques</i>	372
	<i>Results in Total Auricular Reconstruction</i>	372
	<i>Complications in Total Auricular Reconstruction</i>	372
	<i>Complications at the Donor Site</i>	372
	<i>Complications at the Graft Site</i>	372
	Prosthetic Auricular Reconstruction	373
	<i>Indications</i>	373
	<i>Results and Complications of Prosthetic Reconstruction</i>	373
40	Imaging of the Deaf Child	377
	<i>Timothy J. Beale and Gitta Madani</i>	
	Part 1. A Radiological Overview of the Imaging of Children with Hearing Loss	378
	Key Points of CT Imaging	378
	<i>CT Imaging Protocols in Children</i>	378
	Key Points of MRI	378
	<i>MRI Protocols in Children</i>	378
	Sedation	379

CT: Normal Cross-Sectional Anatomy	379
<i>Systematic Review of CT Images</i>	379
Normal MRI Anatomy	381
<i>Systematic Review of MRI Images</i> ..	381
When is Early Imaging Essential?	382
Imaging Algorithms for Conductive, Sensorineural and Mixed Hearing Loss ...	382
<i>Conductive Hearing Loss</i>	382
<i>Sensorineural Hearing Loss</i>	382
<i>Mixed Hearing Loss</i>	382
Part 2. Radiological Abnormalities	
Found in Children with Hearing Loss ..	382
Embryology of the Inner and Middle Ear and the EAC	382
<i>Inner Ear</i>	382
<i>Middle Ear</i>	382
<i>External Ear</i>	382
Conductive Hearing Loss	383
Radiological Features of Congenital Aural Dysplasia	383
Congenital Middle Ear Anomalies ...	383
Syndromes Associated with Congenital CHL	384
<i>Otocraniofacial Syndromes</i>	384
<i>Craniosynostoses</i>	386
<i>Otocervical Syndromes</i>	386
Acquired CHL	386
<i>Trauma: Ossicular Disruption</i>	386
<i>Tympanosclerosis</i>	388
Sensorineural Hearing Loss	388
Congenital SNHL	388
<i>Abnormal Development of the Inner Ear</i>	388
<i>Syndromes Associated with Congenital SNHL</i>	390
<i>The Otoskeletal Syndromes (Bone dysplasias)</i>	392
Acquired SNHL	395
<i>Post-Meningitic Labyrinthitis Obliterans</i>	395
The Future	395

41 Acute Otitis Media in Children 399

Ingeborg Dhooge

Introduction	400
Definitions	400
<i>Otitis Media</i>	400
<i>Acute Otitis Media</i>	400
<i>AOM Without Effusion (Myringitis)</i>	400
<i>Recurrent Otitis Media</i>	400
<i>Otorrhoea</i>	400
Epidemiology	400
Predisposing Factors and Environmental Pressure	400
Host-Related Parameters	400

Age	400
Gender	401
Race	401
<i>Altered Host Defences and Underlying Diseases</i>	401
<i>Genetic Predisposition</i>	401
<i>Prenatal and Perinatal Factors</i>	401
Environmental Factors	401
<i>Climate and Seasons</i>	401
<i>Day Care/Home Care, Exposure to Other Children</i>	401
<i>Passive Smoking and Environmental Pollution</i>	401
<i>Socio-Economic Status/Poverty</i>	401
<i>Use of Dummies (Pacifiers)</i>	401
Pathogenesis of otitis media	402
Eustachian Tube Dysfunction	402
Immature Immune Response	402
Microbial Infection	403
Natural History and Complications	403
Acute Perforation	403
Suppurative Complications	403
Intratemporal (Extracranial) Complications of Otitis Media	404
<i>Subperiosteal Abscess</i>	404
<i>Facial Palsy</i>	404
<i>Labyrinthitis</i>	404
Intracranial Complications of Otitis Media	404
Assessment and Diagnosis	405
Symptoms and Signs	405
Otoscopy	405
Conventional Tympanometry	405
Conventional Audiometry	405
Other Diagnostic Procedures	405
Imaging	406
Treatment	406
Medical	406
<i>Symptomatic Treatment</i>	406
<i>Antibacterial Treatment</i>	406
<i>“Alternative” Medical Treatments</i> ...	408
Surgical Therapy	408
<i>Myringotomy</i>	408
<i>Tympanostomy Tube Insertion</i>	408
<i>Adenoidectomy</i>	408
Follow-Up	409
Prevention of AOM	409
Sequelae	409

