

Contents

VOLUME 1

PART I. GENERAL

1 / Genetics and Dysmorphology by <i>Kenneth N. Rosenbaum</i>	3
Frequency and Impact of Genetic Disorders	3
Dysmorphology—Definitions	3
Principles of Genetics	3
Assessment of the Child With Malformations	5
Selected Genetic Disorders and Syndromes Featuring Surgically Treatable Malformations	6
Chromosomal Aberrations	7
Counseling in Cases of Isolated Surgically Treatable Malformations	8
2 / Teratology by <i>Henry L. Nadler</i>	11
General Principles	11
Categories of Teratogens	11
Summary	13
3 / Prenatal Diagnosis by <i>Michael R. Harrison</i>	14
Amniocentesis and Genetic Counseling	14
Fetoscopy and Fetal Sampling	15
Prenatal Diagnosis and Perinatal Management	16
4 / Fluid and Electrolyte Management by <i>Marc I. Rowe</i>	22
Fetal and Neonatal Physiology	23
Arriving at the Tentative Program—The Rough Draft	27
5 / Management of the Critically Ill Patient by <i>Marc I. Rowe, Barbara J. Pettitt</i>	31
Monitoring	31
Shock	39
6 / Anesthesia by <i>Eugene K. Betts, John J. Downes</i>	50
The Risks of Anesthesia	50
Principles of Pediatric Anesthesia	51
Results	65
7A / Respiratory Support—Ventilators by <i>Russell C. Raphaely</i>	68
Oxygenation Failure	68
Ventilation Failure	68
Respiratory Support Options	68
Techniques	68
Elevation of Transpulmonary Distending Pressure	68
Inspiratory Pressure Elevation (Mechanical Ventilation)	69
High-Frequency Ventilation	70
Management of Acute Respiratory Failure	71
Complications of Respiratory Support	71
Chronic Respiratory Failure	71
7B / Respiratory Support—Extracorporeal Membrane Oxygenation in Newborn Respiratory Failure by <i>Robert H. Bartlett</i>	74
Technique	74
Results	75
8 / Surgical Infections by <i>Ann M. Kosloske, H. Harlan Stone</i>	71
Components of Infection	71
Host Deficiency States	81
Specific Types of Surgical Infection	81
Sepsis in the Newborn	81
Antibiotics	81

- 9 / **The Immunocompromised Child** by *Fred S. Rosen*
 B-Cell Deficiencies
 T-Cell Immunodeficiencies
 Complement Defects
 Phagocytic Defects
- 10 / **Nutrition of the Surgical Patient** by *Arnold G. Coran*
 Nutritional Requirements
 Metabolic Response of the Full-Term Infant to Operation and Stress
 Nutritional Assessment
 Enteral Feeding
 Total Parenteral Nutrition
 Short-Gut Syndrome
 Home Parenteral Nutrition
- 11 / **Surgical Implications of Hematologic Disorders** by *Sherwin V. Kevy*
 Nonhemolytic Anemias
 Hemolytic Anemias
 Platelets
 Nonthrombocytopenic Purpura
 Coagulation
 Transfusion Therapy
- 12 / **New Imaging Technologies** by *Richard M. Heller*
 The New Modalities
- 13 / **Psychological Implications of Operations in Children** by *Stephen P. Hersh*
 Biopsychosocial Principles
 Psychosocial Principles
 Principles of Treatment
 Elective Operation
 Emergency Operation
 Operation and the Exceptional Child
 Disturbed and Disturbing Behaviors
 Use of Medications
 Glossary of Selected Terms

PART II. TRAUMA

- 14 / **Infants and Children as Accident Victims** by *James A. O'Neill, Jr*
- 15 / **Emergency Management of the Injured Child** by *James A. O'Neill, Jr*
 Initial Assessment and Management of Life Support
 Continuing Care of the Stable Patient
 Priorities in Emergency Treatment
- 16 / **Child Abuse** by *James A. O'Neill, Jr*
 Incidence and Epidemiology
 Diagnosis
 Lesions Associated With Physical Abuse
 General and Legal Considerations
- 17 / **Thoracic Injuries** by *J. Alex Haller, Jr*
 Primary Evaluation and Therapy
 Specific Initial Resuscitative Measures
 Treatment Approach
 Potentially Lethal Chest Injuries
 Less Dangerous Chest Injuries
- 18 / **Abdominal Trauma** by *Martin R. Eichelberger, Judson G. Randolph*
 Evaluation of Intra-abdominal Injury
 Exploratory Laparotomy
- 19 / **Genitourinary Injuries** by *Howard McC. Snyder III, Anthony A. Caldamone*
 Renal Trauma
 Ureteral Injuries
 Bladder Injuries
 Urethral Injuries
 Injuries to the External Genitalia
- 20 / **Soft Tissue Injuries** by *Donato LaRossa, Linton A. Whitaker*
 Wound Management
 Management of Specific Injuries
 Injuries With Partial or Complete Soft Tissue Loss