42 Otitis Media With Effusion 413

Peter J. Robb

Introduction	413
Epidemiology	414
Birth Order	414
Gender	414

Breast-Feeding	414
Day-Care	414
Parental Smoking	414
Infection	414
Immune Deficiency and Allergy	415
Reflux of Gastric Acid	415
Symptoms and Signs of OME	415
Diagnosis	415
Audiology	416
Natural History	416
Complications	417
Treatment	417
Medical	417
Hearing Aids	417
Surgical	417
<i>Grommet Surgery (Ventilation</i>	
<i>Tubes)</i>	417
<i>Adenoidectomy</i>	418
Complications of Surgical Management	418
Premature Extrusion of the	
Grommets and Perforation	418
Otorrhoea	418
Myringosclerosis (Tympanosclerosis)	419
Atelectasis	419
Post-adenoidectomy Haemorrhage ..	419

43 Chronic Otitis Media 421

John Hamilton

Introduction	422
Histology of Chronic Inflammation	
of the Middle Ear Mucosa	423
Perforation of the Tympanic Membrane	
and Chronic Suppurative Otitis Media	423
Epidemiology	423
Symptoms	424
Signs	425
Investigation	425
Treatment	425
<i>Prophylaxis</i>	425
<i>Medical Treatment</i>	426
<i>Surgical Closure of Tympanic</i>	
<i>Membrane Perforations in Children</i>	
<i>with CSOM (Tympanoplasty)</i>	426
Tympanic Membrane Atrophy	
and Retraction Pockets: Pars Tensa	427
Pathology	427
Epidemiology	427
Classification	428
Natural History	429
Symptoms	429
Principles of Surgical Intervention	
for Tympanic Membrane Atrophy	
and Retraction Pockets	430
Surgical Treatment of Tympanic	
Membrane Atrophy	430

Grommet Insertion	430
Reinforcement Tympanoplasty	430
Retraction Pocket Excision	430
Attic Retraction Pockets	430
Cholesteatoma	432
Pathogenesis and Classification	432
<i>Congenital Cholesteatoma</i>	432
<i>Acquired Cholesteatoma</i>	433
Epidemiology	433
Anatomy of Paediatric Cholesteatoma	433
Treatment	434
<i>Medical</i>	434
<i>Imaging</i>	434
<i>Surgical</i>	435
<i>Surgical Approaches for the Removal of Cholesteatoma and Restoration of a Stable, Dry Ear</i>	435
Residual and Recurrent Cholesteatoma	436
<i>Residual Cholesteatoma</i>	436
<i>Recurrent Cholesteatoma</i>	437
Changes in the Ossicles Secondary to Chronic Middle Ear Inflammation ..	437
Epidemiology	437
Pathology	437
Symptoms	437
Signs	437
Diagnosis	438
Treatment	438
Outcome Measures	438
<i>Factors that Influence the Success of Ossicular Reconstruction</i>	438
Complications of CSOM in Children ..	439

44 Bone-Anchored Hearing Aid (BAHA) and Softband	441
<i>David W. Proops</i>	
Introduction	441
Conductive Hearing Loss in Children ..	442
Management of Congenital Conductive Hearing Loss	442
Classification of Congenital Ear Disorders	442
Investigations of Congenital Middle and External Ear Malformations	442
Aiding Options for Congenital Conductive Hearing Loss	443
Surgical Management of Congenital Conductive Hearing Loss	443
Reconstructive Surgery of the External and Middle Ear	443
The Bone-Anchored Hearing Aid	443
<i>BAHA Surgical Procedure</i>	443
<i>Results in Bilateral Congenital Conductive Hearing Loss</i>	444