21 / Musculoskeletal Trauma by Richard S. Davidson, Hugh G. Watts	189
Differences in the Musculoskeletal System Between Children and Adults	189
Initial Assessment: Physical Examination	191
Management	193
Complications and Pitfalls	195
22 / Management of Acute Hand Injuries by Richard G. Eaton, John J. Keyser	197
Evaluation of the Injured Hand	197
Treatment	197
Tendon Injury	199
Nerve Injury	201
Skeletal Injuries	201
23 / Snake and Insect Bites by William L. Buntain	203
Snakebites	203
Insect (<i>Hymenoptera</i>) Stings	207
Spider (<i>Arthropod</i>) Bites	208
24 / Central Nervous System Injuries by Derek A. Bruce	209
Diffuse Injuries	209
Focal Injuries	210
Birth Trauma	212
Emergency Medical Management	213
Medications	214
Spinal Cord Injury	215
25 / Vascular Injuries by James A. O'Neill, Jr	216
Causes of Injury	216
Diagnosis	217
Preoperative Management	218
Technical Considerations	218
Results and Complications	219
26 / Burns by James A. O'Neill, Jr	220
Care of Minor Burns	221
Care of Major Burns	221
Treatment	222
Complications Associated With Burn Injury	222
Results	222
27 / Birth Trauma by Peter K. Kottmeier	230
Infants at Risk	230
Nerves	231
Fractures	232
Respiratory Distress	233
Abdominal Distention	234

PART III. PRINCIPAL MALIGNANT TUMORS

28 / Cancer Chemotherapy by Norman Jaffe	240
Indications for Chemotherapy	240
Classification of Chemotherapeutic Agents	241
Vinca Alkaloids	244
Miscellaneous Agents	244
Combination Chemotherapy and Protocol Treatment	246
29 / Radiation Therapy for Solid Tumors by Giulio J. D'Angio	247
General Principles	247
Treatment Modalities	248
Radiation Modifiers	248
Radiation Therapy of Specific Conditions	248
Wilms Tumor	250
Neuroblastoma	250
Soft Tissue Sarcomas	251
Other Soft Tissue Tumors	251
Gonadal Tumors	251
Bone Tumors	255
30 / Hodgkin's and Non-Hodgkin's Lymphoma by Jessie L. Ternberg	256
Hodgkin's Disease	256
Non-Hodgkin's Lymphoma	256
31 / Teratoma by Morton M. Woolley	260
Experimental Embryology	260
Human Embryology	261

	Endocrinopathy and Serum Tumor Markers	
	Clinical Aspects of Teratomas	
	Sacrococcygeal Teratomas	
	Genetic Presacral Teratomas With Associated Sacral, Anal, and Urinary Anomalies	
	Abdominal Teratomas	
	Gastric Teratomas	
	Retroperitoneal Teratomas	
	Thoracic Teratomas	
	Intrapericardial Teratomas	
	Myocardial Teratomas	
	Cervical Teratomas	
	Epignathi	
	Intracranial Teratomas	
32 /	Rhabdomyosarcoma by <i>Daniel M. Hays</i>	
	Incidence and Sites	
	Etiology and Epidemiology	
	Pathology	
	Development of Combined Therapy Programs	
	Clinical Management	
	Characteristics of Tumors in Specific Sites	
	Summary	
33 /	Neuroblastoma by <i>Jay L. Grosfeld</i>	
	Clinical Presentation	
	Diagnosis	
	Immunologic Aspects	
	Staging	
	Therapy	
	Role of Second and Delayed Primary Operations	
	Prognosis	
	Future Directions	
34 /	Wilms Tumor (Nephroblastoma) by <i>H. Biemann Othersen, Jr.</i>	
	Pathogenesis and Epidemiology	
	Pathology	
	Diagnosis	
	Operative Technique	
	Postoperative Care	
	Metastatic Disease	
	Second Neoplasms	
	Bilateral Tumors	
	Mesoblastic Nephroma	
	Results and Conclusions From the National Wilms Tumor Study	
	Future Directions	
35 /	Tumors of the Liver by <i>Judson G. Randolph, Philip C. Guzzetta</i>	
	Diagnosis of the Child With a Suspected Hepatic Mass	
	Benign Tumors of the Liver	
	Malignant Tumors of the Liver	
	Technique of Liver Resection	
36 /	Bone Tumors by <i>Hugh G. Watts, Richard Lackman</i>	
	Malignant Tumors	
	Benign Tumors	
37 /	Tumors of the Brain and Spinal Cord by <i>George B. Udvarhelyi, Jeffrey A. Winfield</i>	
	Brain Tumors	
	Spinal Cord Tumors	

PART IV. TRANSPLANTATION

38 /	Principles of Whole Organ Transplantation by <i>Thomas E. Starzl, Byers W. Shaw, Jr., Shunzaburo Iwatsuki</i>	
	Immunosuppression	
	Tissue Typing	
	Organ Procurement and Preservation	
39 /	Kidney Transplantation by <i>John S. Najarian, Nancy L. Ascher, S. Michael Mawer</i>	
	Selection and Evaluation of Patients for Renal Transplantation	
	Diseases Leading to Kidney Transplantation	
	Evaluation of the Potential Recipient	
	Relative Indication for Hemodialysis, Peritoneal Dialysis, or Transplantation	
	Preparation of the Patient for Transplantation	
	Donor Selection	

Pretransplant Transfusion	364
Anesthesia in the Anephric Patient	364
Intraoperative Management and Surgical Technique	364
Routine Postoperative Care	364
Immunosuppressive Therapy	365
Complications in the Postoperative Period	366
Late Post-Transplant Problems	367
Recurrence of the Original Disease in the Transplanted Kidney	368
Psychological Considerations	369
Rehabilitation	370
Cancer	371
Results	372
40 / Liver Transplantation by <i>Thomas E. Starzl, Shunzaburo Iwatsuki, Byers W. Shaw, Jr.</i>	373
The Laboratory Origins	373
The Pace of Clinical Trials	374
Indications for Liver Transplantation	374
Surgical Technique	377
Immunosuppression and Survival	378
The Place of Liver Transplantation in Pediatric Hepatology	381
41 / Heart and Heart-Lung Transplantation by <i>Bartley P. Griffith</i>	383
Cardiac Transplantation	383
Compatibility of Donor-Recipient	384
Donor Criteria and Procurement	384
Operative Technique for Cardiac Transplantation	385
Immune Suppression	385
Surveillance for Rejection	386
Results of Cardiac Transplantation	386
Selection of Candidates for Heart-Lung Transplantation	386
Criteria for Heart-Lung Donor	387
Procurement of Heart-Lung Bloc	387
Operative Technique for Heart-Lung Transplantation	387
Results of Heart-Lung Transplantation	388
42 / Transplantation of Bone Marrow by <i>George W. Santos</i>	390
Selection of Marrow Donors	390
Preparative Regimens	391
Supportive Care	391
Current Clinical Results	392
Complications of Marrow Transplants	392
Conclusions	394
43 / Transplantation of Pancreas, Parathyroids, Intestine, and Other Organs by <i>Krith Reemtsma, Collin J. Weber</i>	395
Pancreas and Islet Transplantation	395
Parathyroid Transplantation	397
Adrenal Autotransplantation	398
Other Endocrine Tissues	398

PART V. HEAD AND NECK

44 / Ophthalmologic Disorders—Eyes and Lids by <i>John S. Crawford, Robert C. Pashby</i>	401
Injuries	401
Surgery of the Eyelids	402
Preoperative Examination	403
Operative Procedures	403
Surgical Treatment of Congenital Cataracts and Dislocated Lens	403
Operation for Obstructed Tear Ducts	404
Ophthalmologic Disorders—Retinoblastoma and Other Retinal Diseases by <i>Emily Y. Chew</i>	407
Diagnosis	407
Genetics of Retinoblastoma	407
Locus of Retinoblastoma Mutation	408
Retinomas	408
Second Nonocular Tumors	408
Pathology	408
Mortality	408
Investigations	408
Treatment	408
Extracocular Retinoblastoma	408
Follow-up	408
Differential Diagnosis	408