<i>Unilateral Congenital Conductive</i>	
<i>Hearing Loss</i>	444
<i>Fixture Survival and Skin Care</i>	444
Providing a Service for Children	
with Congenital Conductive Deafness	444
Softband	445
45 Cochlear Implantation in Children	447
<i>Mary Beth Brinson and John Graham</i>	
Introduction	448
Team Approach	449
Referral Guidelines	449
Group I: Congenital Profound	
Hearing Loss	450
Group II: Progressive Hearing Loss ..	450
Group III: Severe-to-Profound	
Bilateral Hearing Loss	450
Group IV: Sudden Hearing Loss	451
Assessment	451
Audiological Scientist	452
Speech and Language Therapist	452
Teacher of the Deaf	452
Psychologist	452
Medical Team Members	453
Parents	453
Implantation Criteria	453
The Child	453
The Child's Environment	453
Factors Affecting the Outcome	
of Cochlear Implantation	453
Surgery	454
Approach	454
Preparation of the Bed	
for the Receiver-Stimulator Package	454
Insertion of the Electrode Array	454
Special Medical and Surgical Situations	455
Jervell and Lange-Nielsen Syndrome	455
Meningitis	455
Prematurity	455
Otitis Media	455
Ossified Cochleae	455
Congenital Malformation	
of the Cochlea	456
Surgical "Gusher"	456
Chronic Suppurative Otitis Media ...	456
Complications After Implantation	456
Device Failure	456
Infection	457
Skin Breakdown	457
Facial Nerve Stimulation	457
Further Considerations	457
Bilateral Cochlear Implantation	
in Children	457
Future Prospects	457

46 The Dizzy Child	459
<i>Linda Luxon and Waheeda Pagarkar</i>	
Introduction	460
Clinical Evaluation	461
History	461
General Examination	462
Eye Movement Examination	463
<i>Dix-Hallpike Positional Test</i>	463
<i>Halmagyi Head Thrust Sign</i>	464
<i>Head-Shaking Nystagmus</i>	464
<i>Dynamic Visual Acuity</i>	465
Tests for Posture Control and Gait ...	465
Neurological Examination	466
<i>Investigations for the Dizzy Child</i> ..	466
Other Tests	467
Causes of Vertigo/Dizziness/Imbalance in Children	469
Congenital Disorders	469
<i>Syndromic Hearing Loss and</i> <i>Vestibular Dysfunction</i>	469
<i>Nonsyndromic Hearing Loss</i> <i>and Vestibular Dysfunction</i>	470
Otological	470
<i>Inner Ear Infection or Inflammation</i>	470
Traumatic	471
<i>Head Injury</i>	471
<i>Benign Paroxysmal Positional</i> <i>Vertigo</i>	471
<i>Perilymphatic Fistula</i>	472
<i>Cochlear Implant Surgery</i>	472
Idiopathic	472
<i>Menière's Disease</i>	472
Ototoxicity	472
Neurological	472
<i>Migraine and Migraine Precursors</i>	472
Episodic Ataxia Type 2	474
Neoplasia	475
Cardiovascular Causes	475

<i>Orthostatic Hypotension</i>	475
Miscellaneous Causes	475
<i>Ocular Disorders</i>	475
Psychological Causes	475
<i>Vertigo of Psychogenic Origin</i>	475
Management of Dizziness	475
Treatment of Paediatric Migraine	476
Vestibular Management	476

47 The Facial Nerve 479

Chris Rittey

Anatomy	479
Upper Motor Neurone Input	479
Clinical Assessment	480
Isolated Facial Weakness	481
Bell's Palsy	481
Rare Causes of Isolated Unilateral	
Facial Palsy	481
Congenital Facial Palsy	482
Facial Palsy as Part of Other Cranial	
Nerve Palsies	482
Ramsay Hunt Syndrome	482
Hemifacial Spasms	483
Developmental Data	485
Airway	485

Appendix 485

Robert J. Ruben

Sinuses	486
The Adenoid	486
Language and Speech	486

Subject Index 489