45 /	<i>Craniofacial Abnormalities</i> by Leonard B. Kaban, John B. Mulliken, Joseph E. Murray	4
	Congenital Deformities: Anatomy and Clinical Findings	4
	Correction of Congenital Craniofacial Anomalies	4
	The Future of Craniofacial Surgery	
46 /	<i>The Nose</i> By Ronald M. Zuker	4
	Embryology	
	Anatomy	
	Congenital Anomalies of the Nose	
	Tumors of the Nose	
	Nasal Trauma	
47 /	<i>Cleft Lip and Cleft Palate</i> by William K. Lindsay	
	Cleft Lip	
	Normal Lip Development	
	Cleft Lip Development	
	Epidemiology	
	Risk for Future Siblings	
	Historical Aspects	
	Neonatal Management	
	Age at Operation	
	Choice of Anesthesia	
	Neonatal Maxillary Orthopedics	
	The Unilateral Cleft Lip Repair	
	The Bilateral Cleft Lip Repair	
	Changes With Time in Bilateral Cleft Lip	
	Later Management of Patients With Unilateral Cleft Lip	
	Later Management of Patients With Bilateral Cleft Lip	
	The Role of Alveolar Bone Grafting	
	Cleft Palate	
	Embryology and Development	
	Classification	
	Applied Anatomy	
	Epidemiology	
	Treatment	
48 /	<i>Otolaryngologic Disorders</i> by Francis I. Catlin	
	Surgery of the Tonsils and Adenoids	
	Operative Treatment of Hearing Impairment	
	Choanal Atresia	
	The Infant Larynx	
49 /	<i>The Salivary Glands</i> by Kenneth J. Welch	
	Benign Neoplasms	
	Malignant Neoplasms	
	Mucoepidermoid Carcinoma	
	Undifferentiated Sarcoma	
	Adenocarcinoma	
	Acinic Cell Carcinoma	
	Embryonal Rhabdomyosarcoma	
	Miscellaneous	
	Discussion	
	Treatment	
	Ranula	
	Sialadenitis	
	Miscellaneous Lesions	
	Sialography	
	Techniques of Parotidectomy	
50 /	<i>The Oropharynx and Jaws</i> by Kenneth J. Welch	
	Congenital Anomalies of the Lip	
	Injuries	
	Hemangioma	
	Vascular Malformations	
	Lesions of the Jaw	
	Infections	
	Lesions of the Tongue and Oral Cavity	
	Lesions of the Nasopharynx	
	Cervicopharyngeal Teratomas of the Newborn	
51 /	<i>Glands of the Neck—Lymphadenitis</i> by Peter G. Jones	
	Acute Coccal Adenitis	
	Mycobacterial Cervical Adenitis	
	Cat-Scratch Disease	
	Glands of the Neck—Neoplasia of Cervical Nodes by Theodore C. Jewett, Jr	

52 / Thyroid and Parathyroid by Norman W. Thompson	522
The Thyroid	522
Parathyroid	530
53 / Cystic Hygroma by Mark M. Ravitch, Benjamin F. Rush, Jr.	533
Embryology of the Lymphatics	533
Natural History	534
Pathology	535
Clinical Course	537
Treatment	539
54 / Cysts and Sinuses of the Neck by Robert T. Super, Kevin C. Pringle	539
Embryology	543
Branchial Remnants	547
Midline Masses	550
Low Neck Masses	552
55 / Torticollis by Peter G. Jones	553
Pathology	553
Clinical Picture	554
Secondary Effects	554
Diagnosis	555
Management	555
Experience and Results	555

PART VI. THORAX

SECTION ONE: THE THORACIC PARIETIS	
56 / Disorders of the Breast by John H. Seashore	559
Congenital Anomalies	559
Inflammatory Diseases	559
Diffuse Enlargements of the Breast	561
Discrete Breast Masses	563
57 / The Chest Wall by Mark M. Ravitch	563
Congenital Deformities of the Chest Wall	568
Depression Deformities	578
Protrusion Deformities	581
Poland Syndrome	582
Rib Deformities in Diffuse Skeletal Disorders	583
Tumors of the Chest Wall	589
58 / Congenital Diaphragmatic Hernia by Kathryn D. Anderson	589
Embryology	590
Incidence	590
Mortality	590
Anatomy	591
Physiology of Pulmonary Circulation	592
Diagnosis and Clinical Presentation	593
Associated Anomalies	594
Factors Associated With Increased Mortality	594
Preoperative Treatment	595
Operative Treatment	596
Postoperative Care	596
Postoperative Physiology and Vasodilator Therapy	597
Results and Follow-up	597
New Therapeutic Approaches	599
Foramen of Morgagni Hernias	599
Eventration of the Diaphragm	599
Paraesophageal Hiatus Hernia	602
59 / Mediastinal Cysts and Tumors by Mark M. Ravitch	602
Anatomy	602
Thymus	605
Tumors in Mediastinal Lymph Nodes	606
Mediastinal Cysts and Tumors	606
Superior Vena Cava Syndrome	615
SECTION TWO: THE AIRWAY, LUNGS, AND PLEURA	
60 / Bronchoscopy by Dale G. Johnson	619
Setting	619
Equipment	619

Anesthesia	62
Procedure	62
Clinical Application	62
61 / Lesions of the Larynx and Trachea—Tracheostomy by Dale G. Johnson	62
Clinical Presentation	62
Diagnosis	62
Tracheostomy by Eoin Aberdeen, Dale G. Johnson	62
62 / Congenital Malformations and Neonatal Problems of the Respiratory Tract	63
by Alfred A. de Larosier	63
Respiratory Problems in the Newborn	63
Obstructions of the Upper Airway	63
Intrathoracic Airway Obstruction	63
Cysts and Tumors in the Mediastinum	63
Lobar Emphysema	63
Pulmonary Lesions	63
Pulmonary Agenesis, Aplasia, and Hypoplasia	63
63 / Lung Cysts, Sequestration, and Bronchopulmonary Dysplasia by William K. Sieber	63
Congenital Cysts of the Lung	63
Congenital Adenomatoid Malformation of the Lung	63
Pulmonary Sequestration	63
Bronchopulmonary Dysplasia	63
Congenital Bronchopulmonary Foregut Malformations With Gastric or Esophageal Communication	63
Prognosis After Pulmonary Resection in Children	63
64 / Chylothorax by Judson G. Randolph	63
Etiology	63
Embryology	63
Anatomy	63
Physiology	63
Clinical Findings	63
Treatment	63
Surgical Measures	63
65 / Infections of the Lungs, Pleura, and Mediastinum by Ann M. Kosloske	63
Lung Abscess	63
Bronchiectasis	63
Surgery in Cystic Fibrosis	63
Bronchiolitis	63
Mycobacterial and Fungal Disease	63
Parasitic Diseases	63
Inflammatory Pseudotumor	63
Open Lung Biopsy	63
Empyema	63
Infections of the Mediastinum	63
Esophageal Perforation	63
66 / Tumors of the Lung by Robert M. Filler	63
Primary Tumors	63
Metastatic Tumors	63

SECTION THREE: THE ESOPHAGUS AND MEDIASTINUM

67 / Esophagoscopy by Dale G. Johnson	64
Rigid Versus Flexible Instrumentation	64
Equipment	64
Patient Preparation, Sedation, and Anesthesia	64
Technique	64
Normal Esophagoscopy	64
Indications	64
Complications	64
68 / Esophageal Atresia and Congenital Stenosis—Esophageal Atresia and Associated Malformations, Including Laryngotracheoesophageal Cleft by Judson G. Randolph	64
Embryology	64
Genetic Relations	64
Associated Anomalies	64
Incidence	64
Esophageal Atresia With Lower-Segment Tracheoesophageal Fistula	64
Complications	64
Esophageal Atresia With Upper Pouch Fistula	64
Esophageal Atresia Without Fistula	64
Isolated (H-Type) Tracheoesophageal Fistula	64
Laryngotracheoesophageal Cleft	64

Congenital Stenosis of the Esophagus	695
<i>by Nainar A. Myers, Eoin Aberdeen, Judson G. Randolph</i>	696
Diagnosis	696
Treatment	698
69 / Corrosive Strictures of the Esophagus by William P. Tunell	698
Incidence	698
Etiology	698
Pathophysiology	699
Diagnosis	699
Treatment	701
70 / Replacement of the Esophagus by Kathryn D. Anderson	704
Indications	704
Selection of the "Ideal" Esophageal Substitute	705
Timing	705
The Colon	705
The Gastric Tube	707
71 / Gastroesophageal Reflux by J. Boix-Ochoa	711
Antireflux Barrier	711
Etiologic Factors in Gastroesophageal Reflux	711
Clinical Findings	711
Diagnosis	711
Treatment	711
72 / Neuromotor Esophageal Disorders by J. Laurance Hill	720
Etiology	720
Achalasia	720
Diffuse Esophageal Spasm	721
Vigorous Achalasia	721
Management	721
Scleroderma	724
73 / Esophageal Rupture and Perforation by Thomas R. Weber	727
Classification and Incidence	727
Clinical Presentation	728
Diagnosis	728
Treatment	728
Results of Therapy	729
Index	xxviii